HAEMOPHILIC ARTHROPATHY OF THE SHOULDER

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Among 41 adult haemophiliacs 15 suffered from shoulder symptoms. We examined 12 patients by radiography and ultrasound. Four had bilateral symptoms making a total of 16 symptomatic shoulders. Of these, 10 had abnormal ultrasound scans with eight having evidence of rotator cuff tears. Evidence of bicipital tenonitis was found in two. Pain with loss of range of movement and a positive impingement sign was the most reliable clinical indicator of a cuff tear. Joint incongruity and superior migration of the humeral head were the best radiographic indicators. Rotator cuff tears are a common component of haemophilic arthropathy of the shoulder.

Little has been written about the effects of haemophilia on the joints of the upper limb, particularly the shoulder. The pathogenesis of haemophilic arthropathy in the shoulder has been presumed to be the same as for other joints, repeated spontaneous haemorrhages resulting in widespread destruction. Incomplete adsorption of intra-articular blood leads to retained blood products which produce chronic inflammation of the synovium; a permanently swollen joint with contractures and muscle atrophy results. In the terminal stages the larger joints may go on to fibrous or bony ankylosis, while smaller joints may be completely destroyed (Epps 1983).

The humeral head and adjacent glenoid may develop joint space narrowing and irregularity, interosseous cysts and subluxation (Ahovuo, Paavolainen and Slatis 1986). Post (1988) has cautioned against manipulating such a subluxed humeral head. This condition results from a large volume of blood in the joint 'the piston effect' of Epps (1983). Heim, Horoszowski and Martinowitz (1986) have described an adolescent with a locked shoulder secondary to severe incongruity.

It is not clear from the literature what is the incidence of symptomatic and asymptomatic disease of the shoulder in patients with moderate or severe haemophilia; what are the effects of haemophilia on the soft tissues about the shoulder, including the rotator cuff, the biceps tendon and the synovium; whether there is a relation between inferior subluxation or pistoning and rotator cuff patho-
logy; and whether impingement syndrome is responsible for the symptoms in the haemophilic shoulder.

Many authors (Crass et al. 1984; Middleton et al. 1985; Ahovuo et al. 1986; Middleton et al. 1986) have used ultrasound in the diagnosis of rotator cuff tears, bicipital tendonitis and impingement syndrome. We have used ultrasound as well as radiographic methods to detect those lesions in the haemophilic shoulder.

MATERIALS AND METHODS
A population of 41 mild, moderate and severe adult haemophiliacs was screened at the Haemophilia Clinic in Winnipeg. Patients with shoulder symptoms were given a questionnaire and underwent physical and radiographic examination. We used a scoring system modified from Pettersson, Ahlberg and Nilsson (1980) as recommended by the World Federation of Haemophilia. Subsequently the shoulders were examined by ultrasound. Physical examination included range of motion measurements, recording of muscle atrophy and weakness, and special tests to diagnose rotator cuff or biceps tendon disease. One author (PMcD) performed all the physical examinations and scored all radiographs. Ultrasound evaluations were done by two examiners both specially trained in shoulder arthrosynthesis. Both were present at every examination and agreed on all findings.

RESULTS
The severity of disease, patients' ages and the incidence of symptoms in the shoulders and other joints are given in Table I. This classification of severity was based on factor activity (≤ 1%, severe; 1% to 5%, moderate; > 5%, mild).

Ultrasound. The ultrasound findings in 16 symptomatic shoulders are given in Table II.

Scoring. Joints were scored according to their signs and symptoms (maximum 16 points) and according to their radiographs (maximum 14 points). The higher the score the worse the shoulder. The average signs and symptoms score in the symptomatic shoulders was 6.00 and the average radiological score was 4.50. In cases where the ultrasound was positive the average scores were 8.20 and 8.10 respectively.

Range of motion. Most symptomatic shoulders had some loss of range of motion. This loss was generally > 30% when rotator cuff lesions were present. The loss of motion also correlated with irregularity and incongruity of the glenohumeral joint on the radiograph. When rotator cuff lesions were present, loss of motion was greatest in internal and external rotation.

DISCUSSION
The shoulder joint is often neglected in discussions of haemophilic arthropathy and the incidence of its involvement is cited by many as low. By contrast, we found a 37% incidence of shoulder problems. Högh, Ludlam and Macnicol (1987) considered that the elbow was the principal site of haemophilic arthropathy in the upper limb, but in our series the elbow was involved in only 23% of patients. This is despite the fact that all but one of our patients with shoulder symptoms gave a history of haemarthrosis in the elbow.

Conclusions. The joint most commonly involved in the upper limb in severe haemophilia is the shoulder; rotator cuff tears and impingement symptoms are an important manifestation of this arthropathy.

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REFERENCES