On examination the only positive but unequivocal finding was tremor and incoordination of the arm on performance of the finger-nose test. It appears probable that she had Huntington's chorea in its early stage.

The following are brief notes on the deceased members who were affected.

I 1.—Onset of illness occurred in middle age and death at 65. He suffered from old jerky movements of the limbs and progressively became "very stupid."

II 1.—Onset of illness occurred at about 40 and she died at 50. She had involuntary jerky twisting movements affecting the hands, face, head, limbs, and trunk. She "flung" out her limbs at times, both at rest and when walking. Her mental functions also declined and she became slow in speech and poor in memory. During the last year of illness she was weak and bedridden and had to be spoon-fed.

III 4.—Onset of illness occurred at 29 and he died at 37. The sequence of events was clumsiness in grasp; spontaneous loose abrupt movements affecting all limbs; and progressive generalized weakness. Mental faculties, however, were preserved to the end.

IV 1.—Onset was at 22 and death occurred at 28. Here also the illness started with clumsiness followed by involuntary movements similar to those in the preceding cases and affecting most parts of the body. Articulation became severely defective. He was demented for the last two years.

Comment

In this family there were seven cases (including one probable case) of Huntington's chorea. Four were males and three females. The age at onset ranged from 10 years to middle age. The two cases described in detail had chorea and dementia in degrees varying from moderate to severe. Chorea and dementia were also present in all the other affected members of the family, with the exception of one (III 4) in whom intellectual functions were preserved to the end.

The heredo-familial character of the disease and its Mendelian dominant inheritance are evident. The four choreic parents in the family had 17 siblings between them. Six were affected—that is, about one-third. Of the rest, eight were well (including two below the age of 10) and three died in childhood.

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Sir William Fletcher Shaw, of Manchester, Emeritus Professor of Obstetrics and Gynaecology at Manchester University, President of the Royal College of Obstetricians and Gynaecologists, 1938, left £46,569 gross, £46,058 net (duty paid, £13,742). He left £1,000 and the George II silver salver presented to him by the past and present members of the council of the college "and also the gold medal awarded to me for my M.D. thesis" to the Royal College of Obstetricians and Gynaecologists, and £300 to the Rector and Churchwardens of Swettenham Church, Cheshire, upon trust to apply the income towards the maintenance and repair of the communion table, the choir stalls, memorial tablet, oak panelling, and pavement. (*The Times*, April 13.)

ACUTE TRANSIENT ARTHRITIS OF THE ADULT HIP

ITS RELATION TO RHEUMATOID ARTHRITIS AND TO "PALINDROMIC RHEUMATISM"

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During a period of four years we have seen eight adult patients with an acute arthritis of the hip, accompanied by intense and disabling pain, which resolved, leaving the joint more or less normal. Aspiration was performed on seven of them; cloudy yellow fluid under pressure was obtained in five and proved sterile in each. This paper presents the clinical and laboratory findings, follow-up observations, and probable diagnosis in these eight patients.

Case 1

A woman aged 68 had for 12 years been subject to occasional attacks of pain and swelling in the hands, wrists, knees, and ankles. She presented with severe pain of sudden onset in the right hip, accompanied by vomiting. The right hip-joint was tender and its movement restricted and painful. Temperature 100° F. (37.8° C.); white blood-cell count (W.B.C.), 4,000/c.mm. (differential count normal); erythrocyte sedimentation rate (E.S.R.), 54 mm./1 hour (Westergren). The Rose-Waaler test (Rose *et al.*, 1948) was positive at 1/256.

Aspiration of the right hip-joint yielded 10 ml. of cloudy yellow fluid. Penicillin 250,000 units was introduced into the joint and systemic penicillin given. The aspirated fluid showed numerous polymorphonuclear leucocytes and no organisms. Culture was sterile. Severe pain subsided in three days and full function returned in four weeks.

Four and a half years later there had been no further pain in the right hip, which had full painless movement. The hands showed rheumatoid changes. E.S.R., 22 mm./1 hour. Latex test (Singer and Plotz, 1956) was positive at 1/160.

