HETEROTOPIC OSSIFICATION COMPPLICATING PARALYSIS OF INTRACRANIAL ORIGIN

P. H. ROBERTS, ONEWSTERY, ENGLAND
From the Robert Jones and Agnes Hunt Orthopaedic Hospital, Oswestry

Heterotopic ossification may occur around joints in many pathological conditions of the nervous system, the hip being most often involved. Such new bone is common in paraplegia, and two recent papers have recorded over 170 cases (Damanski 1961, Hardy and Dickson 1963). Other cases have been reported after poliomyelitis (Costello and Brown 1951) and tetanus (Gunn and Young 1959). I have found only one report of this occurrence in hemiplegia (Irving and Le Brun 1954).

Six cases of heterotopic ossification have recently been studied. All of them occurred when the paralysis followed intracranial lesions, three of which were traumatic, two vascular and one neoplastic. In four of the cases the neurological lesion was a hemiplegia but in the remaining two, both of which followed head injuries, the initial paralysis affected all four limbs. Heterotopic bone was seen about the hips in all cases, but in one case it was also present about the shoulder and in another about both elbows.

CASE REPORTS

Case 1—A forty-one year old woman suddenly developed left hemiplegia in April 1961. Angiographs showed two aneurysms on the right middle cerebral artery, and evidence of a temporal haematoma. Craniotomy was performed and clips were applied to the aneurysms. On the third day after operation the patient became unconscious. The skull was reopened: although there was no evidence of further bleeding the brain was so swollen that a small frontal lobectomy had to be performed to allow closure of the wound.

She regained consciousness three weeks later but remained confused for several further weeks. During the eight weeks after operation her physical condition was critical. In spite of repeated blood transfusions and intragastric and intravenous protein infusions, she was constantly in negative nitrogen balance. Her physical condition then improved. However, flexion contractures had developed in the left arm and leg. Persistent lack of co-operation made physiotherapy difficult.

FIG. 1
Case 1—Radiograph of the left hip taken in January 1964. The heterotopic bone is well demarcated.
Over the next year there was no improvement in the contractures of the left arm and leg. Radiographs in July 1962 showed a mass of new bone in the region of the left hip joint. Because the patient had made little attempt to rehabilitate herself, operative correction appeared to be of little value at this stage. During the next year, however, the patient's attitude changed. As the deformity of the hip was obviously hindering her attempts to walk, it was decided to attempt surgical correction.

At the time of admission to hospital in January 1964, the left hip was fixed in 30 degrees of flexion, 45 degrees of abduction and 40 degrees of lateral rotation. There was a 30 degree flexion contracture of the knee and 5 degrees of equinus at the ankle. Some active movement was present at the knee and ankle. The left upper limb was spastic with very little active movement. A radiograph (Fig. 1) showed a mass of heterotopic bone extending from the pelvic brim to the upper shaft of the femur.

Operation and progress—Immediately after excision of the mass of bone flexion to 90 degrees was possible, and adduction to the neutral position. Fixed flexion of 25 degrees and fixed lateral rotation of 40 degrees remained. The deformity was corrected by osteotomy with internal fixation in April 1964. The left foot lapsed into a severe equinovarus deformity and a stabilisation was performed in August 1964.

The patient was discharged from hospital in December 1964. The left hip had remained in neutral position and allowed 20 degrees of flexion and 15 degrees of side-to-side movement. The knee range was from 0-90 degrees and the foot was plantigrade. The patient was walking with the aid of a tripod.

When the patient was last seen, in February 1967, the left hip had remained in neutral position and allowed flexion to 20 degrees, adduction to 20 degrees and abduction to 15 degrees. There was active flexion through 100 degrees at the knee. The patient was still walking with the aid of a tripod but was able to get in and out of chairs unaided.

Case 2—In September 1965 a forty-five year old woman was found unconscious on the floor of her home with a flaccid right hemiplegia. The left carotid pulse was diminished and in the absence of blood in the cerebro-spinal fluid a diagnosis of a carotid artery thrombosis was made.

