DIAGNOSTIC ULTRASOUND IN HAEMOPHILIA


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Ultrasound examination has been recommended for the evaluation of acute haemorrhages into soft tissues or joints in haemophilic patients. We have reviewed the notes of all such patients admitted during one calendar year and find that in 47 separate admissions the ultrasound examination assisted management decisions on 27 occasions. The technique is described with an analysis of the ways in which ultrasound observations may influence the clinician.

There are approximately 4500 individuals with classic haemophilia (Haemophilia A) and 800 with Christmas disease (Haemophilia B) in the United Kingdom. These conditions are clinically identical and are characterised by repeated episodes of bleeding. The severity of the disorder is related to the level of naturally occurring coagulation factor. Those patients with a Factor VIII or Factor IX level of 2% or less are likely to suffer spontaneous haemorrhages. With levels above 10% serious problems are rare and usually related to injury or surgery.

Bleeding may occur into any tissue, the most common sites being intra-articular and intramuscular where they are usually treated by factor replacement, analgesia and immobilisation. The timing, duration and intensity of therapy and subsequent rehabilitation is controlled by the clinician who must base decisions on the degree of pain, clinical examination and laboratory measurement of haemoglobin, coagulation factor and antibodies (Duthie et al. 1972; Rizza 1984).

Although the diagnosis of bleeding is often straightforward there are times when other conditions may masquerade as haemorrhage and vice versa. Those bleeds which occur in the iliopsoas muscle, hip joint and retroperitoneum may mimic a variety of medical and surgical emergencies. Diagnostic ultrasound has been advocated for the detection and follow-up of these haemorrhages (Nowotny et al. 1976; McVerry et al. 1977; Kumari et al. 1979; Wallis et al. 1981; Pettersson and Ahlberg 1982; Belloir, Didier and Well 1983). These patients are often in considerable pain and there is a real risk of precipitating further haemorrhage by injudicious handling. There are clearly advantages in a non-invasive technique which may be performed at the patient's bedside with minimal disturbance.

Since April 1982 it has been our practice to examine with ultrasound all patients admitted to the Nuffield Orthopaedic Centre with haemarthrosis or soft-tissue haemorrhage. This paper summarises our experience, describes the characteristic ultrasound appearances and attempts to assess the practical value of the technique.

METHODS

Technique. Earlier examinations were performed with a Static B scanner (Technicare Unirad EDP 1000) using probes with frequencies between 2.5 and 7.5 MHz. A combination of the two scanning methods has been employed since a real-time machine became available in September 1985 (Diasonics 100; 5.0 and 10.0 MHz mechanical sector).

The first examination after admission is a detailed study to define the location and size of a haemorrhage and to exclude the involvement of adjacent tissue spaces. Real-time probes proved particularly useful in the initial diagnostic examination, but it was often necessary to use the wider field of the B scanner to encompass the full length of limb and retroperitoneal bleeds. Real-time ultrasound allows accurate anatomical definition of individual muscle groups; these can be identified by watching the display while the patient moves appropriate parts of the limb (Fornage et al. 1982).

Previous experience in attempting to measure the precise volume of soft-tissue haemorrhage (Kinnas, Woodham and MacLarnon 1984) showed that it was very time-consuming. We now believe that measurement of length, breadth and depth provides a baseline sufficient to detect clinically important changes in size at subsequent examinations. A record was made of the
Anatomical location, the internal echo pattern of the haemorrhage and the clarity of the borders. To aid the clinician a sketch of the findings was placed in the patient’s notes, indicating the approximate site and dimensions. An example is shown in Figure 1. Whenever possible, follow-up examinations were performed by the radiologist who had performed the initial study and the measurements were taken along the same planes.

Careful examination results in minimal skin pressure and we have not observed any ill effects even in fragile patients with antibodies to Factor VIII or IX. A stand-off jelly block (Kitecho, 3M Corporation) may be useful in reducing the amount of skin contact. This material is also of value when the lesion is relatively superficial and would normally be obscured by artefacts.