Case 2

A man aged 43 presented with severe pain of recent onset in the right hip. The joint was immobilized by muscle spasm. By next morning the left hip and both ankles were affected. Temperature 99° F. (37.2° C.); E.S.R., 43 rising to 82 mm./1 hour; W.B.C., 14,600/c.mm. (neutrophils 79%, eosinophils 1%, monocytes 8%, lymphocytes 12%). Aspiration was not performed. Treatment consisted of bed rest, sodium salicylate for two weeks, and chloroquine 400 mg. daily for five weeks. Severe pain subsided after two days, and at two weeks all pain had gone.

Twenty-seven months after onset there was occasional slight ache in the right buttock and thigh. Both hips had full painless movement. E.S.R., 1 mm./1 hour. Latex test was negative throughout.

Case 3

A man aged 52 presented with right hip pain. Two weeks previously severe pain and "fixation" of the right hip had necessitated admission to another hospital. There had been no urethritis but there had been recurrent conjunctivitis complicated by superficial punctate keratitis. The right hip-joint was slightly tender. Flexion was from 10 degrees to full, rotation was very restricted and abduction slightly

restricted—all with pain at the limit. Temperature 98.4° F. (36.9° C.); E.S.R., 45 mm./1 hour. Rose-Waaler test was negative. Attempted aspiration of the right hip-joint yielded no fluid. Treatment was by bed rest, traction, and short-wave diathermy, and the patient walked after four weeks.

During the next 18 months there was slight ache in the right groin, with two exacerbations, each lasting one week. Later the right shoulder and the toe-joints were affected. Two years after the original attack all movement of the right hip was slightly restricted, with pain at the limit. E.S.R., 2 mm./1 hour. Latex test was negative.

Case 4

A man aged 53 had suffered eight attacks of acute pain in the right shoulder during a period of four years. He presented with acute pain in the right hip. The joint was tender and all movement was limited by pain. Temperature 99° F. (37.2° C.); E.S.R., 32 mm./1 hour; W.B.C., 7,800/c.mm. Rose-Waaler test was positive at 1/256.

Aspiration of the right hip-joint yielded 5 ml. of cloudy yellow fluid, and hydrocortisone 37.5 mg. was injected into the joint. The aspirated fluid contained numerous polymorphonuclear leucocytes and was sterile on culture. Severe pain subsided in one day and the patient walked without pain after a week.

During the next 18 months there were three further episodes of acute pain in the right hip, each lasting one day. Cloudy yellow fluid was aspirated on two occasions. Two years after first presenting the hips had full painless movement. E.S.R., 3 mm./1 hour. Latex test was positive at 1/80.

Case 5

A woman aged 46 presented with severe pain in the left hip. Her face was drawn and she was apprehensive of movement. The left hip-joint was tender and its movement restricted. Temperature 99° F. (37.2° C.). E.S.R., 22 mm./1 hour; W.B.C., 10,800/c.mm. (neutrophils 82%, monocytes 2%, lymphocytes 16%). Rose-Waaler test was negative, but became positive at 1/512 on the seventeenth day. Aspiration of the left hip-joint yielded 4 ml. of cloudy yellow fluid showing numerous polymorphonuclear leucocytes. Penicillin was given until culture of the aspirated fluid was reported sterile, when prednisolone 30 mg. daily was given for three days and 15 mg. daily for a further three days. Severe pain subsided after one day and the patient left hospital walking well on the tenth day.

Sixteen months after the first attack the left hip was again acutely affected. E.S.R., 55 mm./1 hour. Latex test was negative. Aspiration of the left hip yielded 6 ml. of cloudy yellow fluid containing numerous polymorphonuclear leucocytes. Culture was sterile. Acute pain subsided in five days, and two weeks after the onset the patient was free of pain. One month after this attack the E.S.R. was 19 mm./1 hour and the latex test still negative.

Case 6

A woman aged 34 presented with acute pain in the right hip, which had begun four days before. She was greatly distressed. The right hip was held flexed and abducted and movement was very restricted by pain. The front of the hip-joint was swollen and tender. Temperature 99° F. (37.2° C.); E.S.R., 11 rising to 24 mm./1 hour; W.B.C., 13,000/c.mm. (neutrophils 79%, eosinophils 1%, monocytes 9%, lymphocytes 11%). Latex test was negative.

Aspiration of the right hip-joint yielded 6 ml. of cloudy yellow fluid containing 3,840 polymorphonuclear leucocytes per c.mm. Culture proved sterile. Prednisolone 20 mg. daily was given for four days and then 10 mg. daily for two days. Severe pain subsided in one day, and after six days hip movement was full with pain at the limit.

