HAEMOPHILIC CYSTS

Report of Five Cases

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Spontaneous haemorrhage into joints and muscles is a frequent manifestation of severe haemophilia and as a result the older child or adult may be disabled by joint deformities. Although the management of these problems is often difficult there is no direct threat to life or limb. This is not true, however, of the haemophilic cyst, a complication of bleeding into the musculo-skeletal tissues which may have more disastrous consequences. Since Starker's (1918) case there have been forty-seven reports of this lesion. If untreated, progressive enlargement occurs, leading to compression of neighbouring vital structures, and ultimately there may be ulceration of the overlying skin with infection and fatal haemorrhage. This paper reports five further cases (Table I).

CASE REPORTS

Case 1—A twenty-year-old student whose blood contained no factor VIII was admitted in May 1967 to another hospital with a massive haemorrhage of four days duration into the

<table>
<thead>
<tr>
<th>Case number</th>
<th>Age (years)</th>
<th>Clotting factor defect</th>
<th>Site</th>
<th>Previous history</th>
<th>Duration of swelling</th>
<th>Neural or vascular compression</th>
<th>Treatment</th>
<th>Result</th>
<th>Follow-up (months)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>20</td>
<td>VIII</td>
<td>Calf</td>
<td>Cyst incised under plasma cover only</td>
<td>2 months</td>
<td>Posterior tibial nerve</td>
<td>Immobilisation in plaster for 6 months. Evacuation of haematoma</td>
<td>Healed. Calf scarred and contracted. Recovery of nerve</td>
<td>4</td>
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<tr>
<td>2</td>
<td>31</td>
<td>IX</td>
<td>Thigh</td>
<td>Five-year history of recurrent swelling and infection</td>
<td>1 month</td>
<td>None</td>
<td>Immobilisation in bed 1 month</td>
<td>Resolved</td>
<td>24</td>
</tr>
<tr>
<td>3</td>
<td>19</td>
<td>IX</td>
<td>Iliacus</td>
<td>—</td>
<td>6 weeks</td>
<td>Femoral nerve</td>
<td>Immobilisation in bed 2 months</td>
<td>Resolved</td>
<td>3</td>
</tr>
<tr>
<td>4</td>
<td>9</td>
<td>VIII</td>
<td>1. Tibia</td>
<td>Fall 3 weeks before</td>
<td>1 month</td>
<td>Posterior tibial nerve</td>
<td>Immobilisation in plaster for 2 months</td>
<td>Healed. Recovery of nerve lesion</td>
<td>18</td>
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<td>2. Calcaneus</td>
<td></td>
<td></td>
<td>Injury</td>
<td>3 weeks before</td>
<td>3 weeks</td>
<td>None</td>
<td>Immobilisation in plaster for 6 months</td>
<td>Healed</td>
<td>1</td>
</tr>
<tr>
<td>5</td>
<td>15</td>
<td>VIII</td>
<td>Radius</td>
<td>Pseudotumours of metatarsal and finger</td>
<td>6 weeks</td>
<td>Ischaemia of skin</td>
<td>Amputation</td>
<td>Primary wound healing</td>
<td>12</td>
</tr>
</tbody>
</table>
muscle of the right calf. In the first ten days he received 1,200 millilitres of plasma and five units of blood. The girth of the calf subsided from 43·3 centimetres to 40 centimetres. After five weeks the calf still measured 40 centimetres. Two attempts at aspiration were unsuccessful. Since it was feared that extensive fibrosis would ensue, seven weeks after the onset 800 millilitres of clot were evacuated through three small incisions under cover of one litre of plasma followed by daily plasma infusions. Further bleeding followed and the calf girth increased to 48·8 centimetres.

