Orthopaedic care in haemophilia

Haemophilia, affecting 1 in 10 000 males, is the most common congenital coagulation disorder affecting about 6000 patients in the UK. Although most patients inherit the disorder as an X-linked inheritance, one third arise as spontaneous mutations.

The musculoskeletal hallmark of this condition is spontaneous bleeding into joints and soft tissues. Unchecked recurrent haemarthroses lead to a chronic synovitis and eventually joint destruction. Significant arthritis occurs in at least one joint in the majority of patients with severe haemophilia, i.e. a factor VIII level of <1% of normal. Target joints are predominantly the knee, elbow and ankle, with the shoulder and hip much less involved.

However, since the 1960s, treatment of these patients has become increasingly sophisticated in developed countries. The prevalence of disability has diminished with improved prophylactic care and increased potential for orthopaedic interventions. Initially, cryoprecipitates were used followed by plasma-derived factor concentrates. In the future, it is expected that there will be increased use of recombinant factor replacements, which carry no risk of infection with human pathogens.

ORGANIZATION OF HAEMOPHILIA CARE

Haemophilia patients register with a regional centre for monitoring and treatment. Within such units multidisciplinary teams develop. Treatment advice will be influenced by factors such as patient compliance, available resources and the presence of viral infection. Depth of experience of haemophilia as a disease and of each individual patient is required. Ideally, each haemophilic orthopaedic clinic should involve an orthopaedic surgeon, haematologist and physiotherapist. A haemophilic nurse and counsellor can also provide valuable advice relating to the physical and psychological wellbeing of the patient. Only such a team can provide optimal perioperative management of these patients.

CONTROL OF BLEEDS AND TREATMENT OF SYNOVITIS

Usually the haematological team has responsibility for the treatment of acute musculoskeletal bleeds. Increasingly, treatment commences at home by the prompt use of factor replacement. Rest, ice application and pain relief augment the treatment. The aim is to minimize bleeding and prevent the development of complications such as joint synovitis, articular contractions and, more rarely, nerve compression and pseudotumour formation.

However, despite prompt treatment, target joints can develop, with recurrent bleeds producing a chronically swollen articulation. To reduce the synovitis, patients often embark upon a period of regular 'secondary prophylaxis' of factor replacement. The synovium can be further controlled with radioactive synoviorthesis, chemical synoviorthesis or surgical synovectomy.

SURGICAL CONSIDERATIONS

The development and experience of surgical programmes for haemophilic patients received a severe blow with the discovery of human immunodeficiency virus (HIV) in the late 1980s. Many haemophilic patients were exposed to HIV, hepatitis B and C as a result of the contamination of factor concentrates. Concern for the wellbeing of patients and staff led to a reduction in many of the elective orthopaedic procedures undertaken at this time. However, the improved treatment of HIV-positive haemophilia patients has lead to a substantial revision in life expectancy estimates for these patients and most surgical programmes were re-established by the early 1990s (Phillips et al, 1992).

A review of the effects of major orthopaedic procedures on HIV-positive patients did not reveal a comparatively detrimental effect upon CD4 counts (Phillips et al, 1997). An experienced team must undertake surgical procedures on high-risk patients, using surgical protocols and equipment designed to minimize the risk of penetrating injury and aerosol contamination of staff. The risk of infection in such patients following procedures remains high, however, and such complications must be discussed in preoperative counselling. A multicentre study revealed a deep sepsis rate of 18.7% in primary joint replacements in HIV-positive haemophilics – 10–15 times higher than the normal population (Hicks et al, 2001).

Even in the absence of HIV, the risk of infection following joint replacement in haemophilics is greater than normal with an overall infection rate of 7.2% regardless of HIV status derived from a review of the literature (Hicks et al, 2001). This risk may be related to a number of factors including the need for repeated intravenous medication and the potential for increased bleeding at the surgical site, which can act as a culture medium for micro-organisms.

Procedures on haemophilic patients are technically challenging for the surgeon. Dense fibrosis, significant synovitis and severe osteo-articular destruction renders surgical approaches and definitive surgery more difficult than in the normal patient. Before surgery many patients have experienced long periods of painful, stiff joints. Surgery has been more successful in improving comfort and reducing bleeding episodes, and less successful in restorating movement. Optimal results require the involvement of an experienced physiotherapy team.

The cost of orthopaedic procedures upon haemophilic patients is significantly greater than the normal population. An economic review of total knee replacements calculated that surgery was fourteen times more expensive in haemophilic patients, with 85% of costs accounted for by factor replacement therapy. However, the 90% reduction in

postoperative bleeds into the replaced joint produced marked savings in future from reduced factor replacement requirements. Most haemophilic joint replacements are undertaken in patients with an average age of 35–45 years – 30 years younger than most patients who undergo joint replacement.

A review of perioperative management found haemophilic patients required an initially higher level of analgesics postoperatively. This can be explained by a combination of surgery being more difficult and taking longer and the patient possibly developing a degree of habituation to medication (Ribbans and Giangrande, 2002).