Retrospective review. We have attempted to assess the clinical value of ultrasound examination by a review of all the patients admitted to the Nuffield Orthopaedic Centre with a suspected haemarthrosis or soft-tissue bleed in the year April 1984 to May 1985. A clinician and radiologist (PMS and DJW) studied the clinical records at one sitting. When the notes clearly stated that a decision on management had been made as a result of the ultrasound study we noted which ultrasound observation had been most significant in influencing the clinician. If there was no indication that the examination had affected the management of the patient, the ultrasound study was categorised as having had no practical value.

RESULTS

Thirty-eight patients with blood clotting disorders were admitted to the Nuffield Orthopaedic Centre between April 1984 and May 1985 on a total of 51 separate occasions. Ultrasound examination was performed soon after admission and at intervals to assess progress. The frequency of follow-up studies varied, depending on the clinical problem. Four patients were not examined by ultrasound: one was in severe pain and the other three were haemophiliacs with high levels of antibody to Factor VIII. The risk of moving these patients was considered too great; however, later in the series we examined patients with antibodies by bringing the ultrasound equipment to the ward.

There were 31 patients with classic haemophilia, two with Christmas disease and one with von Willebrand’s disease.

Analysis of findings. On five admissions (10.6%) no lesion was identified: this was usually of considerable practical importance. On 22 admissions (46.8%) the ultrasound findings were of no practical value. On 20 admissions (42.6%) there were positive findings which assisted the clinician.

Anatomical location. The site of the haemorrhage usually conformed to the area suspected by the admitting clinician. However, there were 14 occasions when the size and location of the bleed was difficult to detect clinically and the ultrasound assessment of these criteria had a bearing on prognosis or therapy. This was especially true in the measurement of iliopsoas bleeds and in defining whether a limb bleed had extended into adjacent joints.

Echo patterns. We found that the echo pattern of a haematoma varied with its duration. In the soft tissues an early bleed is normally of increased echogenicity compared to the surrounding muscle. The normal tissue interfaces become indistinct and it is often difficult to define precisely the limits of the bleed. Established haematomas were relatively echo-free being less echogenic than the surrounding muscle (Fig. 2). The progressive change to echo-free areas seems to start within three to four days and is usually well established by 10 days. We have observed two broad types of acute haemorrhage: firstly, when the haematoma is of uniform texture and separates muscle planes; secondly, a haemorrhage
that appears to interdigitate between muscle fibres giving a rather mottled texture with very poorly defined borders.

The pattern in haemarthrosis usually showed a mixture of echo-free fluid within the joint and a variable quantity of echogenic material often floating free. Although the very early haemarthrosis was sometimes uniformly echogenic, the pattern was variable and the progress not as predictable as in muscle bleeds. All the acute haemorrhages showed a substantial component of echogenic material within the joint, unlike the echo-free appearance of joint effusions from other causes (Wilson, Green and MacLarnon 1984). On review, there were six occasions when the echo pattern was of value (see Cases 4 and 5).

Follow-up. A serial examination of acute intramuscular haemorrhage showed that both patterns of early bleed may either resolve spontaneously or steadily liquefy, with reduced internal echoes and an increasingly well-defined border. In this latter event, the haematoma may persist for many months, only slowly diminishing in size. Serial girth measurements made at the site of the bleed are potentially misleading as immobilisation results in rapid muscle-wasting. On one occasion the girth measurement diminished when the size of the haematoma increased. A sudden increase in the echogenicity of an established haematoma was taken to indicate fresh bleeding even if there had been no change in its overall size.

There is no difficulty in monitoring an intra-osseous pseudotumour either with plain radiographs or computerised tomography (Pettersson and Ahlberg 1982), but the only satisfactory method of imaging the progression or resolution of a soft-tissue cyst is by ultrasound.

On review, serial examination was of value in eight cases (see Case 1).

CASE REPORTS

Case 1 (aged 36). This haemophiliac with 0% Factor VIII, without antibodies, presented with an established haemophilic pseudotumour of the left thigh. He had sustained a supracondylar fracture of the left femur which had been treated by traction and then a weight-relieving caliper. Fourteen months later a transverse stress fracture had developed proximal to the original fracture. Despite continued use of the caliper there was no sign of bony union seven months later and he

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**Fig. 3**

Plain radiograph of the pseudotumour.

**Fig. 4**

Ultrasound scan of the left thigh (F, femur; H, haematoma).
anterior to the scapula (H).

