

Joint Surgery in the Adult Patient with Hemophilia

E. Carlos Rodríguez-Merchán
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E. Carlos Rodríguez-Merchán
“La Paz” University Hospital-IdiPaz
Madrid
Spain

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Foreword

A reminiscence of an elective orthopaedic repair performed in San Francisco in the early 1960s suggested that in the pre-concentrate era, surgical operations were performed only to try and save a life, and if the patient survived, the postoperative course was stormy and lengthy. In 1968, Dr. J. Vernon Luck, Sr., did succeed in inserting a vitallium cup into a patient's damaged acetabulum under cover of the newly available factor VIII concentrate. Although the patient subsequently did very well with a Charnley 'total' hip replacement, he died of AIDS in 1988 [1].

This early report shows the importance of the collaboration needed between the haematologist and the orthopaedic surgeon in order to provide good haemostasis during joint surgery in haemophilia. Almost half a century later, this book reviews the tremendous advances that have been made in such surgery. The use of continuous infusion and recombinant products has allowed for results similar to those in the general population [2]. These issues are addressed in Chap. 2.

The knee is the most common joint to be affected in haemophilia, followed by the elbow, ankle, hip and shoulder [3]. All these joints are considered in Chaps. 5, 6, 7, 8 and 9 in this book. Total knee replacement provides the most extensive experience and is reviewed in Chap. 8 by the editor of this book, Carlos Rodriguez-Merchán, a surgeon with enormous experience in haemophilic patients.

Although radiography has a place in the management of joint disease in haemophilia, the use of other imaging modalities has revolutionized therapeutic decision making, with magnetic resonance imaging and ultrasonography in particular coming to play an important role. Imaging, reviewed in Chap. 4, is now essential for the good management of joint disease including surgery.

Thrombo-prophylaxis remains a contentious issue for patients with an inherited bleeding disorder, and this is discussed in Chap. 11. There is no doubt that the management of haemophilia requires a multidisciplinary team approach including the physiotherapist and the rehabilitation specialist. These important aspects of care are covered in Chap. 12.

Pain relief for haemophilic individuals with arthropathy can be problematic in the face of the many analgesics exhibiting anti-platelet effects. Inclusion of Chaps. 13 and 14 is therefore very relevant. It is interesting to note that in the 1968 experience, narcotics were given in high doses, and these could be tapered quickly following joint replacement and pain relief [1].

It was Inga Marie Nilsson from Malmö, Sweden, who pioneered prophylaxis in the 1960s, and there is now widespread acceptance of this as a ‘state-of-the-art’ treatment in the well-resourced world following the publication of two random controlled trials [4]. However cost-effectiveness remains an issue, and a discussion of this is included in Chap. 15.

The advent of safe clotting factor concentrates together with the widespread use of prophylaxis raised the prospect of a generation of patients with haemophilia who would not have joint disease. It was even thought that the orthopaedic surgeon would be redundant in this group of patients. Clearly, as this book attests, that is not the case. Patients with haemophilia are living much longer and approaching a normal life expectancy, and this holds true even for those with HIV infection. Many of these patients have severe arthropathy because in their younger life, treatment was unavailable or inadequate. Although modern recombinant products are safe from transfusion-transmitted disease, inhibitors remain a challenge, and these patients may develop arthropathy through ineffective treatment. Those who have had the benefit of modern treatment and prophylaxis demand a normal lifestyle – young boys play football and the ankle will remain a challenging joint. Furthermore, this book addresses the well-resourced world: many parts of the world can only dream of joint surgery.

Much has been achieved in the last 50 years, but the orthopaedic surgeon is likely to be required in the comprehensive care team for many years to come. This book will help those providing the essential joint surgery.

London, UK

Christine A. Lee,
MA, MD, DSc(Med), FRCP, FRCPATH, FRCOG

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Articular Pseudotumors and Bone Cysts in the Adult Hemophilic Patient

10

Manuel Peleteiro-Pensado
and Eduardo J. Ortiz-Cruz

10.1 Introduction

Pseudotumors and bone cysts are a serious, but very rare, complication in hemophilia. The hemophilic pseudotumor is an encapsulated hematoma. They may arise from bleeding into muscle, under periosteum, or into the bone. Once established, the pseudotumor has a tendency to progress and produce clinical symptoms by mass effect in relation to its anatomical location, leading to bone and soft-tissue lesions, or neurovascular complications.

