

A REVIEW OF HYDATID DISEASE OF THE BRAIN

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Of all the parasites infesting the brain, the larval stage of one or other of the cestodes or tapeworms is by far the commonest. By and large, parasitic infestation of the brain is of rare occurrence and does not lend itself to ready diagnosis and, at best, can only be strongly suspected in sheep-rearing countries, when the doctor is confronted with a patient under middle age, with raised intracranial pressure and angiographic changes suggestive of a large cyst out of all proportion to the amount of disability produced. It is clear that the condition should be suspected on clinical grounds, and further investigated by angiography rather than ventriculography, in order to eliminate the danger of puncturing the cyst with resultant dissemination of daughter cysts and the almost inevitable recurrence. This is only true when dealing with hydatid disease caused by the larval stage of *Echinococcus granulosus*, and this paper will deal with this type in some detail; the other types will merely be mentioned, as they are rarer, present a different clinical picture and offer a different diagnostic problem.

1. The larval stage of *Echinococcus granulosus*:

The adult tape worm is commonly found in the duodenum of the dog and other carnivora, and when full-grown is 3-6 mm. long, consisting of four segments—a scolex, which carries four suckers and a double row of hooklets; the immature proglottid; the mature proglottid, followed by the terminal gravid segment, which carries from 500 to 800 ova in various stages of development.

The ovum, 45 μ in diameter, and brownish in colour, has a chitinous envelope which is resistant to heat and cold. When swallowed by the intermediate host, it hatches into an active hexacanth embryo, which makes its way into the portal vein and is carried throughout the body. The majority of the embryos are filtered out in the liver, the commonest site for hydatid disease; the next commonest is the lungs and the spinal column and the brain follow in lesser frequencies.

Within a very short time a cyst develops with fluid on the inside of the germinal epithelium and a laminated membrane of hyaline material on the outside, which constitutes the true capsule of the bladder-like cyst. A false capsule forms around the developing cyst, which is a fibroblastic reaction by the host; this is seen in the brain as a flimsy neuroglial reaction. Not all of the germinal epithelium is fertile, but

budding occurs at a thousand places, growing towards the cyst cavity as scolices, daughter cysts and grand-daughter cysts in various stages of development. They become characteristically detached from the parent cyst wall and occur as 'hydatid sand' or sediment in the parent cyst.

There is always a tendency for a hydatid cyst to become multi-vesicular, but multi-loculation or exogenous budding of brood capsules appears to be an effect of pressure exerted by the tissues of the host, particularly irregular pressure. This is well demonstrated by our series, where all the cysts except one was single when the site was in the brain, in contrast to the multiple cysts encountered in the spinal column.

The cysts are slow-growing and may reach a large size before producing symptoms. If lodged in the spinal column they produce symptoms earlier and do not reach a size greater than 1 cm. in diameter before the patient seeks medical advice.

The nerve tissues are not infiltrated but are pressed aside by the developing cyst. It shows some predilection for the cerebrum, and is decidedly rare in the cerebellum. The symptoms are characteristically intermittent due to intermittent oedema, but the reason of this phenomenon is still unknown.

2. *Cysticercus cellulosae*, the larval stage of *Taenia solium*, a cestode or tape worm commonly found in man. Humans can be infected by swallowing the eggs passed in human faeces, or by regurgitation of the intestinal contents into the stomach as a result of gastric disturbance, which is equivalent to swallowing thousands of eggs, or by ingesting pig's flesh in the uncooked form.

The liberated oncosphere penetrates the intestinal wall and gains access into the lymphatic or blood stream to be disseminated into the muscles, subcutaneous tissue, eye and brain. The resulting bladder worm, *Cysticercus cellulosae*, measuring 5 \times 8 mm., shows a great tendency to calcify and be diagnosed with great facility by taking plain X-rays of the brain and muscles of a suspected case. When lodged in the brain they tend to produce epilepsy.

3. *Coenurus cerebralis*, the larval form of the tape worm, *Multiceps multiceps*, found in the brain of man, sheep, goats, cattle and antelopes. The cysts are smaller, up to 2 cm. in diameter, and are usually multiple. They are found in the