The patient suffered two further severe attacks of pain, each lasting a few days, in the left hip four months later

and in the right hip a year later. Subsequently, twinges of pain occurred in the right hip. The joint retained full movement, though with pain at the limit. Latex test remained negative.

Case 7

A man aged 47 suffered pain in the left groin which caused loss of sleep. The next morning he was obliged to lift his left leg out of bed by hand and, standing, had difficulty in lifting the left foot from the ground. By evening the pain was less and by next morning had disappeared. One week later there was sudden pain in the left wrist with swelling. Recovery was complete in two days.

Two weeks later, on awakening, there was again pain in the left groin which became so severe that the patient "nearly passed out." When we saw him the next day the left hip-joint was tender. Flexion was 60 degrees, abduction was 20 degrees, and rotation was very restricted—all with pain. The left wrist was slightly swollen but not tender. Temperature 99.6° F. (37.6° C.); E.S.R., 94 mm./1 hour; W.B.C., 10,400/c.mm. Latex test was positive at 1/20.

Aspiration of the left hip-joint yielded 6 ml. of cloudy yellow fluid, under pressure, so that the piston of the syringe was pushed up in the barrel. Aspirated fluid contained 25,400 polymorphonuclear leucocytes per c.mm., and was sterile on culture.

During the next few days the left hip improved, but other joints, large and small, were transiently affected. Six weeks after onset the patient complained of stiffness in the left side of the abdomen and in the left hip, and of "pins and needles" in the lateral part of both thighs. There were well-defined areas of hypoalgesia in the territory of both lateral cutaneous nerves of the thigh. These symptoms and signs disappeared in two weeks.

Six months later there was evidence of rheumatoid type of arthritic change in the small joints of the hands. Other joints were clinically normal. E.S.R., 28 mm./1 hour. Latex test was positive.

Case 8

A man aged 21 began to have attacks of pain in his right groin five years before presenting. Onset was rapid, and pain lasted 24 hours. Incidence had been every two weeks but no attack had occurred in the last two years. There was no family history of gout.

In the present attack, pain came on in the early morning, "as though somebody were kicking his right groin." The right hip was held flexed 30 degrees, abducted 30 degrees, and in 10 degrees lateral rotation. Flexion was to 100 degrees and rotation was 20 degrees in each direction. All movement caused pain in the limit.

On admission to hospital his temperature was 98.4° F. (36.9° C.); E.S.R., 2 mm./1 hour; W.B.C., 8,400/c.mm.; serum uric acid, 8 mg./100 ml. Latex test was negative. Attempted aspiration of the right hip-joint yielded no fluid. The patient left hospital the next day, free of pain and with full movement, but limping slightly.

Two weeks later, in the evening, there was pain in the hips and knees, with "pins and needles" in the groins, thighs, and knees, and some interference with walking. Symptoms lasted only half an hour. When seen two days later his hips and knees had full painless movement and were not tender. There was well-defined hypoalgesia on the front of the thighs, in the area supplied by the femoral nerves. Quadriceps strength was normal and knee-jerks were present and equal.

Two months after onset he was free of pain and had full hip movement. E.S.R., 1 mm./1 hour; serum uric acid, 6.4 mg./100 ml.

Nine months after the first attendance there had been no further attack except twinges of pain from the right groin to the knee. Sensation in the thighs was normal. E.S.R., 1 mm./1 hour; serum uric acid, 4.7 mg./100 ml.

Discussion

In these eight patients no radiographic abnormality was seen in hip or sacro-iliac joints.

The L.E. cell phenomenon was sought in five patients but was not found.

Results of guinea-pig inoculation and culture of the aspirated fluid were negative for tuberculosis in all.

Differential Diagnosis

Before considering the diagnosis in each case, five differential diagnoses may be examined.

Acute Bacterial Arthritis.—This is the most important differential diagnosis and, at the onset, the most likely on clinical grounds. However, in retrospect there is no evidence that any of these hip-joints was infected with pyogenic organisms.

Gout.—Serum uric acid estimations were within normal limits (taking 6.5 mg./100 ml. as the upper limit of normal in men, and 6 mg./100 ml. in women), except for a reading of 8 mg./100 ml. on a single occasion in Case 8.