In 1965, Fernández de Valderrama and Matthews [1] described a hemophilic pseudotumor as a progressive cystic swelling involving muscle, produced by recurrent hemorrhage and accompanied by radiographic evidence of bone involvement. In a review of his experience at Oxford, in 1966 Gunning [2] estimated its incidence to be about 1 % of all severe hemophiliacs.

The majority all reported hemophilic pseudotumors involve the musculoskeletal system. A few intra-abdominal pseudotumors have been reported, but these were in fact tumors of the pelvis that had extended into the abdomen [3, 4]. Seldom retroperitoneal pseudotumors have been reported in the literature [5].

The presence of a slowly enlarging mass in the limb or pelvis of a patient with hemophilia should rise suspicion of a possible pseudotumor, despite there have been rare reports of malignant tumors mimicking pseudotumors [6, 7].

Most pseudotumors are seen in adults and occur near the large bones of the proximal skeleton. However, a number develop distal to the wrist and ankle in younger patients before skeletal maturity. If untreated, proximal pseudotumors will destroy soft tissues, erode bone, and produce vascular or neurological lesions. Pathologic fractures can be associated.

10.2 Pathogenesis

Pseudotumors result from repetitive bleeding resulting in an encapsulated mass of clotted blood and necrosed tissue. The pathogenesis of pseudotumors has been much discussed by many authors, most of whom agree that their formation differs according to the anatomical site [8, 9].

Proximal pseudotumors occur in the proximal skeleton, especially around femur and pelvis; they appear to start in the soft tissues, erode bone secondarily from outside, and develop slowly over many years. They occur in adults and do not respond to conservative treatment. Repeated and unresolved hemorrhages, mostly caused by indirect trauma to the muscles, which all have a large area of origin like iliacus, vastus lateralis, and soleus, are the likely mechanism for formation

M. Peleteiro-Pensado (✉) • E.J. Ortiz-Cruz
Department of Orthopaedic Surgery,
“La Paz” University Hospital-IdiPaz,
Paseo de la Castellana 261, Madrid 28046, Spain
e-mail: m_peleteiro@yahoo.com;
ortizcruzej@gmail.com

and development of these lesions. They present as a painless expanding mass which is firm, often multilocular, and not tender, but is adherent to the deep structures. Such pseudotumors frequently remain painless and asymptomatic until there is a pathological fracture. The radiographic picture is typical with a large soft-tissue mass and areas of adjacent bony destruction. Calcification within the mass is common.

Distal pseudotumors predominantly affect younger, skeletally immature patients (children and adolescents) and are generally the result of direct trauma. It is not unusual to see such tumors distal to the wrist and ankle, especially in the small cancellous bones as the calcaneus, talus, and metatarsals of the feet but seldom in the carpus or other locations. These distal lesions develop rapidly, are painful, and appear to be secondary to intraosseous hemorrhage.

10.3 Pathology

A pseudotumor consists on blood products in different evolution stages surrounded by a fibrous capsule that contains macrophages charged with hemosiderin (Fig. 10.1). Calcification and later ossification may be seen within its wall. On histological examination, hemophilic pseudotumors resemble hematomas with a dense fibrous capsule. The cyst wall is formed by collagenous connective tissue, and the cavity contains a variable amount of organized fibrous tissue, thick “toothpaste-like” debris and liquefied clots. Other features that have been observed within the cyst cavity include bone fragments, foci of hemosiderin-loaded histiocytes, vascular neoformation within the cyst wall, osteoid neoformation, giant multinucleated cells, and foreign body-type cells [10, 11].

