

Vacuum assisted closure for the treatment of pelvic pseudotumors in patients with hemophilia

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Introduction: Hemophilic pseudotumor is a rare complication occurring in 1–2% patients with hemophilia, especially in those presenting severe cases of hemophilia and those who have developed antibodies against factor VIII or IX. In such cases, surgical procedure has to be performed under the supervision of a hematologist in order to manage the proper intravenous factor infusion.

Materials and Methods: We present a case of a twenty year old hemophilic patient diagnosed with hemophilic pseudotumor in the left iliac wing, totally destroyed. Surgical management with vacuum assisted closure (VAC) and articular reconstruction, artery embolization, radiotherapy and factor infusion were used.

Results: Optimal results were achieved in the post surgical stage: patient reported a reconstruction of approximately 50% at three months and approximately 80% at six months.

Conclusions: Few reports on such cases are found in literature which allows us to modify and innovate several techniques, with a correct and thorough performance of a presurgical planning on an individual basis and using a technique in which soft tissues are mostly preserved.

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

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Introduction and Objectives: Haemophilic arthropathy is an important cause of morbidity in patients with severe haemophilia. The ankle joint is the second most common morbid site, and joint pain and stiffness are the most frequent findings. The stiffness of the ankle joint is the most functional disabling sequelae of gait cycle. The main causes of the joint stiffness are osteophytes bone block and the surrounding soft tissue contracture. Osteophytes bone block that occur along the anterior lip of the distal tibia and the neck of talus is a consistent radiographic finding in the ankle joint of haemophilia. The purpose of the present study was to investigate the role of anterior osteophytes bone block to cause limitation of the ankle motion in haemophilic patients.

Material and Methods: Thirty seven cases (Thirty five patients) were enrolled. Two patients had bilateral procedures. Mean age was 30.7 years. Male was thirty four and female was one. Haemophilia A was 28 patients, B was six patients, and one was factor VII deficiency. A mean follow-up was 29.3 months (10 to 83). No patient was lost to follow-up.

Results: Osteophytes resection was carried out through an anterior ankle approach. Some of patients were combined with joint debridement, synovectomy, and articular cartilage shaving. The mean AOFAS (American Orthopedic Foot and Ankle Society) score had improved from 62.5 points preoperatively to 82.6 points at the final follow-up ($P < 0.01$). The mean VAS decreased from 8.2 pre-operatively to 1.6 at the final follow-up ($P < 0.01$). The mean dorsiflexion increased significantly from 4.1° pre-operatively to 10.2° at final follow-up ($P < 0.01$). Mean plantar flexion improved from 35.3° pre-operatively to 39.4° at final follow-up ($P < 0.01$). A total of 30 patients (89.2%) were very satisfied or satisfied with the outcome.

Conclusion: Anterior osteophytes formation was very common, and impingement between the anterior lip of the distal tibia and the head-neck junction of the talus by osteophytes was one of the main causes to block ankle dorsiflexion. Anterior osteophytes resection of the ankle joint was very effective and valuable surgical procedure to improve function of the ankle joint in haemophilic patients.

Femoral neck fracture in hemophilia patients – A single center experience

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Introduction: Femoral neck fractures are frequent injuries especially in elderly population. Because of presence of osteoporosis, arthropathy and impaired stability, patients with hemophilia are at increased risk of bone fractures. In contrast to general population, data about incidence, clinical course and management of femoral neck fracture in hemophilia patients are scarce in the literature.

Methods: In the period from October 2012 to October 2015 five hemophilia patients with femoral neck fracture were treated in our institution. We present clinical course and management of these five patients.

Results: Five patients, 4 with hemophilia A, and one with hemophilia B, mean age 44 years (range from 25 to 58) suffered femoral neck fracture. Three patients were with severe hemophilia, one with mild and one with moderate. In all patients fracture occurred after trivial trauma. Four patients presented within a few hours after the injury, with Garden classification grade I. One patient presented 2 months after the injury with a Garden grade IV. Three patients were treated with opened reduction and fixation, one with femoral head replacement, while one patient with hemophilia B was treated conservatively. Osteodensitometry was performed in 3 hemophilia A patients, and significant osteoporosis has been found in all of them. Average time of hospital stay of operatively treated hemophilia A was 30 days with mean total consumption of FVIII concentrate 992 IU/Kg per patient. Total consumption of FIX concentrate for

conservative treatment of the hemophilia B patient was 1333 IU/kg. All four operated patients recovered completely, while patient who was treated conservatively developed limb deformity and gait disability.

