Refer to: Banta JV, Boone DC, Smith CF: Arthrocentesis of the knee in acute hemophilic arthropathy. West J Med 122: 285-288, Apr 1975

Arthrocentesis of the Knee in Acute Hemophilic Arthropathy

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In 27 children and young adults with hemophilia presenting acutely painful distended intra-articular hemorrhages of the knee, aspiration was carried out and the patients were followed for a minimum of 24 months. Seventeen patients with classical hemophilia were found to have less than 1 percent of normal plasmal level of antihemophilic factor (AHF). Of the remainder, five were Factor IX, plasma thromboplastin component (PTG), deficient, whereas two patients had Von Willebrand's disease. Aspiration was routinely done in an outpatient clinic, followed by immediate discharge with return to regular activity levels within 48 hours. There were no infections nor rehemorrhages attributable to aspiration technique.

MANAGEMENT OF CHRONIC DISABILITY caused by joint hemorrhage in patients with hemophilia has been a concern for many years. Recurrent blood loss, especially into the knee joint where incidence is highest,[1-5] creates a vicious cycle of inactivity and progressive disability.

Many clinicians treating patients with hemophilia mention aspiration of the acutely distended joint[2,3,6,7] as a desirable adjunct to therapy but little mention is made of technique, hematologic control or long-term effectiveness. Petersen[8] reported one instance of death from streptococcal infection following knee joint aspiration and it is presumed that many physicians have abandoned aspiration for similar though less catastrophic reasons.

Trueta[9] described the vicious cycle of joint destruction. Intra-articular blood under pressure produced painful joint distention and muscle imbalance about the knee with compression of the opposing hyaline cartilage surfaces. Further damage to the articular cartilage was caused by subchondral and synovial ischemia due to decreased blood flow. De Palma[2] classified four stages of a progressively destructive process; irreversible changes were found by Stage Three.

Although the often quoted figure of 20,000 to 40,000 persons with hemophilia in the United States is unsubstantiated, the general incidence is estimated at one per ten thousand males.[10] Of this group, 80 percent have Factor VIII—antihemophilic factor (AHF)—deficiency (hemophilia A, classical hemophilia) and 15 percent have Factor IX—plasma thromboplastin component (PTC)—deficiency (Christmas disease, hemophilia B). In both types, there is recurrent hemarthrosis. Lacking a documented study of aspiration of the acutely distended joint and facing a large hemophilic population, this feasibility study was under-

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Submitted, revised, September 27, 1974.

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taken to attempt to decrease the morbidity and the long convalescent period inherent in this joint disorder.

Material

Subjects were selected from the population of 400 patients with hemophilia cared for at the Regional Hemophiliac Rehabilitation Center at the Orthopaedic Hospital in Los Angeles. Patients were candidates for this study when they were seen for administration of plasma products for acute hemarthroses of the knee and when distention, limitation of motion and acute pain or discomfort were noted in the joint. The suprapatellar pouch was uniformly distended in the less chronically affected joints. Distention was less evident in those joints with advanced degenerative changes of soft tissue scarring and contracture; however, pain appeared more severe in the more severely degenerated joints. Plasma levels of Factor VIII as determined by the two-stage technique were known and the severity of the disease was classified according to Nilsson's rating.[11] (See Table 1.)

Variables studied were duration of hemorrhage, x-ray rating of the aspirated joint, knee circumference, range of motion, volume, cell count and hematocrit of the aspirated material and the walking-activity level. Measurements were taken of the circumference of the knee at both the midpatella and one and two inches above and below this site, plus joint ranges of motion immediately before and at regular intervals after aspiration.