10.4 Clinical Presentation and Diagnosis

Patients present with painless palpable masses or with painful crises due to episodic acute bleeding into the tumor. Most of the morbidity from

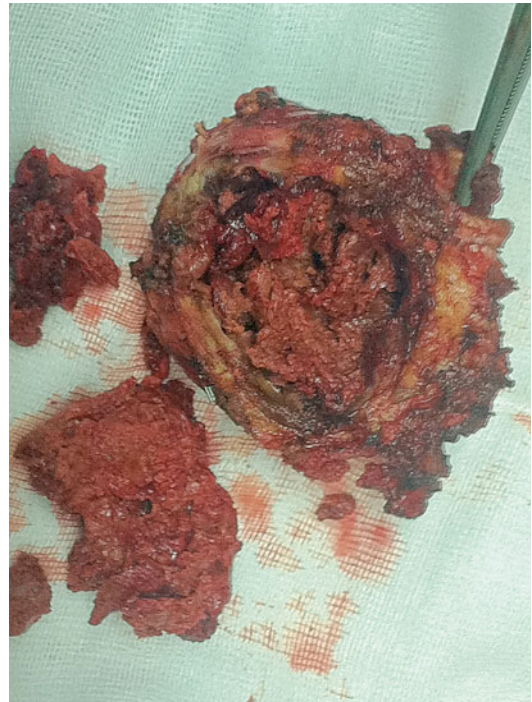


Fig. 10.1 Hemophilic pseudotumor opened after surgical resection

pseudotumors is due to their compressive effect on surrounding structures. Depending on the area involved, the symptoms include palpable masses, numbness, weakness, and neuralgia. Complications occur because progressive enlargement occurs, leading to compression of neighboring vital structures, destruction of soft tissues, and bone erosion, which may produce neurovascular complications. Ultimately there may be perforation through the skin or into adjacent organs, abscess and fistula formation, fatal hemorrhage, pathologic fractures due to bone destruction, and compartment syndromes due to vascular compromise and joint contractures [5].

In X-rays, lesions in proximity to long bones, there is a large soft-tissue mass with areas of adjacent bone destruction. The bone loss may be extensive, involving diaphysis and metaphysis and even destroying adjacent bones by crossing the joints. Periosteal elevation with new bone formation can be seen at the periphery of these lesions. Calcification and ossification within the soft-tissue mass are frequently noted [2, 12]. In

pelvic pseudotumors, the radiographic picture differs from those previously described in that the soft-tissue mass, and the extent of pelvic bone destruction, most of which occurs in the iliac wing, may be difficult to appreciate on plain X-ray films. Periosteal elevation and calcification within these lesions are much less common. An intravenous pyelogram should be performed if there is any concern about displacement of the ureters.

CT scans define the attenuation value of the tumor and thus help to differentiate it from other

masses. It clearly delineates the size and extent of the tumor and compression of the adjacent skeleton, tissues, and organs. CT may prove useful in determining the origin of the pseudotumor and help follow its course. CT scans have been proven to be particularly efficient in the detection of daughter cysts, deep-seated cysts, and tumor extension into adjacent tissues, information that is useful in planning surgery [13].

Ultrasonography is ineffective for the detection of bony changes. It can be used after surgery to monitor for recurrence of pseudotumors in soft

