PARAPLEGIA DUE TO HYDATID DISEASE

T. J. Mills, Liverpool, England

In Great Britain hydatid disease is uncommon, and instances of hydatid cysts causing pressure on the spinal cord or nerve roots are rare. There appear to be only fifteen published cases in the medical literature in this country. The last review of spinal hydatid disease seems to be that of Colman (1899). The three new cases here recorded may therefore be of interest.

As an illustration of the rare occurrence of hydatid disease in Great Britain, Table I shows the deaths from this disease during the years 1921-50.

| TABLE I |
| DEATHS FROM HYDATID DISEASE IN GREAT BRITAIN FROM 1921-50 |

<table>
<thead>
<tr>
<th>Deaths</th>
<th>1921-30</th>
<th>1931-40</th>
<th>1941-50</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total</td>
<td>296</td>
<td>260</td>
<td>172</td>
</tr>
<tr>
<td>Per million of population</td>
<td>6:68</td>
<td>5:63</td>
<td>3:58</td>
</tr>
</tbody>
</table>

* Figures of liver disease only

The rarity of the disease in the spine is also indicated by figures from Australasia, where a "hydatid registry" was commenced in 1930. In the succeeding eighteen years there were twenty-four cases of hydatidosis of the spine, representing 1.2 per cent of all cases of hydatid disease (Woodland 1949). It is not recorded how many of these cases were complicated by paraplegia, nor was any mention made of paraplegia from extra-osseous hydatid disease.

It is likely that the number of cases in which the spinal column is attacked will always be small in view of the usual distribution of hydatid cysts in man, as recorded in the Hydatid Registry of the Royal Australasian College of Surgeons (Barnett 1945) (Table II).

| TABLE II |
| DISTRIBUTION OF HYDATID CYSTS IN MAN |

<table>
<thead>
<tr>
<th>Site</th>
<th>Per cent of all cases</th>
<th>Site</th>
<th>Per cent of all cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>Liver</td>
<td>63.3</td>
<td>Kidney</td>
<td>2.1</td>
</tr>
<tr>
<td>Lung</td>
<td>24.5</td>
<td>Spleen</td>
<td>1.3</td>
</tr>
<tr>
<td>Muscles</td>
<td>4.6</td>
<td>Brain</td>
<td>0.9</td>
</tr>
<tr>
<td>Bone</td>
<td>2.6</td>
<td>Other sites</td>
<td>0.8</td>
</tr>
<tr>
<td>(Vertebrae 1-2)</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

PATHOLOGY

Hydatid disease in the peripheral organs begins as a single primary cyst. This may contain daughter cysts which are regarded as the parasite's defensive reaction to trauma. Rupture of a cyst into muscular tissue may be followed by a crop of secondary hydatid cysts originating from daughter cysts, scoles or brood capsules. Sometimes these small secondary cysts may be forced by muscle movement some distance along connective tissue planes.

884 THE JOURNAL OF BONE AND JOINT SURGERY
HYDATID DISEASE

Hydatid infection in bone is always primary (Dew 1928). Secondary erosion of bone by an extra-osseous cyst may occur, but the cyst does not infiltrate into the bone substance. Owing to the peculiar structure and the resistant nature of bone, true hydatid disease in bone differs from hydatid disease elsewhere in two respects—namely, exogenous vesiculation occurs and there is no adventitious layer (ectocyst). As the embryo grows, some absorption of bone occurs with dilation of the bony space in which the parasite lies, but this occurs very slowly. The parasite grows along the line of least resistance, along the bony canals, to form a diverticulated type of "cyst." These cysts do not contain fluid but are composed of solid masses of hyaline material surrounding a central nuclear mass. The diverticula may become separated from the parent cyst—the so-called "beading-off" of secondary cysts. When "beading-off" occurs into the surrounding soft tissues, the secondary cysts assume the typical univesicular type of growth. Avascular necrosis may occur in the more central part of the bony lesion, leading to cavitation or pathological fracture.

When the disease occurs in or near the vertebral column the spinal cord may be compressed either by a cyst whose primary situation is in the spinal canal or by cysts arising from primary disease of the vertebrae or paraspinal tissues, which have secondarily invaded the extradural space.

**Classification**—Five types will be recognised: 1) Primary cyst in the spinal cord; 2) intradural cyst; 3) extradural cyst; 4) hydatid disease of the vertebrae; 5) paravertebral hydatid disease.