PERIOPERATIVE MANAGEMENT

Safe surgery requires the patient to coagulate normally in the perioperative period. Preparation begins several days before surgery and, in severe haemophilics, concentrates are required until at least 7–10 days postoperatively. Continuous infusions are a more efficient and safer way to administer factors as they avoid trough levels, which may allow breakthrough bleeding.

SPECIFIC JOINTS

The knee is most commonly affected. In the early stages, when synovitis predominates, arthroscopic debridement and synovectomy may be helpful in slowing joint destruction. Open synovectomy occasionally is indicated. Osteotomies have been described but joint stiffness pre- and post-surgery remains a problem. Before joint replacement, arthrodesis was a reliable means of obtaining a comfortable, stable articulation free of bleeding episodes. However, the frequent presence of affected neighbouring joints and patient expectations have rendered the procedure rare in developed countries. Total knee replacement is the procedure of choice for end-stage knee arthropathy. Most major units in the UK have experience of this procedure. The procedure is technically difficult and expensive to undertake. Patients must be advised of the infection risks and should be realistic in their expectations of knee movement. However, in terms of pain relief and reduction in joint bleeds, it has become a reliable procedure.

The ankle often becomes a problem in teenage years. By the fifth decade, 80% of the author's patients reported that ankle arthropathy was having a significant effect upon activities. Most symptoms can be controlled by attention to activities of daily living, physiotherapy to maintain mobility and prevent contractures, especially to the tendo Achilles, and the judicious prescription of boots and orthotics. Like the knee, in the early stages, arthroscopic or open synovectomy may be required. Excision of restricting osteophytes and tendo Achilles lengthening for equinus deformities can improve function. By the time end-stage arthropathy has been reached, the joint is so ankylosed that some patients describe a reduction in pain and bleed patterns. There is minimal experience of total ankle replacement and ankle arthrodesis is the procedure of choice for the painful end-stage haemophilic ankle arthropathy.

The elbow can develop significant deformity and stiffness. Synovectomy, as in other joints, should delay joint destruction. A deformed radial head can be excised to promote forearm rotation. Arthrodesis, in a position of function, has proved successful and there is now increasing experience with elbow replacement in this group.

While the hip is less often affected than the aforementioned three joints, replacement is well described in the haemophilia group. Iliopsoas bleeds can produce significant flexion contractions but the procedure is not as technically challenging as knee replacement. However, infection risks remain and, because the joint is replaced in a relatively young group, increased loosening rates have been reported. Major series indicate a 20–30% revision rate within 10 years.

The final joint to be considered is the shoulder. Surgery is usually reserved for end-stage cases when the choice lies between arthrodesis and replacement.

THE FUTURE

Increasing availability of home treatment with lyophilized coagulation factors, primary prophylaxis programmes, access to physiotherapy, and education facilities have combined to reduce musculoskeletal disability in haemophilia (Nilsson et al, 1992). This should be reflected in reduced requirements for orthopaedic interventions, particularly for end-stage arthropathy. Published data on outcomes of different procedures have helped refine indications and timing of interventions. Cross-fertilization of management protocols via the World Federation of Hemophilia (www.wfh.org) have helped extend optimal management throughout the world. HM

WJ Ribbans

Consultant Orthopaedic Surgeon

Northampton General Hospital

Northampton NN1 5BD

Hicks JL, Ribbans WJ, Buzzard B et al (2001) Infected joint replacements in HIV-positive patients with haemophilia. *J Bone Joint Surg (Br)* **83:** 1050–4

Nilsson IM, Berntorp E, Lofqvist T, Pettersson H (1992) Twenty-five years experience of prophylactic treatment in severe Haemophilia A and B. *J Intern Med* **232:** 25–32

Phillips AN, Eldford J, Sabin C et al (1992) Immunodeficiency and the risk of death in HIV infection. *JAMA* **268:** 2662–6

Phillips AM, Sabin C, Ribbans WJ, Lee CA (1997) Surgery in hemophilic patients with human immunodeficiency virus. *Clin Orthop Rel Res* **343:** 81–7

Ribbans WJ, Giangrande PLF (2002) Hemophilia. In: Bulstrode C, Buckwalter J, Carr A, Marsh L, eds. *Oxford Textbook of Orthopaedic Surgery*. Oxford University Press, Oxford: 1455–70

KEY POINTS

- There is no such thing as minor surgery in haemophilia.
- Orthopaedic surgery should take place in specialized haemophilia units.
- Home treatment and primary prophylaxis have reduced disability from haemophilic arthropathy in childhood.
- Orthopaedic surgery is challenging because of the fibrosis, synovitis and joint destruction.
- Postoperative deep sepsis rates are appreciably higher in haemophilics, especially those with human immunodeficiency virus infection.
- Joint replacement has had a significant effect upon the quality of life for haemophilics in terms of pain relief and reduced bleeds.