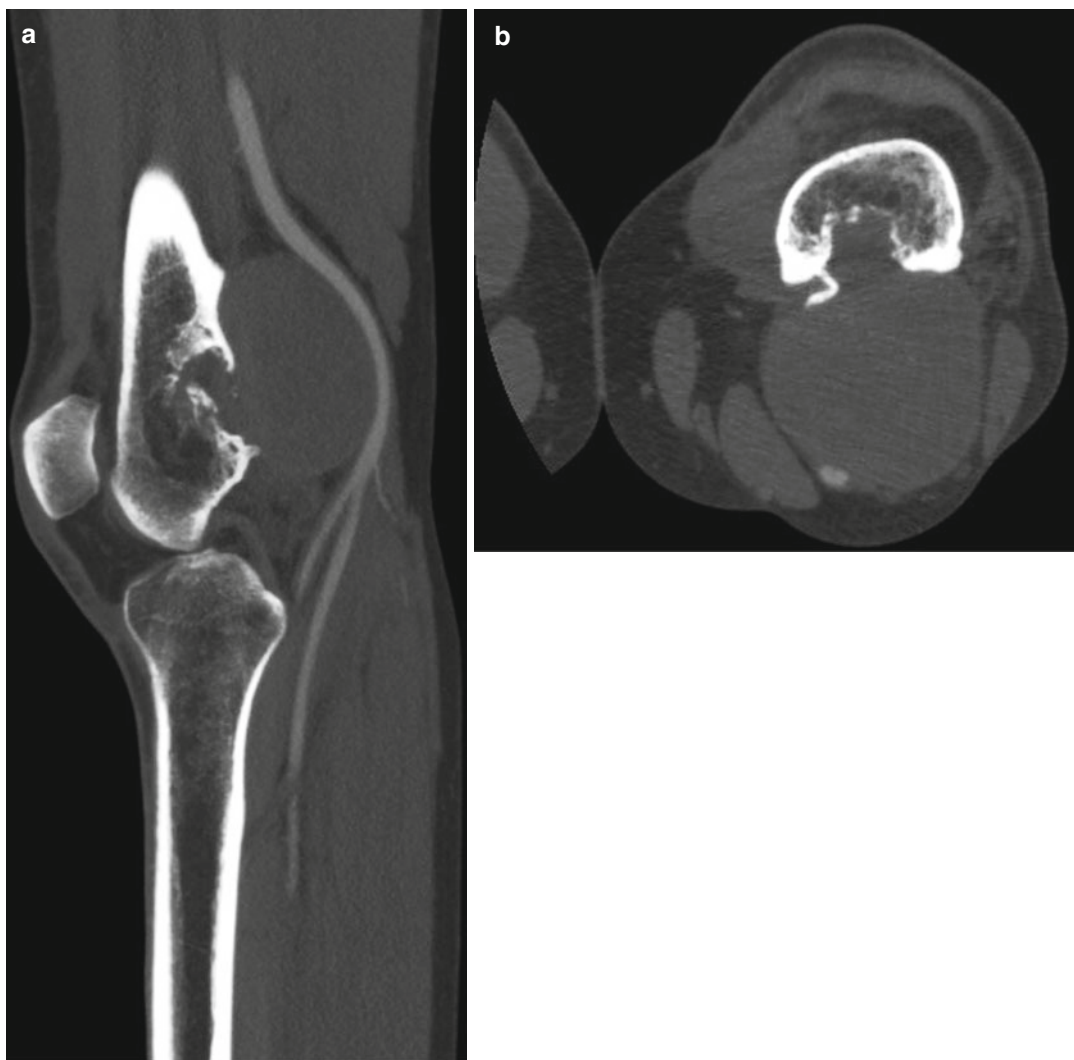


Fig. 10.2 CT of a hemophilic pseudotumor and its relationship with the popliteal artery: (a) sagittal view; (b) axial view

tissues. It has also been suggested as a low-cost alternative to monitor the progress of an existing lesion before obtaining a CT scan prior to repeated surgery [13].

MRI has been shown to be superior to other modalities for the imaging of soft tissues. MRI is sensitive for the detection of pseudotumors and can provide useful information for decisions regarding therapy and may be used to follow the response of a tumor to treatment. The appearance on MRI seems nonspecific with a similar appearance in tumors and abscesses, but invariably there are heterogeneous low and high signal intensity areas on both pulse sequences, reflecting the presence of blood products in various stages of evolution [14, 15].

CT and MRI scans are considerably more useful than plain X-rays and may be sufficient to evaluate displacement of the large vessels (Fig. 10.2). Imaging characteristics of pseudotumors are rather nonspecific, but given a history of hemophilia, radiologists must be aware of these to avoid misinterpretation.

10.5 Treatment

The management of the patient with a hemophilic pseudotumor is complex and carries a high rate of potential complications. It is important that they are diagnosed early, and prevention of muscular hematomas is key to reducing their incidence. Muscle hematomas should be avoided by primary prophylaxis, and if a muscle hematoma does develop, it must be treated hematologically on a long-term basis until total shrinkage and disappearance of the hematoma have occurred [16].

Hemophilic pseudotumor are uncommon; therefore, there is no consensus about specific management. There are different treatment options (surgical removal and exeresis, and filling of the dead cavity, percutaneous management, embolization, radiation), and it would be necessary to individualize therapy. With the increasing availability of factor replacement, surgical removal is feasible.

Preoperative biopsy or aspiration of the fluid within the cysts for either diagnostic or therapeutic

purposes is contraindicated. The cystic contents are too thick to permit successful aspiration, and there is a high risk of relapse, infection, or the development of a persistent fistula [10].

Conservative treatment includes long-term factor replacement and immobilization of the affected area. Distal pseudotumors in children respond well to conservative approach. However, it fails to prevent progression of the lesion especially in proximal lesions, and recurrence and progression is the rule [6].