Following baseline measurements, an infusion of intravenous glucose and water was started. Under sterile conditions with 1 percent lidocaine hydrochloride (Xylocaine® hydrochloride) anesthesia, arthrocentesis was carried out with a 15 gauge needle from the most accessible site adjacent either to the medial or lateral superior poles of the patella. Following aspiration, 1 ml each of dexamethasone sodium phosphate (Decadron®) and prednisolone tertiary-butylacetate (Hydeltra®-TBA) diluted to 6 ml with 1 percent hydrochloride were instilled into the joint in an effort to reduce the local inflammatory response.[12] Plasma products were then administered, with AHF deficient patients receiving sufficient concentrate or cryoprecipitate to raise their AHF levels to 50 percent. PTC deficient patients received the maximum amount of lyophilized plasma, calculated by body weight, that could be administered without overloading the cardiovascular system. This raised their PTC levels to between 15 and 20 percent. A sterile compress and an elastic wrap were applied to the knee, followed by a posterior molded thermoplastic splint for 24 hours.

Patients were then seen following aspiration (defined as day 0) on days 4, 7, 14 and 28, and at six and eight weeks. Plasma products were administered to attain factor levels of 20 percent on days four and seven, walking with crutches was begun on day one and crutches were discontinued by day four. The molded thermoplastic splint with an open anterior thigh was used for 24 hours and supervised exercises under a therapist's direction were begun day four.

TABLE 1.—*Description of Subjects*

1. *Type of Hemophilia*			*Patients*
Antihemophilic factor (AHF) deficient			20
Plasma thromboplastin component (PTC) deficient			5
Von Willebrand's disease			2
TOTAL			27
2. *Severity of Hemophilia*			
Plasma levels of AHF or PTC factors (2 stage technique)	*Hemo A (AHF Deficient)*	*Hemo B (PTC Deficient)*	*Von Will.*
5 to 25% of normal (mild)	1	0	0
1 to 5% of normal (moderate)	2	0	0
Less than 1% of normal (severe)	17	5	2
Untested	0	0	0
TOTALS	20	5	2
3. *Age*			
Mean	13.4 years		
Median	11.5 years		
Range	6-35 years		
4. *Duration of Hemorrhage*			
Mean	2.2 days		
Median	1 day		
Range	3 hours to 14 days		
5. *Cause*			
Spontaneous			25
Traumatic			2

Results

During the first year of this investigation, aspirations were carried out on 27 patients, equally on the right and left knees. Characteristics of the joint aspirate are listed in Table 2.

A mild elevation in cell count would indicate that hemophilic arthropathy should be included in the Grade 1 classification of Ropes and Bauer,[13] along with traumatic effusions and osteoarthritis.

Current anteroposterior and lateral x-ray views

were evaluated for the severity of degenerative changes. De Palma's[2] classification ranging from earliest detectable changes (Grade 1) to far advanced fixed deformities (Grade 4) were used. The median grade of degenerative changes in 27 patients was 2 with a range of 0 to 4.

Loss in knee extension varied between 5 degrees and 60 degrees at the time of aspiration with improvement to between 0 and 45 degrees by day four. Total range of motion ranged from 0 to 110 degrees at the time of aspiration and improved to between 40 and 140 degrees on day four. A mean gain of 21 degrees of knee extension was noted and a mean overall increase of range of motion of 65 degrees was evident. By eight weeks the total range of motion averaged 97 degrees and loss of knee extension measured only 8 degrees. Circumference measurements decreased by an average of 2.2 cm by the fourth day after aspiration, with little change thereafter (see Table 3).

Within the eight week period arbitrarily designated as recovery from an acute hemarthrosis, there was spontaneous recurrence of blood loss in nine knees; in two knees during the fourth week, five during the sixth week and two during the eighth week. The mean age of the rehemorrhage group was 12.7 years, with a mean x-ray rating grade of 2. One patient was lost to follow-up due to hepatitis developing in the second week after aspiration.

Most significant was the return of all aspiration candidates to normal school and employment activities within 48 hours of aspiration. In no instance could rehemorrhage be attributed to the operation technique. There were no instances of infection.

Attempting numerous correlations of the first year's findings showed an inverse relationship between x-ray rating and volume of aspirate. As noted earlier a knee joint with advanced degenerative changes has a reduced joint volume. Similarly, a decreased cell count was noted with increasing aspirate volume; this may represent a dilutional effect.