Conclusion: With the increased life expectancy in hemophilia patients, the incidence of femoral neck fracture is expected to increase in the future. According our experience, in hemophilia patients femoral neck fracture may occur after trivial trauma and at relatively young age what is rare in general population. Under appropriate coverage with factor concentrate orthopedic treatment should not be delayed and may be conducted in the similar way as in patients without hemophilia. Early diagnosis and management of osteoporosis plays important role in prevention of femoral neck fracture in hemophilia patients.

Drain after total knee arthroplasty in patients with hemophilia: A necessary practice?

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Introduction and Objectives: There is controversy around using drain in total knee arthroplasty (TKA) especially in patients with hemophilia. Use of drain is proclaimed to reduce the rate of hematoma (which will impede rehabilitation), wound problem, limitation in range of motion and infection. We conceived this study to see if no-drain-protocol has any effect on the outcome of TKA in hemophilia.

Materials and Methods: In a prospective study, we compared the results of drain-protocol (42 TKAs in 39 patients, mean age 35.5 years) with no-drain-protocol (38 TKAs in 27 patients, mean age 35.7 years). Peri-operative variables (the level of reported pain, rate of complications, estimated blood loss, ...) was observed in the two groups. The time (days) to regain a 90 degree of range of motion was also registered. Patients were followed for at least 12 months.

Results: There was no statistical difference between two groups in terms of knee scores, blood loss, postoperative pain, fever, time to regain the range of motion and infection. Two patients in drain group and one patient in no drain group were re-operated because of peri-prosthetic joint infection. No patients needed blood transfusion in each group.

Conclusion: Our study suggests that 'no drain' protocol is safe after TKA in hemophilia. It decreases the cost of surgery and facilitates ambulation. Further prospective randomized study is necessary to determine the exact role of drain in TKA in hemophilia.

Long term follow-up on activity level of multiple joint procedures based on case series

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Introduction: Elective orthopedic surgery is a well-accepted option for the treatment of end stage haemophilic arthropathy (HA). If conservative treatment no longer guarantees participation in society, orthopaedic surgery has to be considered. Single procedures will lead to pain relief in one joint only and limitations in activities and participation remain. Long-term effects of multiple surgeries on activities (self-reported and performance-based) have not been reported. A case series was used to define which measurements on activity level are suitable to complete follow-up.

Materials and Methods: Multiple joint procedures (MJP) are defined as any combination of Total Hip Arthroplasty (THA), Total Knee Arthroplasty (TKA) and ankle arthrodesis (AA) during one hospital admission (one session or staged). Pre-and post-operative data from three patients were selected – based on availability of pre-operative assessments on activities – including the Hemophilia Activities List (HAL) [sum score 0–100 points, optimum 100], the McMaster Toronto Arthritis patient disability questionnaire (MACTAR), the 50 meter walking test (50MWT), the Timed Up & Go test (TUG), Figure 8 test and walking up and down a standardized stairs (4 steps).

Results: Included were two cases from subgroup bilateral TKA and one of bilateral AA; all had severe haemophilia A; mean age at time of surgery was 57 years old; mean follow-up was 6 years (range 4 – 8). The HAL showed improvement in both basic (23–40 points) as well as complex lower extremity activities (3.5–40 points) with an effect size over one standard deviation. The MACTAR showed individual progress in all patients, with emphasis on walking, standing, riding a bicycle and walking stairs. Performance tests all improved: 50MWT and TUG improved by a mean of 3 s, Figure 8 test by 1.4 s. Walking up and down the stairs showed a more fluctuating pattern.

Conclusion: In addition to measuring outcome of MJP on body and participation level (*Haemophilia*, 2014, 20 (2):276–81) we recommend to measure on activity level the HAL, 50 MWT, Getup & Go and Figure 8 (preferred and maximum speed).

Factors associated with perioperative complications in orthopedic surgery for patients with hemophilia

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Introduction and Objectives: Hemophilic arthropathy causes pain and restriction of range of motion, and results in an impairment of activity of daily living. When conservative treatments proved to be ineffective, orthopedic surgery is required for patients with severe hemophilic arthropathy. However, surgery for patients with hemophilia is challenging due to high complication rate such as infection, delayed wound healing and mortality. The aim of this study was to assess the incidence of