**PREVIOUSLY REPORTED CASES**

**PRIMARY CYST IN THE SPINAL CORD**

No case has been reported, but theoretically such cysts can occur.

**INTRADURAL CYST**

No case of primary cyst has been reported in Great Britain but a typical case was reported in South America (Allende and Luque 1935). The cyst was single, it occurred in a young subject, symptoms appeared early, and laminectomy with removal of the cyst led to almost complete recovery.

Instances have been reported of small multiple intradural cysts, but these were certainly secondary, as in Case 16 in the series reported later.

**EXTRADURAL CYST**

The primary cyst commonly occurs in young people and arises in the loose fatty extradural tissue. It is single and soon assumes an elongated shape as it grows along the spinal canal, and gradually produces increasing cord compression. The following recorded cases illustrate the clinical picture.

**Reported Cases**

Case 1 (Colman 1899)—Boy aged ten years. History of pain in left shoulder for three months, weakness in the left arm for three weeks, and weakness in the left leg for one week. Soon after admission to hospital he lost the power of movement in the right arm and two days later in the right leg. He then developed flaccid paralysis of the arms and spastic paralysis of the legs. At post-mortem examination, a year later, a hydatid cyst, one and a half inches long, was found lying extradurally to the left of the cord at the level of C.2–4 vertebrae. There was also a hydatid cyst in the liver.

Case 2 (Rogers and Tudhope 1938)—Girl aged nine years. History of pain in the thighs and lower back for eight months, and of flaccid paralysis of the legs with sensory loss for one month. Laminectomy was performed and a single extradural hydatid cyst, two inches long, was found at the level of the second lumbar vertebra. Complete recovery in three months.

Case 3 (Ransom and Anderson 1891)—Man aged forty-two years. History of pain in the back and shooting pains in the legs to the soles of the feet for eighteen months. Symptoms severe at first but after four months became less severe and intermittent, then returned to their severity after a further twelve months. Weakness in the legs for one month, flaccid paraplegia for two weeks, with difficulty
in passing urine. Laminectomy T.11–L.2 revealed nothing abnormal. Post-mortem examination showed a single extradural cyst attached to the lamina of T.10 vertebra. There were two cysts in the right erector spinae muscles.

This type of cyst, if diagnosed or explored early, is particularly amenable to surgery, in that it can be removed completely and recurrence is unlikely (Case 2).

If the cyst is allowed to continue growing it may make its way out of the canal by eroding the laminae or through the intervertebral foramina, and enter or rupture into the thorax or spinal muscles. If rupture occurs the patient may experience a temporary relief in symptoms (Case 3).

HYDATID DISEASE OF THE VERTEBRAE

Paraplegia occurring as a complication of hydatid disease is most frequently due to hydatidosis of the vertebral column.

The disease may begin in the bodies, pedicles or laminae, and spreads in the cancellous tissue in the same way as in other bones. The articular cartilage does not offer much resistance to the spread of the disease, and the intervertebral discs, although more resistant, are also invaded and allow spread to adjacent vertebrae (Woodland 1949).

Dew (1928) stated that usually the disease remains confined to the bone for years, but sooner or later extra-osseous extension takes place. Often the disease spreads to the paravertebral muscles, and fairly large multiple cysts may develop in them. Cysts may also extend into the mediastinum, retroperitoneal tissues or extradural space; the latter give rise to compression of the spinal cord.

Reported Cases

Case 4 (Ogle 1860)—Specimen at St George’s Hospital of a hydatid cyst within the spinous process of C.7, projecting upon the spinal cord and producing symptoms “not unlike cancer.” Few cysts in the cancellous structure of the vertebrae.

Case 5 (Moxon 1871, described by Fagge 1888)—Woman aged fifty-eight. Pain in side for eleven months, paraplegia for six weeks. Post-mortem examination showed a multilocular hydatid cyst to the left of the spine, which made its way into the canal “through” L.2 and L.3 vertebrae and compressed the cord but did not penetrate the dura.

Case 6 (Maguire 1888)—Boy aged sixteen years. History of pain and weakness in arms for three years, spastic paraplegia for two weeks. Died three weeks after admission to hospital with symptoms of meningitis. Post-mortem examination revealed many small extradural cysts between C.7 and T.6 vertebrae, and some cysts embedded in the bone of the vertebral arches. The meninges were congested.