10.5.1 Surgical Removal

Surgical removal is the treatment of choice when it can be carried out in major hemophilia centers and careful preoperative planning can be done, because these procedures are fraught with difficulty and carry a high complication rate. Surgery is the most effective treatment and should be performed upon diagnosis when the pseudotumor is still small and relatively easily resected. Lesions in proximity to the long bones and lesions of the pelvis should be managed in different ways, and in fact, the surgical approach to these lesions must be individualized.

When the lesion is in the proximity of long bones, the aim should be complete resection of the lesion, stabilization and bone grafting if required, hemostasis, and closure of the dead space. Implantation of a tumoral prosthesis for reconstruction of massive juxta-articular bone lesions can be necessary; although the authors have no experience with this type of reconstruction in hemophilic pseudotumors, it is routinely used for reconstruction after resection of bone sarcomas, and primary arthroplasty in hemophilic patients is also a common procedure in our hospital. The incision has to be planned to allow access to the neurovascular structures, removal of the lesion, and fixation of bone if necessary. Neurovascular structures should be identified and retracted prior to remove the blood cyst (Fig. 10.3). It usually is within a muscle mass and during dissection tries to stay on the fibrous capsule and then as much muscle as possible should be preserved. The portion of muscle wall that is directly adjacent to the



Fig. 10.3 Intraoperative view of the pseudotumor seen in Fig. 10.2

bone can be removed easily with cautery dissection and curettes. Bleeding from the bone or other surfaces can be controlled during surgery by the use of fibrin glue. Bone fixation is an important part of the procedure when necessary; intramedullary nailing has been most often used to stabilize diaphyseal or proximal femur fractures, but when the lesions are around the knee, fixation with periarticular plates can be more useful. Reconstruction of the bone defect after removing the pseudotumor can be done with bone graft, bone substitutes, or bone cement (Fig. 10.4), especially in metaphyseal or epiphyseal defects that involve mechanical risk. At this point, meticulous hemostasis should be obtained. Coverage of the remaining bone should then be addressed, using a muscle flap if necessary. A suction drain is always used and a bulky pressure dressing applied. The use of plaster and the weight-bearing status



Fig. 10.4 Postoperative radiograph. The pseudotumor was removed and the bone defect filled with bone cement

must be calculated for the individual patient, depending upon the lesion itself and the extent of arthropathy in other joints. Nonunion of pathological fractures has not been a problem, because large denuded bone surfaces result in abundant new bone formation.

Pseudotumors of the pelvis develop following hemorrhage into either the iliacus or iliopsoas muscles. It is wiser to perform this procedure in connection with general surgeon. The surgical approach to the pelvic blood cyst is different from that for the extremities. Before starting the procedure, a ureteral catheter is placed so that the ureter can be more easily identified during surgery. The patient is placed on his or her back with the affected side of the pelvis on two sandbags with the lower limb draped free. A flank incision, starting at the proximal aspect of the mass and approximately 3 cm above the iliac crest is made, extending distally to the inguinal ligament and then continuing distally in order to expose the femoral vessels and nerve which should be identified. The flank muscles are then divided and the cysts can be identified. If possible, the dissection is kept within the retroperitoneum. If complete excision of the cyst is impossible, the cyst can be opened and evacuated removing sections of the cyst. Manipulation of vessels, the ureter and the femoral nerve must be very careful to avoid iatrogenic injuries of such structures. Fibrin glue should be used as an adjuvant to control bleeding. One can leave a small cuff of capsule attached to these important structures and do not damage them. Massive pseudotumors in the pelvis can affect the hip joint; these challenging cases can be impossible to successfully treat or reconstruct. Many methods have been employed to eliminate the dead space, including the use of omentum, muscles, dextran mesh, and bone grafting. Pelvic pseudotumors distort natural anatomy causing major displacements of bowels, ureters, nerves, and blood vessels thus increasing the likelihood of injury during surgery. It is our belief that pseudotumors should

be excised in order to prevent them from reaching inoperable dimensions. The management of patients with massive pseudotumors, where the aim of treatment is improvement of the quality of life and not total eradication of the tumour, requires that the patient and their family understand the treatment goals. It is essential that psychological counselling be available before, during, and after the surgical procedure. One of the major problems with surgery in patients with massive pseudotumors is that one knows the starting point but one is never sure of where and when the procedure will end. The decision to operate should not be taken lightly, for the complications include fistula formation, regrowth of the pseudotumor, sepsis, bleeding, and death [17, 18].