When the patient was transferred to our care a week later there was oozing from the wounds, the calf measured 48·7 centimetres, and there was a posterior tibial nerve palsy (Fig. 1). The limb was immobilised in plaster, and after six weeks free clot was removed at weekly intervals under plasma cover, with care to prevent fresh bleeding. After twelve weeks it was felt safe to carry out a thorough operative clearance of the calf under human AHG protection. Secondary suture of the wounds was done two weeks later. Slight serosanguinous discharge continued for three months; thereafter the wounds remained healed and plaster protection was discontinued. There was residual equinus deformity of 30 degrees but recovery from the posterior tibial palsy was complete (Fig. 2). It is intended to correct the equinus deformity later, but the calf will remain extensively scarred and it is unlikely that calf function will be restored.

**Case 2**—A thirty-one-year-old man whose blood contained no factor IX was admitted in August 1966 with a large brawny swelling of the left thigh. His history included many bleeds into the knees and hips and he had severe loss of movement in all four joints. In 1961 he had developed a swelling in the left groin which had become infected after aspiration. In 1965 a flare-up of the infection in the left thigh had occurred. A month before his admission the thigh had again become swollen and it was feared that he had developed osteomyelitis of the
femur. On admission the thigh, although swollen to 64 centimetres, was not tender and there was no radiological evidence of bone infection or reaction. Blood was oozing from a posterior sinus but no organisms were grown on culture (Fig. 3). Sinography demonstrated a large cavity in the lateral part of the thigh (Fig. 4).

![Case 2—Haemophilic cyst of the thigh. Blood oozing from the sinus.](image)

**Fig. 3**
Case 2—Haemophilic cyst of the thigh. Blood oozing from the sinus.

![Case 2. Figure 4—Sinogram showing extent of the cyst, but no bony reaction. Figure 5—The same thigh after one month of complete bed rest.](image)

**Fig. 4**
Case 2. Figure 4—Sinogram showing extent of the cyst, but no bony reaction. Figure 5—The same thigh after one month of complete bed rest.

After complete bed rest for a month the sinus had closed and gradual activity was resumed, the patient being fitted with a thigh corset. At the time of discharge the thigh measured 54 centimetres (Fig. 5). Sinography showed closure of the cavity. The patient has remained free from further trouble at this site.

**Case 3**—A nineteen-year-old clerk with Christmas disease was admitted in December 1967 with a history of spontaneous pain in the right groin for twelve hours, and paraesthesia of the
anterior and medial aspects of the thigh. On examination the hip was held in 75 degrees of flexion. A small mass was felt in the right iliac fossa. There was loss of sensation over the anterior and medial parts of the thigh, and the quadriceps was paralysed (Fig. 6). A diagnosis of iliacus haematoma with femoral nerve palsy was made. The limb was supported in the flexed position and treatment with Christmas factor begun.

After two weeks the pain had settled and the hip flexion deformity had been reduced gradually to 45 degrees. Non-weight-bearing exercises were begun before mobilisation in a caliper.

![Fig. 6](image)

*Fig. 6  Case 3—Haemophilic cyst in the right iliac fossa with femoral nerve palsy. The area of sensory loss is mapped out.*

Six weeks after the onset a large mass was noted in the right iliac fossa. The patient had no pain and there was no increase in the neurological lesion. Intravenous pyelography showed displacement of the right ureter but no obstruction and no bony involvement.

Treatment was by complete rest. A month later traction was applied to correct the flexion contracture of the hip and active exercises were begun. After a further two weeks walking in a caliper was encouraged. When the patient was seen as an out-patient a month later the mass was much smaller, the hip flexion deformity measured 15 degrees, the quadriceps was able to sustain the leg against gravity and the area of sensory loss reduced to a small patch. Three months later the mass had resolved completely.

**Case 4**—A boy aged nine years whose blood contained no factor VIII was admitted in July 1966 to another hospital with a swelling of the right tibia. He had fallen on this leg three weeks previously. Knee movements were restricted but there was no haemarthrosis. Despite frequent infusions of fresh frozen plasma and immobilisation in a plaster the mass increased in size and a high fever persisted.