Ankylosing Spondylitis.—Radiographs of the sacro-iliac joints showed no abnormality.

Reiter's Syndrome.—No history of urethritis was obtained in any case. One patient (Case 3) suffered from recurrent conjunctivitis and punctate keratitis.

not involved. Rheumatoid nodules were not found in any of our patients.

Differential agglutination titres for rheumatoid arthritis, using either the Rose-Waaler sheep-cell agglutination or the latex agglutination, were positive in four patients (Cases 1, 4, 5, and 7).

Transient and symmetrical neurological symptoms and signs occurred in two patients (Cases 7 and 8), but did not amount to typical rheumatoid peripheral neuropathy, and neither patient had received steroids (Hart and Golding, 1960; Steinberg, 1960).

Six of our patients had more than one acute joint attack. Two patients (Cases 4 and 8) demonstrated repeated attacks characteristic of "palindromic rheumatism." Hench and Rosenberg (1941, 1944) defined this condition as "an oft-recurring disease of joints (arthritis, peri-arthritis, para-arthritis) apparently producing no articular residue," and regarded it as distinct from rheumatoid arthritis. However, many cases of "palindromic rheumatism" have been observed eventually to develop into rheumatoid arthritis, and most authorities now consider "palindromic rheumatism" to be one variety of this disease. Hench and Rosenberg found the hip affected rarely.

Each of our eight cases may now be considered with its particular diagnostic features (see Table).

Details of Cases

Case No.	Age when Presenting	Sex	Hip Affected First	No. of Acute Attacks		Small Joints Chronically Affected	E.S.R. mm./1 Hour (Westergren) Highest	Rheumatoid Agglutination Test	Serum Uric Acid (mg./100 ml.)	Aspiration	Follow-up Period (Months)	Diagnosis
				Hips	Other Joints							
1	68	F	No	1	—	Yes	54	Pos.	4.5	Yes	54	Rh. arthritis
2	43	M	Yes	1	—	No	82	Neg.	5.9	No	27	Uncertain
3	52	M	"	3	—	Yes	45	"	3.1	Yes (Dry)	26	Possible Reiter's syndrome
4	53	M	No	4	8	No	32	Pos.	4.3	Yes	24	Rh. arthritis "palindromic"
5	46	F	Yes	2	—	"	55	"	3.2	"	17	Possible rh. arthritis
6	34	F	"	3	—	"	24	Neg.	3.8	"	12	Uncertain
7	47	M	"	2	1	Yes	94	Pos.	5.6	"	21	Rh. arthritis
8	21	M	"	Numerous	—	No	2	Neg.	8 6.4 4.7	" (dry)	9	"Palindromic rheumatism"

Acute Pain due to Calcification in Region of Hip-joint (Jones, 1955).—Tenderness is over the deposit rather than over the joint. Radiographs in our patients revealed no abnormality.

Relation to Rheumatoid Arthritis and to Palindromic Rheumatism

Affection of the hip in rheumatoid arthritis is well recognized, but writers comment on its rarity. Forestier *et al.* (1951) state that localization in the hip is relatively rare, the onset insidious, and the course progressive; also that it occurs nearly always in cases affected with polyarthritis. Short *et al.* (1957), in a collected series of cases of rheumatoid arthritis of monarticular onset, found that the disease started in the hip-joint in 13%. They state that rheumatoid arthritis should occupy a high place in the differential diagnosis of a persistent or remittent monarticular arthritis.

Criteria for the diagnosis of rheumatoid arthritis, based on 332 cases, were drawn up by the American Rheumatism Association (Ropes *et al.*, 1956), with the object of classifying cases as definite, probable, or possible, or of excluding them.

In two of our patients the finger-joints were chronically affected (Cases 1 and 7), and in one patient the toe-joints (Case 3). Otherwise the small joints were

Treatment

It is of great practical importance to exclude acute bacterial arthritis, and in the first attack, or when it is suspected that acute bacterial arthritis may be complicating established rheumatoid arthritis (Kellgren *et al.*, 1958), the hip-joint is aspirated, under a general anaesthetic, for culture. Penicillin may be injected into the joint at the time of aspiration. Systemic antibiotic treatment is given until the next day, by which time culture is reported sterile, and even then is continued if there is any reason to believe that the arthritis may be bacterial.