Case 7 (Barss and Trevelyan 1899)—Man aged fifty-two years, with symptoms of acute myelitis. At post-mortem examination the cancellous tissue of the vertebrae was extensively occupied by minute cysts. Extradural cysts were found between T.1 and L.2 vertebrae.

Case 8 (Theobalds 1909)—Male aged seventy-five years. History of having thirteen worms removed from abscess over sacrum years before; paraplegia with bowel and bladder involvement for seven months. Post-mortem examination showed a swelling in the right lumbar region to be a suppurating hydatid cyst extending into the right sacrospinalis muscle as high as L.2, and into the pelvis to the left of the sacrum. L.3 vertebra was carious with cysts in the bone.

Case 9 (Ruskin 1939)—Man aged twenty-seven years. History that hydatid cysts were removed from left side of neck three years previously; paraesthesia and weakness on the left hand for three months, spastic paraplegia for two weeks. Radiographs showed erosion of C.7 and T.1 vertebrae. At laminectomy three extradural cysts were removed at level of T.1. Patient died.

Case 10 (Horsley 1897)—“... a man with symptoms of compression at the origin of the third lumbar nerve. On opening the spine at the twelfth dorsal vertebra we found it was due, not to a new growth, but to multiple hydatid cysts of the vertebral column.” Gowers (1899), referring to this case, stated that the damage had been too great to allow recovery of the paralysis in the thigh and hip muscles.

Case 11 (Tytler and Williamson 1903)—Woman aged twenty-seven years. History of removal of a hydatid cyst from the inferior angle of the left scapula four years previously; mid-thoracic pain for two months and flaccid paraplegia with bowel and bladder involvement for one month. There was a subcutaneous cyst to the left of T.3 and T.4 spines. At laminectomy fifteen extradural cysts were removed. Two years later a cyst in the scar was drained, and two and a half years later the patient still had a spastic paraplegia but sensation and bowel and bladder function were normal.

THE JOURNAL OF BONE AND JOINT SURGERY
Case 12 (Rogers 1938 and Wolfe 1943)—Boy aged ten years with spastic paraplegia for four months. At laminectomy an extradural cyst at T.6 level was removed. Complete recovery in three months. Six years later a gradually increasing kyphosis with paraplegia occurred. Laminectomy at T.3-4 level revealed a mass of hydatids arising in the bodies of the vertebrae and extending into the spinal canal and pleural cavity. No recovery after seven months. The cord was re-explored and freed from the scar tissue. Patient died later.

The prognosis in vertebral hydatid disease appears hopeless as regards complete cure. Laminectomy and removal of cysts and debris from the spinal canal may relieve the compression symptoms temporarily but recurrence of symptoms is the usual fate (Cases 10 to 12).

The cases reported by Gowers (1899) and by Tytler and Williamson (1903) cannot be regarded as cured because they were not followed up for long enough to show the ultimate fate of the patients.

It is possible that infection occurs in childhood, but because of the slow growth of the hydatid cyst in bone the symptoms do not usually occur before adult life, when destruction of the bone leads to vertebral collapse, or extra-osseous extension causes a palpable tumour or cord compression.

The average age at death in the cases reviewed was forty-one years and the average duration of life after onset of symptoms was five years.

PARAVERTEBRAL HYDATID DISEASE EXTENDING INTO THE SPINAL CANAL

A hydatid cyst, primarily in the mediastinum, retroperitoneal tissues or spinal muscles, may erode the vertebrae by pressure or may extend or rupture into the extradural tissues through the intervertebral foramina.

It may be difficult to differentiate this type of case from the primary extradural hydatid which makes its way out of the canal. However, when the size and position of the cysts and the time of onset of the nervous symptoms in relation to the other symptoms and signs are taken into consideration, the types can usually be differentiated.

Reported Case

Case 13 (Brailsford 1931)—A large hydatid cyst of the superior mediastinum eroded the vertebral bodies and caused pressure on the cord, and eventually ruptured into the spinal canal.

UNCLASSIFIED CYSTS

The location of the primary cyst in the following two cases could not be determined from the published data.