On his transfer to our care three weeks later the swollen limb was hot, tense and tender. It measured 35 centimetres in circumference. There was pyrexia of 38·7 degrees Centigrade. A posterior tibial nerve palsy was present. Radiographs showed a lesion of the upper tibial metaphysis with extensive periosteal elevation (Fig. 7). Plaster immobilisation was continued and for the first few days daily infusions of human AHG were given. During the ensuing two months the leg decreased in girth to 29·4 centimetres and there was radiographic evidence of periosteal new bone formation but no sign of ischaemia of the tibial shaft (Fig. 8).

Walking with crutches was allowed at two months and in November 1966 the boy went home walking in a long leg caliper. By May 1967, almost a year after the onset, knee flexion was 0 to 120 degrees, the posterior tibial palsy had recovered and the upper calf measured 28·1 centimetres; caliper protection was continued (Fig. 9).

At a routine follow-up visit a year after discharge a warm, tense swelling of the right heel and sole was noted (Fig. 10). It had appeared three weeks before, after an injury in which he struck his foot. Radiographs showed another pseudotumour, this time of the calcaneus (Fig. 12). There was no attendant neurological or vascular impairment. The leg was rested in a full length plaster. Once again resolution took two months, by which time the circumference of the swelling had decreased from 30 to 27·8 centimetres. Non-weight-bearing walking with
Case 4. Figure 7—Radiograph of a haemophilic pseudotumour of tibia in boy of 9. Figure 8—Three months later the radiograph shows extensive periosteal new bone. Figure 9—Ten months later remodelling of the tibia is seen.

Case 4. Figure 10—A pseudotumour of the heel in the same boy. Figure 11—The foot three months later showing resolution of the pseudotumour.

Case 4. Figure 12—Radiograph showing the destructive lesion of the calcaneus. Figure 13—Radiograph after three months' immobilisation. There is healing of the cyst.
crutches began in January 1968 and the boy went home two weeks later in a below-knee plaster. After three months radiographs showed healing of the cyst (Fig. 13), and the foot had regained its normal shape (Fig. 11). External protection was stopped.

**Case 5**—A fifteen-year-old boy with severe classic haemophilia and mongolism was admitted to another hospital in June 1967. For three weeks a swelling of the right wrist had been seen, although his mother felt later that slight enlargement might have been present for six months. Radiographs showed a pseudotumour of the lower end of the radius (Fig. 14).

Five years previously a Syme's amputation and later a below-knee amputation of the same leg had been carried out for a similar lesion of the third metatarsal (Jones 1965) (Fig. 16), and a year after that the middle finger of the right hand was amputated for a pseudotumour of the proximal phalanx (Fig. 17).

The radial mass was treated initially by immobilisation in a plaster and 10 units of cryoprecipitate were given daily for twelve days. This seemed to control the bleeding, but two weeks later there was a further increase in size of the swelling which was not controlled by a second course of cryoprecipitate.

On his transfer to Oxford in July 1967 the wrist circumference was 24·4 centimetres. It increased to 27·5 centimetres over the next twelve days and the skin began to show ischaemic changes (Fig. 15). Further deterioration occurred despite daily human AHG administration and fifteen days later amputation was inevitable if ulceration and infection were to be avoided. On the following day the limb was amputated at mid-upper level. The coagulation defect was managed by the administration of human AHG daily through a catheter in the inferior vena cava for fourteen days. The amputation wound healed by first intention but the course after operation was complicated by staphylococcal septicemia which may have been due to colonisation of the catheter. The patient left hospital in October 1967 and has remained well.
**PATHOGENESIS**