Aspiration, in addition, seems to relieve pain and may aid resolution. Bed rest until acute symptoms subside is necessary. Traction is perhaps helpful. Analgesics are given.

The hip condition shows a natural tendency to resolution, and there is no evidence from this small series that steroids, chloroquine, or salicylates affected progress. Intra-articular hydrocortisone at the time of aspiration was given in one patient (Case 4), whose clinical diagnosis was palindromic rheumatism, and after immediate examination of the joint fluid had shown no organisms. This measure is of doubtful benefit, would be harmful in the case of misdiagnosed acute bacterial arthritis, and is not recommended.

Summary

Eight adult patients with acute transient arthritis of the hip are described. Follow-up periods varied from 9 to 54 months; average, 24 months.

It is important to exclude bacterial arthritis by aspiration of the hip-joint.

Two patients are diagnosed confidently, and one provisionally, as cases of rheumatoid arthritis. One patient is diagnosed as a case of rheumatoid arthritis (palindromic) and one as "palindromic rheumatism." One patient possibly suffered from Reiter's syndrome. As yet two patients remain undiagnosed.

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A FATAL CASE OF PROGRESSIVE VACCINIA

BY

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Progressive vaccinia (vaccinia necrosum, vaccinia gangrenosa) is a rare complication of vaccination which has recently been reviewed by Kempe (1960) and by Erichson and McNamara (1961). The present case is reported because of the failure of passively transferred antibody or *N*-methylisatin β -thiosemicarbazone (compound 33T57) to influence the course of the disease.

Case History

The patient, the second child of healthy parents (whose first baby had died soon after birth with a spina bifida), was vaccinated against smallpox by multiple pressure over the left deltoid region at the age of 10 months. The primary vaccination began to develop normally, but at about the same time a papule appeared on the sole of her left foot which, like the primary lesion, became vesicular in six days. These vesicles increased in size, and when we saw the child four months after vaccination the posterior half of the sole of the left foot was covered by a crusted lesion which extended on to the lateral side and also medially as far as the malleolus. The edge of this lesion was vesiculated and surrounded by a reddish areola. On the outer side of the left arm there was an irregular lesion 5 cm. in diameter with an extension towards the shoulder

consisting of raw and crusted areas with a rolled vesicular edge and a surrounding wide zone of erythema (Fig. 1).

Vaccinia virus was recovered from the lesions on the arm and the foot. The child's serum taken at this time contained no demonstrable neutralizing or haemagglutination-inhibiting antibody to vaccinia virus. The serum gamma-globulin was 0.51 g./100 ml., calculated from paper electrophoresis of the serum (Oberman, Gregory, Burke, Ross, and Rice, 1956). Examination of the blood cells showed a hypochromic anaemia (Hb 6.6 g./100 ml.) of the iron-deficiency type and a lymphopenia.

Treatment

The treatment of vaccinia complications with gamma-globulin or immune serum has been discussed by Kempe (1960). We began treatment of our patient with intramuscular injections of antivaccinia gamma-globulin (batch EPG 101) and a transfusion of fresh whole blood from an immune donor. The antibody levels and the gamma-globulin which developed in the child's serum are shown in Table I.

TABLE I

Day of Lesions	Procedure	Material Tested	Antibody Titres*		Serum γ -globulin (g./100 ml.)
			H.I.†	Neut.‡	
126	Test	Patient's serum	<5	<5	0.51
127	1 g. γ -globulin	γ -Globulin	640	1,600	—
129	1 "	"	—	—	—
130	Test	Patient's serum	5	—	0.55
131	300 ml. blood	Donor's "	80	—	—
138	Test	Patient's "	20	—	0.78

* The antibody titres are the highest effective dilutions of serum before the addition of virus.

† Haemagglutination inhibition, using the method described by Kempe (1956).

‡ Neutralization tests were kindly carried out by Professor A. W. Downie, University of Liverpool.

FIG. 1.—Vaccinial lesions 18 weeks after vaccination.

A small haematoma occurred on the back of the child's hand where the transfusion needle was placed, and 5,000 units of lyophilized crystalline alpha-chymotrypsin was given orally three times daily for five days to increase the rate of absorption. On the seventh day after the transfusion the child developed a fever of 103° F. (39.4° C.), and on the following two days it was 105° F. (40.6° C.), after which it fell to normal. It is not known whether the febrile