There is a significant amount of hemorrhage during and following these procedures. Replacement therapy and transfusion requirements should be carefully considered by the hemophilia team before undertaking surgery.

10.5.2 Radiation

Although surgical excision of hemophilic pseudotumors is currently considered as the preferred management modality, there are instances in the literature that because of certain impediments surgery could not be easily performed and radiotherapy may be beneficial. The exact mechanism of action of radiotherapy is unknown; however, it presumably acts by causing injury to blood vessels and fibrosis. Despite the limited data some conclusions can be pointed out. There was a failure rate of around 25 %, most of which are related to adjunctive treatment modalities implemented, location of pseudotumor, and the size of pseudotumor [19]. Most of treatment failures occurred in patients with proximally occurring masses within pelvis and femur. Distal pseudotumors and the ones which were located in upper limbs or skull generally were more amenable to radiotherapy. This treatment modality may be beneficial for

unresectable lesions. Irradiation prior to surgery is not, in Heim's opinion [17], recommended, as the ensuing fibrosis will make surgery more difficult. Once the pseudotumor has been excised, focal irradiation to any remaining capsule may be beneficial.

10.5.3 Embolization

Pseudotumors are avascular internally but have a very rich blood supply in the capsule. This is most probably the reason for repeated hemorrhages inside the capsule, which result in the expansion of the mass. The rich vascular supply of the capsule is the cause of excessive bleeding during and after the surgery. Regarding the role of arterial embolization in hemophilic pseudotumor management, it should be considered in lesions of large size, especially in pseudotumors of pelvic region, as it may effectively reduce its size and decrease the risk of bleeding complications during surgery. Nevertheless, in view of its temporary effect, embolization had better be performed as a preparatory procedure, at best about 2 weeks prior to surgery. This time lapse will allow for mass shrinkage but is insufficient for vessel restoration [19].

10.5.4 Percutaneous Management

Curettage and filling with fibrin seal and cancellous bone graft via the percutaneous insertion of a trocar into the cyst has been recommended by Caviglia et al. [20]. There may be some value in this treatment modality in small lesions. In advanced or potentially inoperable pseudotumors, there is a role for percutaneous aspiration, but evacuation of the contents of the pseudotumor may be difficult and there is a risk of infection and permanent fistula. In soft-tissue pseudotumors, evacuation and filling of the cavity are carried out assisted by ultrasound, while in

pseudotumors affecting bone, this is performed with an image intensifier or CT scan and video assistance. If the cavity content is liquid, it is easy to aspirate, but solid contents cannot be evacuated by aspiration and will need to be washed out and removed with curettes. Cavities of <3 cm may be filled with fibrin seal; larger cavities should be filled with a lyophilized bone graft or a bone substitute. When the pseudotumor is in the bone, the location, size, and degree of its cortical compromise will be evaluated to confirm the true loss of the bone stock. When the pseudotumor is diaphyseal or diaphy-metaphyseal, it generally consists of a single cavity. In contrast, with a pseudotumor that is epiphyseal or epiphy-metaphyseal (in cancellous bone), the cavities are multiple. Lack of aspiration of one cavity may lead to only partial cure of the lesion. It is very important to clarify the bone damage to prepare bone reconstruction or mechanical support if needed.

Conclusions

A pseudotumor is basically an encapsulated hematoma; it has become rarer over the years with better treatment modalities for bleeding disorders like factor replacement. Its prevention is paramount; this goal can be achieved by primary prophylaxis to avoid muscle hematomas and by adequate and long-term hematological treatment of muscle hematomas in case they appear. There are a number of therapeutic alternatives for this dangerous condition: surgical removal, percutaneous management, exeresis and filling of the dead cavity, irradiation, and embolization. The management of the patient with a hemophilic pseudotumor is complex and with a high rate of potential complications. Surgical excision is the treatment of choice but should only be carried out in major hemophilia centers by a multidisciplinary surgical team. The main postoperative complications are death, infection, fistulization, and pathological fractures (requiring even amputation of the affected limb).

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