During the second year of this study 24 of the original 27 patients were followed for a total of 24 months. Ten remained free of blood loss in joints, whereas 14 required repeat aspiration. At the end of 24 months, 12 patients were walking without artificial support, whereas, 12 required the assistance of a posterior splint or crutches, or a brace.

A review of hospital records was made to ascertain the frequency of blood losses into the affected joints for the immediate two years before beginning aspiration treatment. The chi-square of analysis was of only marginal significance in analyzing the frequency of blood losses in the aspiration versus preaspiration groups.

TABLE 2.—*Aspirate Analysis*

1. *Volume (N=27)*	
Mean	73.2 ml
Median	70.0 ml
Range	15 to 200 ml
2. *Cell count (N=23)*	
Mean	3,460 cells per cu mm
Median	2,750 cells per cu mm
Range	600-9,000 cells per cu mm
3. *Hematocrit (N=26)*	
Mean	31.9%
Median	33.5%
Range	6.0%

TABLE 3.—*Range of Knee Motion*

	Aspiration Day (degrees)	*Day 4 (degrees)*	*Weeks 6 to 8* (degrees)*
Loss of terminal knee extension (mean)	35.4	14.6*	8.7
Total motion (mean)	28.9	94.4	106

*26 patients only.

TABLE 4.—*Comparative Knee Circumference Measurements*

	Aspiration Day (centimeters)	*Day 4 (centimeters)*	*Weeks 6 to 8* (centimeters)*
Maximum circumference (mean)	32.6	30.4	30.1

*N=26 subjects.

Discussion

Aspiration of the acutely distended, painful, joint in hemophilic arthropathy is a safe and effective procedure when undertaken with rigid aseptic technique and adequate replacement therapy. Before instituting aspiration techniques in a patient with a serious intra-articular bleeding episode, an average of three to seven days in the hospital would be required with cast or traction immobilization followed by an often prolonged convalescence. The 27 patients making up this series represent a consecutive prospective series selected only by virtue of the relative severity of their intra-articular hemorrhages. All were treated as outpatients and discharged to their homes within three hours. All patients followed for the 24

months duration of the study were able to resume their usual and customary activity level within 48 hours of aspiration. There was no instance of infection or rehemorrhage attributable to this procedure. Strict aseptic technique, replacement therapy to the 50 percent concentration level and appropriate temporary splintage were essential to this rapid rehabilitation.

The exact nature of the toxic agent in recurrent hemarthrosis is unknown. Hemosiderin deposition throughout the synovium and in macrophages beneath the synovial layer, with fibrosis and scarring of the synovium and capsule, precedes articular destruction.[14-17] Recent investigations[18] of the role of lysosome in articular cartilage degeneration have confirmed the original studies of Fell and Dingle,[19] who first delineated the lysosomal acid protease in which the lysosome was shown to be capable of breaking down the polysaccharide of cartilage matrix in vitro. Roy[20] recently described the earliest degenerative changes in the cartilage matrix by using electron microscopy. Many iron-containing bodies, called siderome, were seen in the chondrocytes of the superficial and middle zones of the articular cartilage of hemarthritic joints. These bodies were seen before any cartilage changes were visible by light microscopy techniques. Roy postulated that lysosomal liberation from these sideromes may initiate the process of cartilage destruction by causing chondrocyte damage.

The mean cell count of 3,460 cells per cu ml, calculated on a sample of 23 aspirants, is only slightly above the normal range for nonspecific traumatic arthritis. This low concentration would suggest a rather small reservoir for lysosomal enzyme liberation.

It would appear from review of current literature that prevention of the initial intra-articular deposition of hemoglobin is a key factor in preventing the often insidious progression of hemophilic arthritis. Results of preliminary review of prospective prophylactic studies now under way at Orthopaedic Hospital would appear most promising. However, in the treatment of established severe hemarthrosis, the role of aspiration should be considered a safe and effective means of restoring early joint function. The incidence of intra-articular hemorrhage in its relationship to seasonal variations, trauma, type of hemophilia, severity and activity is difficult to document. Although it was the clinical impression that aspiration of the knee decreased the number of recurrent hemarthroses, statistical analysis on this admittedly small number of patients did not verify our clinical impression.

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