Orthopaedic procedures in haemophilia

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Summary

Haemophilia may nowadays be considered an “orthopaedic” disease given due to the involvement of musculoskeletal system in almost all haemophilic subjects. The modern haematological prophylaxis has dramatically improved the quality of life reducing bleedings and life-threatening complications; however, joint bleedings, progressive and irreversible arthropathy and osteoporosis are still now common challenging issues to be faced. One of the tissues involved by Haemophilia is the bone, particularly in the periarthicular zone: poor bone quality and decrease of bone stock are typical patterns, and the worse is the arthropathy, the greater the bone loss. The orthopaedic management of such condition is now mandatory and characterized by several surgical techniques. The purpose of this work is to provide an overview of these options derived from our experience in managing haemophilic patients.

KEY WORDS: bone; bone loss; bone grafting; orthopaedic management; haemophilia; knee arthroplasty; revision; joint fusion.

Introduction

Haemophilia is the most common bleeding disorder, related to a congenital or acquired deficiency of factor VIII or IX. Decades ago it was associated with fatal events due to massive bleedings, even in children (1). Nowadays, thanks to a better knowledge and advances in the haematological substitutive therapy and prevention, such conditions are uncommon. On the other hand, the most common complication is the musculoskeletal bleeding, particularly in specific joints, the so-called “target joints”. Haemophilic patients, particularly those suffering from moderate to severe Haemophilia and in case of inhibitors (antibodies against recombinant substitutive factors), have often a serious joint involvement. Such condition, called “haemophilic arthropathy”, is the result of a vicious circle that starts in the target joints as a response to the first episode of haemarthrosis, generally during childhood (2). The presence of blood within the joint triggers the synovial tissue and induces a direct damage to the cartilage. The result is a progressive and irreversible arthropathy, which early affects the bone, until the ultimate disabling and painful condition for the daily life activities of generally young subjects (3). Arthropathy mainly involves synovial joints, as elbows, ankles, and knees, and from a clinical point of view it may present differently according to the stage of the disease (4). Haemarthrosis may be spontaneous or subsequent to trauma. The joint is warm, swollen, sore, with functional limitation of the whole limb. Early stages of arthropathy are associated to morning pain diminishing during the day, and worsening in the evening. A high risk of chronic synovitis after recurrent or persistent bleedings is present, inducing a vicious circle of recurrence of bleedings. Finally, advanced stages of arthropathy are characterized by pain (progressively worse), dramatic reduction of range of motion (ROM), malalignment, instability, muscular tone compromission, and severe functional impairment.

At the radiologic study, early abnormalities are not detectable, and patients should be evaluated by ultrasound (children and young subjects) and MRI (young and adult subjects). By standard radiology, it is possible to detect: local osteoporosis, subchondral irregularities and cysts, joint space narrowing, loss of the physiological axis, and osteophytes (5). By US it may be assessed the presence and the activity of synovitis, the type of swelling (blood, synovial fluid), and even the continuity and width of the chondral layer (6). Haematological prophylaxis, lifestyle modifications, physical and medical therapy may be useful for the modern prevention of arthropathy (7). In early stages of arthropathy, physical, medical therapy and conservative treatment by intraarticular injections (viscosupplementation and synoviorthesis with rifampicine) are usually successful (8). When such strategies are no longer effective or severe arthropathy realizes, orthopedic surgery is needed. There are several surgical approaches depending on age and degree of joint involvement. The indications for orthopedic surgery in haemophilic patients are:

• Symptomatic chronic synovitis not controlled by medical or conservative treatment;
• Severe joint contracture with significant functional impairment, particularly in young subjects;
• Bone deformities or loss;
• Severe joint changes associated with pain, disability, and recurrence of bleedings;
• Presence of pseudotumor;
• Failure of an implant.

Any surgery in these patients should be carefully planned by a multidisciplinary evaluation. This is mandatory because such type of surgery has been historically associated to early failures due to the higher risk of bleeding (especially for patients with inhibitors) and infectious risks (particularly in case
Arthroplasty consists in the substitution of an altered joint by the implantation of metallic, ceramic, and plastic components. The aim is to reduce pain and the recurrence of bleedings, restore the function, and ensure the recovery of daily activities. Knee and hip arthroplasty now are fully reproducible and efficient with long-term effects and acceptable survival rates (15-17). Ankle replacement has to be improved, even the good mid-term outcomes and implant survival (17). Total Knee Arthroplasty is one of the most common surgical procedures in haemophilic patients: it allows the correction of the malalignment and the reduction of the number of bleedings by the intra-operative synovectomy. Nowadays, by the use of modern modular knee systems, it is possible to restore bone loss with reconstructive devices (wedges, cones, offsets, stems, different choices of component) associated with the application of bone grafts (15, 18, 19). The hip is less frequently involved by haemophilic arthropathy, due to the paucity of the synovial tissue. During last years, by the use of modular cementless implants, several studies have demonstrated better results with respect to the past in haemophilic subjects (16, 20, 21).

Revision arthroplasty is the procedure in which bone reconstruction has the highest expression. The failure of a previously implanted prosthesis, due to aseptic or septic loosening (both unfortunately very common in patients with Haemophilia) induces the alteration of soft tissues but mainly the loss of bone stock (7). This loss is then amplified by the removal of the failed components (particularly if cemented, the standard until the ‘2000s). The use of properly prepared homologous bone graft and of osteoconductive metallic devices such as wedges, cones, and stems allow to fill the gaps in order to provide a solid base for the implant of a new prosthesis, usually characterized by a higher constraint with respect to the failed one (22, 23). Two-stage revision is generally indicated in case of septic failure, when also scar dehisence and skin alterations are present: before the implantation of a definitive prosthesis, a first surgical step is mandatory in order to reconstruct the soft tissue coverage (local or a free flap). Sometimes the use of a megaprosthesi (generally used in oncologic surgery) is necessary because of the extreme bone loss and mortification typical of the presence of bone pseudotumours (24). Based on our experience as a national reference center for the treatment of Haemophilia and related issues, we can conclude that in case of severe arthropathy associated with persistent symptoms, orthopaedic surgery is a viable option to manage a target joint. Regarding the bone tissues management, surgery may correct and even reconstruct the bone loss, particularly in case of failure of a previous joint implant. This allows a restoration of the bone stock that represent the basis for a revision arthroplasty. Good results may be achieved only in Haemophilia-dedicated centers where skillful multidisciplinary teams are prepared to manage such type of complex patients by tailored approaches.

References
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