**Muscle cysts**—Our first three cases with no direct involvement of bone are examples of simple cysts as described by Fernandez de Valderrama and Matthews (1965). Intramuscular haemorrhage is common in haemophilia and certain anatomical considerations determine the site and extent of such bleeding. The major flexors of the limbs, especially the iliopsoas, calf, and forearm flexors, seem to be prone to involvement. It may be that these groups are more vulnerable to the effects of minor trauma. Massive haemorrhage in these large muscle masses rapidly leads to widespread destruction of muscle fibres, both by the mechanically disruptive effect of haemorrhage and by ischaemic necrosis from compression. The final wasted appearance of the calf in Case 1 bears witness to the severe loss of muscle bulk in the gastrocnemius and soleus that had occurred (Fig. 18). Infarction of muscle from haemorrhage into the closed fascial compartment of the leg, leading to a Volkmann’s type of contracture, sometimes complicates fracture of the tibia; so it should not be surprising to find similar ischaemic changes and contractures in haemophilic haemorrhage, particularly when the haematoma is confined by fascial and periosteal attachments. Indeed Volkmann’s contracture of the calf and forearm muscles is not infrequent in haemophilia.

Despite the prevalence of intramuscular haemorrhage in haemophilia large cysts are unusual. It may be that the size of the haematoma is critical. The commoner, smaller lesions are readily absorbed, but when the haemorrhage extends throughout the muscle compartment,
resorption is slow and incomplete. Once a fibrous capsule has formed around the clot, absorption is further precluded. An adverse influence affecting resolution of haemophilic cysts may be inferred by comparing these lesions with haemarthroses in haemophilia. In the latter the tissue responsible for absorption of the haemorrhage—the synovium—is specially adapted for this purpose, whereas the fibrous capsule encompassing a large haematoma is less vascular, less differentiated, and ill-suited to the absorption process.

Progressive increase in the size of the cyst with the threat of neurological or vascular embarrassment to the whole limb is often the factor that leads to admission to a special haemophilia centre. Three of our five patients had complete nerve palsies; these are not uncommon after serious intramuscular bleeding, particularly a lesion of the femoral nerve accompanying iliacus haematoma. Such neural or vascular compression is reversible and has invariably recovered once the haemorrhage is controlled by the administration of coagulation factors. Further bleeding into a cyst may arise spontaneously due to the inherent lack of clotting factors, and ineffective immobilisation may contribute to this. Even when plasma or more potent plasma fractions are exhibited bleeding may continue (as in Case 5). This may be due to insufficient entry of clotting factor into the fibrous cyst wall. Estimation of the AHG levels in cysts after replacement therapy is needed to confirm this. A more alarming increase in size is sometimes the result of surgical intervention by aspiration, incision or evacuation when such procedures are covered only by plasma. The AHG levels achieved by daily plasma infusions never reach the level necessary to prevent bleeding after operation.

The contents of these muscle cysts have been observed on several occasions. Despite the lack of circulating factor VIII or IX, clotting does take place and large fleshy clots have been evacuated. Organisation has been noted in older cysts. Increase in size of the cyst by accumulation of serum has not been encountered.

Bone cysts—Many authors have subscribed to the view that subperiosteal bleeding is the cause of the haemophilic pseudotumour. Starker’s (1918) patient demonstrated periosteal elevation of the femur, and the second case reported by Reinecke and Wohlwill (1929) was also attributed to bleeding under the periosteum. Our fourth case showed unmistakable evidence of periosteal stripping, subsequent new bone formation and eventual remodelling. Here, undoubtedly, subperiosteal bleeding was the cause of the cyst. Why haemorrhage in this site is so rare in haemophilia is difficult to explain. When one considers spontaneous bleeding into the musculo-skeletal system, it is noteworthy that most episodes occur in sites where movement occurs, that is, in muscles and joints. The more immobile bone and periosteum, despite their rich vasculature, are relatively immune. A further striking feature is that apart from Starker’s (1918) and Echternacht’s (1943) cases, pseudotumours in children have invariably involved the peripheral parts of the skeleton. The adherent adult periosteum is stripped much less readily than that of the child, and in many of the adult cases it seems more likely that destruction of bone has been due to bleeding within the bone or haemorrhage into a muscle with extensive bony attachments.