Case 14 (Morgan 1877)—Woman with symptoms and signs of caries of the lumbar vertebrae. Post-mortem examination showed the spinal cord to be atrophied in its last one and a half inches to the size of a crow quill from pressure of a hydatid cyst. The cyst occupied the spinal canal to the right of the cord and, escaping through T.10-11 foramen, swelled out into a large elongated mass which entirely filled the hollow of the abdomen to the right of the spine.

Comment—This might well have been primarily an extradural cyst. On the other hand, the description could be compatible with cysts in the paravertebral tissues which secondarily penetrated through T.10-11 foramen (compare Case 16).

Case 15 (Murchison 1877)—Woman aged forty years with symptoms of paraplegia and retention of urine. Post-mortem examination showed the pleura to be separated from the ribs and sides of the vertebral bodies by two hydatid cysts, one on each side. The spinal cord at this place is considerably smaller than elsewhere.

Comment—The primary disease may have been in a thoracic vertebra or it may have been an extradural cyst which had made its way out of the canal through the intervertebral foramina on either side (compare Case 18).
NEW CASE REPORTS

In 1952, while attached to the Royal Infirmary, Liverpool, I had personal experience of a case of paravertebral hydatid disease which terminated in paraplegia. The sacrospinalis muscle was the primary site of the disease.

Case 16 (Mr Norman Roberts’s case)—Man aged forty years who had been in close contact with dogs all his life complained of low back pain and right-sided sciatica for eleven weeks. History—Ten years ago, swelling right lumbar region, occasional tenderness, fluctuated in size. Symptoms disappeared with physiotherapy. One year ago attack of low back pain with swelling in right lumbar region. Symptoms disappeared after physiotherapy.

Examination—Patient distressed; flat, rigid lumbar spine, tilted to right. Straight leg raising right 30 degrees, left 50 degrees. Right knee jerk diminished. Radiographs normal. Diagnosed as a disc lesion.

Treatment—Bed rest followed by plaster jacket. Temporary improvement, then symptoms recurred. Subsequent radiographs showed generalised osteoporosis of lumbar spine. Blood chemistry and urine normal. Symptoms responded to weight traction and plaster jacket. Three months later the patient was readmitted with a complaint of three weeks’ difficulty in defaecation, one week’s difficulty in micturition. Previous day had pain in thoracic region. On examination, semi-conscious, evidence of loss of weight, bladder grossly distended, weakness in legs (tendon jerks not elicited, plantars equivocal), neck rigidity. Blood urea normal. Lumbar puncture—bloodstained fluid, low pressure. After epileptiform attacks the patient died.
Post-mortem examination—A large multilocular hydatid cyst occupied the right sacrospinalis muscle from T.12 to L.5 (Fig. 1). The main body of the cyst was occupied by daughter cysts, and it had grown in an infiltrative manner, passing through the intervertebral foraminae in the lumbar region. There were a number of small cysts lying extradurally up to the level of T.12. Posteriorly, the continuity of the dura was lost, and some small cysts were found in the subarachnoid space in the lumbar region (Fig. 2). Brain and meninges normal; other organs normal.

Mr Sutcliffe Kerr told me of a patient on whom he operated during the war years. This was a case of hydatidosis of the vertebral column which presented with paraplegia. Unfortunately the case notes and radiographs cannot be traced and re-examination of the patient has not been possible.

![Fig. 3](image_url)

Case 18—Myelographs showing a space-occupying lesion (hydatid cyst) at the level of the fourth thoracic vertebra.

Case 17 (Mr Sutcliffe Kerr's case)—Middle-aged female with symptoms and signs of a spastic paraplegia. Radiological examination showed lesions in some thoracic vertebrae and adjacent ribs, which were diagnosed as being due to hydatid disease. Laminectomy was undertaken to relieve the symptoms and multiple extradural cysts were found and removed. She left hospital in good general health.

The following case of an extradural hydatid cyst giving rise to paraplegia is under the care of Professor Norman Dott at the Royal Infirmary, Edinburgh. It illustrates the excellent results and good prognosis which follow timely surgery in this type of case.

Case 18 (Professor Norman Dott's case)—Man aged thirty-nine years. First admitted to hospital in 1948 complaining of difficulty in walking for three months.
History—Three months ago had a minor injury to back. Noticed his legs felt stiff and difficult to control; occasional “jerking in bed.” For five weeks urgency and frequency of micturition, desire to defaecate with little result. No history of contact with dogs.