![Fig. 2](image1.png)
![Fig. 3](image2.png)

Case 2. Figure 2—Radiograph of the right hip seven weeks after the onset of paralysis. The new bone is not clearly defined. Figure 3—Three months later. There has been no extension of the bone but a sclerotic margin has developed and a trabecular pattern has appeared within the bone.
She regained consciousness within a few hours, and apart from some emotional instability her mental state returned to normal. There was, however, no sign of recovery from the hemiplegia. Apart from minor urinary infections, which responded promptly to treatment, her general condition remained good. Seven weeks after the thrombosis a contracture had developed in the right hip and was not responding to physiotherapy. A radiograph showed immature new bone about the hip (Fig. 2).

Three months later there was still no return of function in the right side. The right hip was fixed in 30 degrees of flexion, 30 degrees of abduction and 45 degrees of lateral rotation. A radiograph showed no extension of the heterotopic bone, but its margins were now clearly defined (Fig. 3).

Operation and progress—Because there had been no return of motor function, it was decided that correction of the deformity to allow the limb to be used as a prop was all that should be attempted. An osteotomy was performed immediately distal to the lowest limit of the new bone and satisfactory correction was achieved.

The early progress after operation was satisfactory but during the sixth week a deep vein thrombosed and caused a pulmonary embolus from which she died. Necropsy confirmed the diagnosis. Unfortunately, although the new bone was recorded it was not examined further.

Case 3—A woman began to have epileptic attacks in 1956 at the age of 30. No cause for the attacks was found and out-patient treatment with anticonvulsant drugs was started.

Six years later, in 1962, she was admitted with a four-months history of loss of memory and personality changes, together with recent weakness of the left arm and leg. Arteriography showed a space-occupying lesion of the right temporal lobe and two days later a biopsy established the diagnosis of astrocytoma. The temporal lobe was swollen but it was not thought possible to remove the lesion surgically. After operation her condition deteriorated and hemiplegia became profound. Radical mega-voltage radiotherapy was followed by a
little improvement. It was noted at this time that the left leg was swollen, presumably from deep vein thrombosis.

Improvement was slow and by May 1963, six months after the biopsy, she had developed a fixed contracture in the left hip. A radiograph showed immature new bone in the region of the joint (Fig. 4). Conservative treatment was continued until by October 1964 some active movement had returned to the left arm and the lower leg. The hip was fixed in marked flexion, abduction and lateral rotation. The knee had a flexion contracture of 60 degrees with further active flexion to 100 degrees. No deformity was present in the foot. A radiograph of the hip showed that there had been no extension of the bone but that the margins were now clearly demarcated (Fig. 5).

Operation and progress—Because the patient was young and because some active movement had returned to the limb, it was decided to excise the heterotopic bone. Immediately after excision an almost full range of movement returned to the hip joint.

The early progress after operation was satisfactory, and with traction the contractures of the hip and knee were fully corrected. On the fourteenth day after operation the patient became unconscious. In spite of intensive treatment her condition deteriorated and she died on December 1. Permission for necropsy was refused. The probable cause of her unconsciousness was bleeding into the tumour.

Case 4—A twenty-one year old woman was admitted to hospital in April 1966, deeply unconscious after a road traffic accident. The right arm was spastic but the left side was flaccid. There were fractures of the right leg and of the left pubic rami and a dislocation of the left acromioclavicular joint.

The patient did not regain consciousness for two months. By this time there were flexion contractures in both elbows and the left hip. Apart from a mild urinary infection her physical condition remained good.

Improvement continued and by September she was able to walk with help. The flexion contractures had remained in spite of physiotherapy and radiographs showed mature new bone in the region of both elbows and the left hip (Figs. 6 and 7). Equinus deformity of the left foot was relieved by tenotomy.
By January 1967 there had been some return of power to the arms although spasticity was present. The right leg had returned to normal and in the left good motor function was present below the hip. The right elbow was fixed in 80 degrees of flexion, with 80 degrees of pronation but no supination. The left elbow had a range of flexion of from 90 to 100 degrees but there was a fixed supination of 45 degrees. The left hip was fixed in 45 degrees of flexion, 60 degrees of medial rotation and 30 degrees of adduction. A flexion range of from 20 to 90 degrees was present in the knee.