After two weeks a haematoma formed and discharged through the wound and this became infected with faecal streptococci and Pseudomonas. In the succeeding 11 months he had three episodes of severe sepsicaemia and was transferred to the Nuffield Orthopaedic Centre. Sinograms and ultrasound showed an extensive loculated pseudotumour, and radiographs showed gas in the soft tissues and established non-union at the fracture site (Fig. 3). The sinuses were explored and excised, the pseudotumour removed and the limb immobilised in a compression spica.

Over the following 12 months the progress of the pseudotumour was followed by serial ultrasound examinations through windows made in the plaster spica (Fig. 4). The condition resolved to the point where it was no longer life-threatening and he was discharged home, walking in a specially adapted cast-brace. In this case the use of ultrasound showed that the pseudotumour was gradually but definitely diminishing and the patient was not subjected to further surgery.

Case 2 (aged 19). This haemophilic with 0% Factor VIII, without antibodies, was admitted with a bleed into his dominant right shoulder. He had sustained a dislocation of the shoulder three days before with little trauma. An ultrasound scan showed the haematoma to be within the capsule of the shoulder joint, also extending along the anterior aspect of the scapula. This suggested a recent anterior dislocation with a Bankart lesion (Fig. 5), and we therefore immobilised the shoulder for a period of three weeks, as against our usual practice of mobilising a joint with haemarthrosis as soon as comfort allows. In the succeeding 10 months the patient suffered one bleed into the deltoïd muscle but no further episodes of dislocation.

Case 3 (aged 39). There was 0% Factor VIII without antibodies in this patient who had severe haemophilic arthropathy in numerous joints (Fig. 6). He was admitted with a severe exacerbation of pain in his left ankle. An ultrasound scan confirmed the clinical impression that there was no bleeding within the joint. A confident diagnosis of an acute exacerbation of degenerative arthritis was made and there was no need for a course of Factor VIII, with its attendant risks and cost.

Case 4 (aged 8). This child with 0% Factor VIII and 4% Factor IX, without antibodies, was admitted having had a spontaneous bleed into the non-dominant left forearm 10 days earlier. There was no distal neurological deficit and he was treated with factor cover and immobilisation. The site of the bleed was confirmed by ultrasound examination (Figs 7 and 8). Ten days later he developed pyrexia and there was clinical suspicion of a middle ear infection. There was concern that he might have developed an infection in the haematoma but a further ultrasound examination confirmed that the size of the haematoma had decreased considerably and the pattern of echogenicity remained that of a resolving bleed. The middle ear infection responded to treatment and the ultrasound examination enabled us to manage the patient without a diagnostic aspiration of the haematoma.

Case 5 (aged 8). This boy with 0% Factor VIII, without antibodies, developed a spontaneous bleed in the right groin which did not settle with a dose of Factor VIII given at home. He had recently suffered an upper respiratory tract infection. Clinical examination led to a differential diagnosis of a bleed into the hip or an irritable hip. An ultrasound scan showed echogenic fluid in the hip suggestive of blood or pus rather than a simple effusion (Fig. 9). As there were no clinical signs of infection it was decided to defer joint aspiration and treat him as for an acute haemarthrosis. Two days after admission a mild pyrexia caused concern that there might be metastatic infection of the joint.
Follow-up studies should be performed at intervals when the clinical circumstances dictate, but once a week is usually sufficient. Repeat studies are much less time-consuming and generally take around 5 to 10 minutes. The initial diagnostic examination normally lasts approximately 30 minutes.

Not least of the benefits of early ultrasound examination is that the patient himself commonly expresses relief at "seeing" the haemorrhage. We suspect that there are occasions on which even the best-informed haemophiliac worries that he is suspected of "crying wolf".

One patient developed a mild allergic reaction to the ultrasound jelly; we have not observed any other complications of the procedure. We suggest that ultrasound may be undertaken safely in almost all haemophiliacs. The patient does not need to leave his bed and the machine may be brought to the ward in the more severe or fragile cases.

At the Nuffield Orthopaedic Centre diagnostic ultrasound is now considered a routine part of the management of soft-tissue haematomas in the haemophiliac.

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REFERENCES