We feel, therefore, that true subperiosteal bleeding with clear evidence of periosteal stripping and new bone formation is rare. Ischaemia with sequestration of the underlying cortex and the formation of an involucrum as in untreated osteomyelitis did not occur in our case, presumably because the medullary circulation remained intact. There is now ample evidence that the cortex of a long bone will survive loss of either the periosteal or medullary circulation but not both.

Intra-osseous haemorrhage has seldom been implicated in the literature as a cause of pseudotumour, although Favre-Gilly, Chatain, Trillat and Saint-Paul (1965) favoured this explanation in discussing a case of a cyst in the calcaneus. The early massive destruction of the internal trabeculae in our case was, we believe, due to bleeding within the cancellous bone. The eventual reconstitution of the calcaneus with no evidence of periosteal bone formation in our patient and those reported by van Creveld and Kingma (1961) and Favre-Gilly et al. (1965)
favours this explanation. Furthermore, the dense attachment of the periosteum to the calcaneus makes it difficult to envisage bleeding at this site. There are in the literature four recorded cases of pseudotumours involving the calcaneus, all occurring in children. The vulnerability of the heel to trauma is an attractive theory to explain the predilection for this bone over other tarsal or carpal bones, although only in our patient was any history of injury obtained.

A second site of election in these haemorrhagic bone cysts is the ilium, involved in twelve cases. Our third patient may offer an explanation for this. Haemorrhage into the iliacus muscle with femoral nerve involvement is a well recognised complication of haemophilia (Goodfellow, Fearn and Matthews 1967). Although a mass may be felt in the inguinal area it is usually small, as it was initially in this man. The painless development of a large mass in the iliac fossa which ensued eight weeks after the initial bleed suggests to us that the haemorrhage had tracked under the iliacus along the inner side of the ilium. Progressive destruction of the ilium did not occur in our case, but it is significant that the tumours of the ilium reported by Schwarz (1960), Nelson and Mitchell (1962), Caen, Leclerc, Patrux, Bard, Bourdon and Bernard (1964), Eibl, Fischer and Kühlböck (1965) and Revol (1965) had histories of four, two, four, nineteen and eight years respectively.

Other authors have previously suggested that haemorrhage into muscle with wide fibrous attachments to bone may progress to cortical destruction and bony involvement. The numerous cases in which the femur is the site of haemophilic cysts support this view because in this area there are extensive muscle attachments.

**PATHOLOGY OF RADIAL PSEUDOTUMOUR**

In our fifth case the opportunity arose to make a detailed study of the pseudotumour. Longitudinal sections (Fig. 19) of the amputation specimen showed that the lesion was largely extra-osseous. There was very little proximal extension of the haemorrhage through the medulla of the radius, suggesting that the bleed was not initially intra-osseous. Periosteal elevation with new bone formation was evident proximal to the main area of haemorrhagic destruction and a large haematoma under the periosteum of the radius on its medial aspect extended proximally almost to the limit of the section. The pseudotumour, however, seemed to have burst superficially into the subcutaneous tissue and deeply into the radius at the point of maximum tension. Further extension of the haemorrhage into the epiphysial plate and beyond that into the wrist and proximal carpal joints was observed and indicated the potential destructive quality of the process.

The reaction of the tissues to this process was noted on histological section. Externally in the soft tissues the expected pseudo-encapsulation by fibrous tissue was seen. Of more interest, however, was the presence in this fibrous tissue of woven bone—a strong argument in favour of the subperiosteal origin of the lesion. At the edges of the haematoma inflammatory cells were seen, many of them being histiocytes containing red cells and haemosiderin.

We were particularly interested in the reaction of bone to this haemorrhagic process. Where trabeculae were in contact with blood, resorption was taking place and osteoclasts were noted, but away from this line of resorption the bone was viable, and at more distant sites new bone formation was seen (Fig. 20). At the epiphysial plate islands of viable cartilage were seen still undergoing
ossification, despite widespread disruption by the haemorrhage. In the centre of the area of haemorrhage necrotic trabeculae and fragments of cartilage which had been engulfed in the massive bleeding process were seen.