Operation and progress—It was thought that improvement in the position and movement of the right elbow would help the patient to use crutches. The heterotopic bone was excised from around the right elbow in January 1967, and immediately following this a passive range of flexion from 0 to 130 degrees was possible. During the next four weeks some of this range was lost but the patient was finally able actively to flex the elbow from 40 to 90 degrees and had full supination and 20 degrees of pronation.

Case 5—A sixty-five-year-old man, a known alcoholic, was living the life of a recluse. In March 1966 he was found lying on the floor of his house, having apparently had a heavy fall. He was admitted to hospital and was found to be confused and to have a right hemiparesis. During the subsequent months his condition slowly deteriorated. In September a chronic left subdural haematoma was confirmed by angiography and drained.

His condition then improved considerably and by the end of November useful active movement was present both in the arm and the leg. However, movements of the shoulder and hip were restricted and had failed to respond to physiotherapy. Radio-
graphs showed immature new bone in the region of the right shoulder and hip (Fig. 8).

Improvement continued and by January 1967 motor power was almost normal with minimal spasticity. There was good scapulo-thoracic movement but very little gleno-humeral movement. There was a 20 degree flexion contracture of the right hip, with active flexion to 90 degrees but only a trace of rotation. No contracture was present in the knee or foot. Radiographs of the hip and shoulder (Figs. 9 and 10) showed that the bone below the hip had not extended but was more clearly defined; in the shoulder it was still ill defined.

Because the bone about the hip is now mature it is unlikely that further restriction of movement will occur. Should the flexion deformity prove to be disabling, simple correction by osteotomy would be appropriate. The deformity in the shoulder is almost fixed, and because the bone is still immature it may become completely fixed. If this then causes considerable disability, excision of the bone may be necessary.

Case 6—A youth aged eighteen was admitted in April 1966, unconscious after a road traffic accident. There was spasticity of all limbs, the right arm being the least affected. There was also a double fracture of the mandible together with multiple abrasions. During the following twenty-four hours his condition failed to improve and he was transferred to a neurosurgical centre. Treatment by hypothermia was started and tracheostomy performed. At the end of six weeks there had been no improvement and the treatment was discontinued.
There was little change in his condition until six months had elapsed, when a rapid improvement occurred, so that he was soon able to recognise relatives and to talk. The improvement continued and in a further three months he was able to walk with help. The left arm was spastic, with little movement in the shoulder but some useful movement in the hand. There was equinus deformity of both feet, that of the right being severe. There was no deformity in either hip; flexion was full but medial rotation and adduction were both limited to 10 degrees. Almost full power had returned to both legs.

Radiographs showed a small plaque of mature new bone in the adductor region of the right hip (Fig. 11). It is unlikely that there will be any further extension of the bone or greater limitation of movement, and excision is not indicated.

**DISCUSSION**

Although heterotopic ossification may occur in association with many conditions of the nervous system it is most familiar in paraplegia. The rarity of its report in other circumstances may well be due to unfamilirarity and therefore, failure of recognition (Hardy and Dickson 1963). In the absence of motor recovery, contractures may be accepted without arousing the suspicion of underlying bone.

A full review of the literature is included in the papers by Damanski (1961) and Hardy and Dickson (1963), and it is not proposed to repeat or enlarge upon these. It is, however, of interest to compare certain features of the six cases presented with those seen when the bone occurs in association with paraplegia.

Hemiplegia and paraplegia have in common the loss of normal motor function. It does not appear to matter whether the paralysis be spastic or flaccid. This was noted in paraplegia by Hardy and Dickson. In Cases 1, 3, 5 and 6 spasticity was present from the outset, but in Case 2 the paralysis was flaccid. In Case 4 similar bone developed around both elbows although one was initially flaccid and the other spastic.