Examination—Signs of mild spastic paraplegia. Impairment of sensation below eighth rib. Tenderness over T.6 spine. Lumbar puncture—Pressure 200 millimetres, rapid rise on jugular vein compression but slow fall. Protein 90 milligrams per cent.

Radiographic examination—Thinning of pedicles T.4. Myelography—Block between lower border T.3 and upper border T.5 (Fig. 3).

Operation—Laminectomy, with total removal of T.4 lamina, and partial removal of T.3 and T.5 laminae. A hydatid cyst was found lying in an excavation in the vertebral bodies, anterior to the cord and extending in dumb-bell form through the intervertebral foramina on either side. The cord was compressed and indented from the front, and the T.4 nerve roots were stretched over the lateral extensions of the cyst. The operation field was draped off, the cyst opened and its contents (including daughter cysts) were removed by suction. The lateral extensions of the cyst were freed with difficulty from the intervertebral foramina, the right T.4 nerve root was severed extrathecally and used to rotate the cord gently to facilitate removal of the central part of the cyst wall from the anterior surface of the dura. The cord now lay like a bridge across the excavation in the vertebral bodies. Formalin was not used for fear of damage to delicate structures. The wound was closed, completing the operation.

Histological examination—Echinococcal cyst, complete with brood capsule and two rows of hooklets.

Progress—Within four weeks the patient was walking normally. No bowel or bladder trouble. Six years later he was well, and had no recurrence of any symptoms.

TREATMENT

In the absence of any efficient medicament treatment, complete surgical extirpation of the parasite is the only possible policy. This is more easily accomplished before the primary hydatid cyst has ruptured, and therefore successful treatment depends on the early recognition of the disease.

Extraspinal hydatid disease should be dealt with radically by surgery and the cavities swabbed out with 5 per cent formalin to kill any live parasitic tissue that may remain. Minor symptoms and signs may be present for years in extraspinal disease, and if cord compression supervenes it is a late complication (Case 16).

In osseous hydatidosis, nothing less than complete excision of the affected bone is curative. Curettage and formalinisatin (Dew 1928) and deep x-ray therapy (Coley 1932, Howorth 1945) have both proved ineffective. When symptoms of vertebral disease occur extra-osseous extension has usually taken place and extirpation of the disease may be impossible.

As regards the management of a case of cord compression from primary or secondary hydatid cysts, Professor Dott has kindly sent me the following observations on the surgical treatment.

1. One can only occasionally suspect the presence of this condition, from the history and background of the patient, from the presence of cysts elsewhere, from the Casoni test, and possibly from the radiographic appearance of the bone (Case 17).

2. With knowledge of the above, recognition of the disease at operation (the usual laminectomy for spinal tumour) is very important.

3. The treatment then is like that of these cysts elsewhere in the body, thus: carefully drape the wound. Very carefully remove all the cysts (suction is particularly useful): great care must be taken to prevent the dissemination of small daughter cysts and scolices in the wound. The dura should also be opened to allow careful intradural exploration, as not infrequently intradural cysts are present. Special care must be taken to remove any intervertebral foraminal extensions. In contradistinction to operative treatment for this disease elsewhere in the body no fixative agent such as formalin or alcohol should be used, because of the obvious danger to the nervous tissues.

4. Careful follow-up is necessary to check any local recurrence and to observe if any hydatids are developing elsewhere.
PARAPLEGIA DUE TO HYDATID DISEASE

SUMMARY

1. Cases of hydatid disease causing paraplegia since 1860 in Great Britain have been briefly reviewed and three recent cases have been added.
2. The types have been classified, the clinical pictures outlined and the treatment discussed.

It is a pleasure and a privilege to thank Professor Norman M. Dott for his great kindness in allowing me to use his clinical records and radiographs, and for his contribution to the discussion on treatment. I am most grateful also to Mr Sutcliffe Kerr for permission to record his case and to Mr Norman Roberts, whose encouragement made this paper possible. To Professor Bryan McFarland and Mr Robert Roaf my thanks are due for their advice and most helpful criticism. The pathological specimens were prepared by Dr H. T. G. Strawbridge of the University of Liverpool. The Registrars-General of England and Scotland and the Welsh Board of Health kindly supplied me with statistics.

REFERENCES