Of interest also was the reaction of the neighbouring joints to the presence of blood. The articular cartilage in places was covered by an adherent fibrinous tissue (Fig. 21) under which the cartilage was thinned and fragmented (Fig. 22). This type of reaction has been noted by Swanton (1959) in her observations on haemophilic dogs and by Rodnan (1959) and Young and Hudacek (1954) in their experimental work. It bears a resemblance to the pannus formation in rheumatoid arthritis.

The pathological findings may be summarised as representing a subperiosteal haemorrhage with reactive new bone formation but progressing externally and internally to cause extensive tissue destruction involving the distal radial metaphysis and epiphysis. There was no evidence of infarction or sequestration except in those bony fragments which were mechanically disrupted. The bony reaction was essentially that of resorption and repair.

An attempt was made by intravascular injection using Micropaque to see if any abnormality could be found in the local vasculature. Numerous thin-walled vessels were seen in the periosteum proximal to the lesion but their significance is not known.

It has been suggested in the foregoing discussion that the cystic lesions of bone in haemophilia are the result of internal and external factors, one or all of which may be operative in any one case and that no single explanation such as subperiosteal haemorrhage can be implicated in every instance. Intra-osseous bleeding into cancellous bone, subperiosteal haemorrhage, bleeding into muscle with extensive bony attachments and pressure necrosis may all result in destruction of bone with the formation of a pseudotumour.

The role of trauma in the production of haemorrhage in the haemophilic is always difficult to apportion. There is no doubt that haemarthroses occur spontaneously and the very frequency of joint bleeds suggests a special mechanism which we do not understand, but in the more severe haemorrhages causing cysts injury seems to play a greater part. In almost half the cases in the literature trauma is implicated and seemed to be the direct cause of the calcaneal cyst in our series.

**TREATMENT**

Despite several case reports and the Mayo Clinic series of Ghormley and Clegg (1948), it was not until the paper by Fraenkel, Taylor and Richards (1959) that accurate diagnosis

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**FIG. 20**

Case 5. Figure 20—Histological section at the edge of the haemorrhage shows osteoblastic activity and new bone formation. (x 260.) Figure 21—Fibrinous exudate is seen overlying articular cartilage. (x 260.) Figure 22—The surface of the articular cartilage shows thinning and fragmentation. (x 260.)
before operation was made regularly in patients with haemophilic cysts. In the thirty cases reported before 1961 biopsy or aspiration was employed in twelve; in ten of these the patient died of haemorrhage and infection. In the present decade the mortality has been less alarming and several successful amputations have been possible as a result of the availability of human AHG (Tables II and III).

Our experience with two pseudotumours and three simple cysts which have been controlled by conservative measures leads us to advocate this approach when the cyst is first diagnosed. We were, of course, fortunate in seeing these cases within a month of onset. Replacement therapy by infusion of plasma or plasma fractions was only required for a short period in the first few days, and we relied on prolonged immobilisation with careful limb measurement and radiological progress films to ensure that resolution was taking place. Repeated evaluation of the neurological and vascular status of the limb served as a further guide to progress. Similar experience of this policy of non-intervention has been recorded by MacMahon and Blackburn (1959), van Creveld and Kingma (1961), Caen et al. (1964) and Favre-Gilly et al. (1965).