In paraplegia there is also loss of sensory and autonomic function and often metabolic change from urinary infection or pressure sores. Damanski (1961) postulated that profound hypoproteinaemia and severe urinary infection contributed to the production of the bone. In Case 1 hypoproteinaemia was profound and prolonged and may have been significant. In the other cases it was not evident and the urinary infection was mild.

The notable feature of all cases is the absence of known local trauma. In Case 4, although there were fractures of both pubic rami, the heterotopic bone was formed in the abductor compartment on the far side of the hip joint.

Hardy and Dickson noted new bone as early as nineteen days after the spinal injury. Damanski found none before four weeks. In paraplegia formation of bone is preceded by a soft-tissue swelling and limitation of movement. In the case of hemiplegia described by Irving and Le Brun (1954) a swelling appeared in the region of the hip four weeks after the onset of the paralysis. This was painful and was treated initially as an abscess, the diagnosis becoming apparent when the hip became fixed and the swelling bony hard. In Case 2, the bone was seen to be present seven weeks after the carotid thrombosis. There had been no pain and a swelling had not been noticed. In the remaining cases it is impossible to date the appearance of the new bone. In Case 4 mature new bone was present at five months and in Cases 3 and 5 immature new bone was seen at six months and was presumably of recent onset. In no cases was there any suggestion that the onset had been painful.

Once the heterotopic bone has appeared extension does not occur. The radiographs in Cases 2 and 3 demonstrate mature bone with the appearance of a sclerotic margin and a trabecular pattern within the bony mass. If the bone is excised when mature it does not recur. This was seen in Case 1 where there was no apparent recurrence after three years.

If the heterotopic bone is extensive it is usually associated with fixed contractures. This was seen in the hips of Cases 1, 2, 3 and 4 and in the elbows in Case 4. In Cases 5 and 6
the bone was limited in extent, there were minimal contractures and movement was still present.

_Treatment by operation_—Operation is indicated when the deformity is severe enough to prevent further rehabilitation or interfere with normal activities. In paraplegia operation is hazardous because of the labile blood pressure, local hyperaemia and poor skin healing. These objections do not usually apply in other lesions of the nervous system.

Correction of the deformity at the hip may be achieved by excision of the bone or by osteotomy. In the absence of motor recovery it may be better to perform osteotomy, allowing the limb to be used as a stable prop. However, spica fixation is usually required and this is accompanied by a considerable risk of venous thrombosis and pulmonary embolism, as occurred in Case 2. If there has been return of motor function, excision of the bone will usually produce a good range of movement, and even though all of this may not be retained a limited range is of considerable value. If a contracture persists after excision of the bone, in the presence of movement in the hip corrective osteotomy may be accompanied by internal fixation with early mobilisation.

Fixed contractures in the elbows may, apart from producing disability in the arm, hinder rehabilitation of the lower limbs. Excision of the bone should produce a good range of movement which, as in Case 4, allows the patient to use crutches.

Stiffness of the gleno-humeral joint alone may not cause severe disability and excision of bone in this region may be difficult because of the proximity of nerves and vessels. In Case 5 the heterotopic bone around the shoulder was immature and excision at this stage would in any case be inadvisable.

**SUMMARY**

1. Six cases of development of heterotopic bone around joints in association with paralysis from intracranial lesions are presented. It is suggested that such bone may occur more commonly than is realised.
2. The features of these cases are very similar to those seen in association with paraplegia.
3. Extensive new bone is usually associated with fixed contractures.
4. Operation is hazardous in paraplegia but should not necessarily be so in other paralytic conditions.
5. In the presence of returning motor function excision of the bone, allowing correction of the deformity together with some movement, is a worthwhile procedure. In the hip, osteotomy alone usually requires plaster fixation with the attendant risks of venous thrombosis. Previous excision of the bone allows internal fixation of the osteotomy with early mobilisation.

I am very grateful to Mr D. Ll. Griffiths, Mr D. Graff, Mr Rowland Hughes and Mr R. Owen for allowing me to report their cases and for their help in preparing this paper.

**REFERENCES**