Although we believe that the progressive course of haemophilic cysts may be arrested in the early stages by replacement therapy and immobilisation, the patient may present, as has

### TABLE II

<table>
<thead>
<tr>
<th>Site</th>
<th>Age</th>
<th>Duration of cyst before treatment (years)</th>
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<tbody>
<tr>
<td>Femur</td>
<td>20</td>
<td>Over 18.32</td>
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<tr>
<td>Pelvis</td>
<td>12</td>
<td>Under 18.12</td>
</tr>
<tr>
<td>Calcaneus</td>
<td>4</td>
<td></td>
</tr>
<tr>
<td>Tibia</td>
<td>3</td>
<td></td>
</tr>
<tr>
<td>Foot</td>
<td>2</td>
<td>&lt; 1</td>
</tr>
<tr>
<td>Thumb</td>
<td>2</td>
<td>1 - 5</td>
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<tr>
<td>Others</td>
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<td>Total</td>
<td>47</td>
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### TABLE III

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<th>Post 1961 17 cases</th>
<th>Complications and mortality</th>
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<td>Biopsy</td>
<td>8</td>
<td>2</td>
<td>Infection</td>
</tr>
<tr>
<td>Aspiration</td>
<td>6</td>
<td>4</td>
<td>Bleeding</td>
</tr>
<tr>
<td>Evacuation</td>
<td>2</td>
<td>0</td>
<td>Ulceration</td>
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<td>7</td>
<td>2</td>
<td></td>
</tr>
<tr>
<td>Amputation</td>
<td>8</td>
<td>5</td>
<td></td>
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<tr>
<td>Excision</td>
<td>2</td>
<td>4</td>
<td>Before 1961</td>
</tr>
<tr>
<td>Conservative</td>
<td>0</td>
<td>3</td>
<td>After 1961</td>
</tr>
<tr>
<td>No treatment</td>
<td>8</td>
<td>0</td>
<td>Mortality</td>
</tr>
</tbody>
</table>

30 cases 17 cases

- Infection 8 4
- Bleeding 8 3
- Ulceration 2 0
- Before 1961 16 2
- After 1961 2 0
been reported in many other reports, with a cyst too large or too advanced for resolution to be expected. Alternatively the limb may be endangered by progressive neural or vascular compression. In such circumstances, providing sufficient supplies of human or animal AHG are available, excision of the cyst should be undertaken. Hall, Handley and Webster (1962) reported the successful removal of a cyst in the loin and Bailey, Penner and Korte (1965) excised one in the calf. When progression does occur delay is dangerous, and when spontaneous rupture ensues as in Eibl et al.‘s (1965) case the prognosis is very grave. If the lesion is in the extremity amputation may save the patient’s life, but when cysts of the ilium reach this stage they have been invariably fatal. We advocate radical surgery for iliac pseudotumours before the overlying skin becomes necrotic and ulcerated.

Simple evacuation of cysts not involving bone might seem an attractive proposition, preventing increased muscle damage, contracture, and compression of nerve or vessel. We have recently employed this procedure in a boy with Christmas disease who presented with haemorrhage into the knee joint but also extending beyond the joint into the lower third of the thigh deep to and into the quadriceps muscle. Aspiration was unsuccessful four days after onset, so under cover of Christmas concentrate a small incision was made and 900 millilitres of clot were evacuated. Replacement therapy and repeated compression bandaging prevented further bleeding and spared the quadriceps from further damage. Although the suprapatellar pouch became obliterated, knee movement has been restored.

Our first case from another hospital demonstrates the danger of this technique when plasma only is available. Our experience of open evacuation is still limited and we would certainly advise caution in its use. It may be effective only in the very early stages of a large soft-tissue haemorrhage before the haematoma becomes walled off and forms a cyst.

SUMMARY AND CONCLUSIONS

1. Haemophilic cysts are a rare but serious complication of bleeding into the musculo-skeletal system. Five cases are reported.
2. The cysts may arise from bleeding into muscle, under periosteum, or into bone.
3. In early cases conservative treatment by immobilisation and replacement therapy should produce resolution.
4. When alarming increase in size or progressive neurovascular compression occurs, excision of the cyst or amputation should be carried out to prevent the dangerous consequences of rupture, sinus formation or chronic infection.

These patients were treated jointly with Dr Rosemary Biggs, Dr Charles Rizza and Dr James Matthews of the Oxford Haemophilia Centre, and we gratefully acknowledge their invaluable help.

REFERENCES