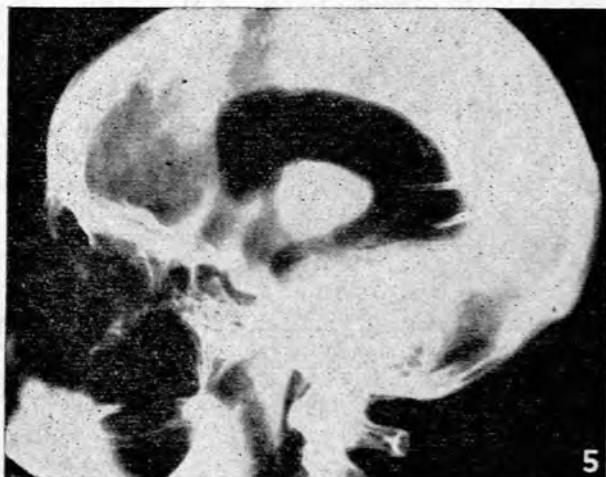
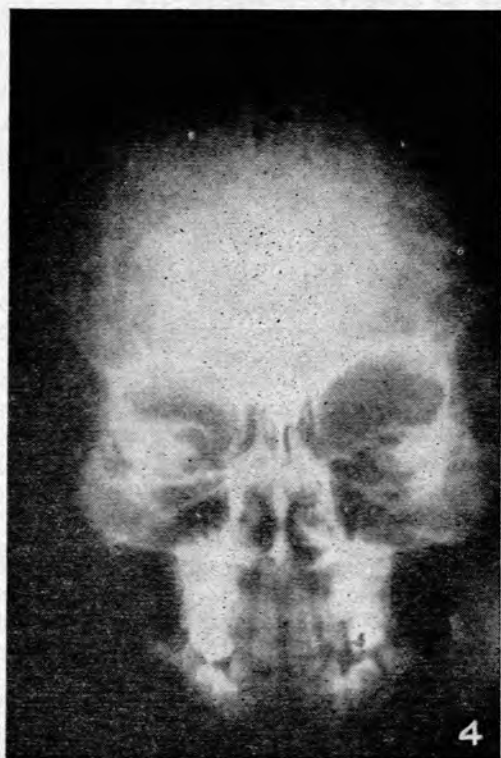
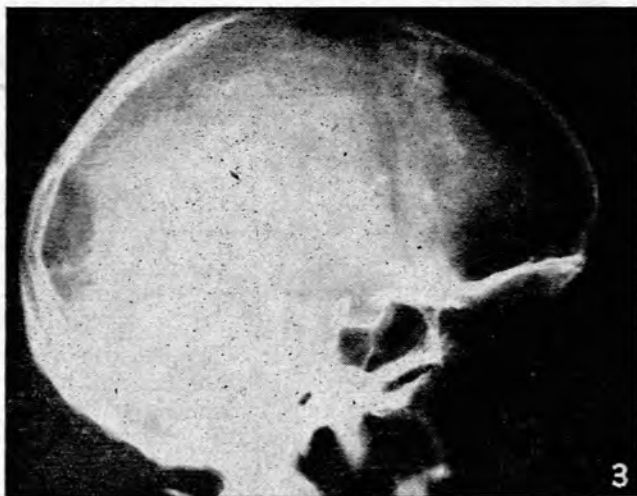
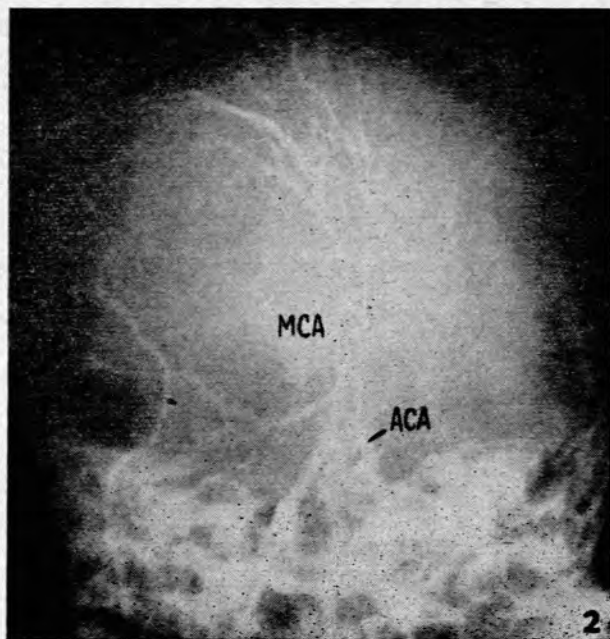
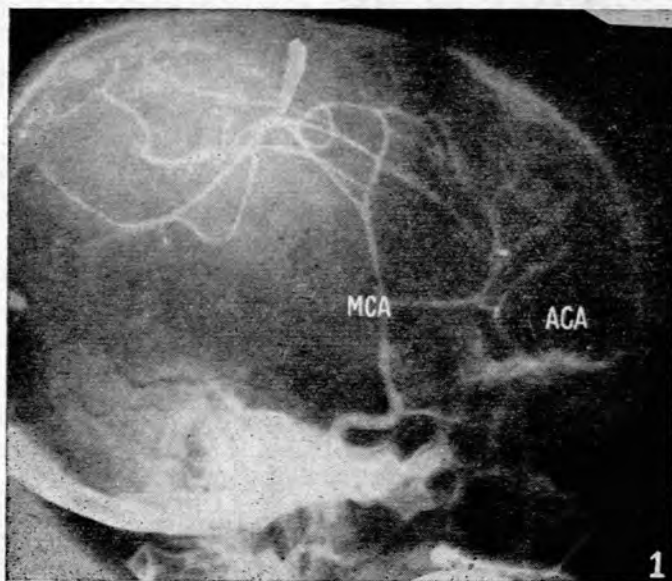


Fig. 1. The lateral projection shows elevation of the middle cerebral vessels (M.C.A.) actively stretched over an avascular space-occupying mass in the right fronto-temporal region.

Fig. 2. A.P. Projection shows marked displacement of the middle cerebral artery (M.C.A.).

Fig. 3. Lateral projection showing numerous calcified cysticerci.

Fig. 4. A.P. projection showing a calcified extra-dural hydatid cyst.

Fig. 5. Lateral projection showing a ventriculogram and a large hydatid cyst in the third ventricle.

ACA=Anterior cerebral artery.

cerebrospinal fluid pathways causing a hydrocephalus, with increased protein and cells in the cerebrospinal fluid. They vary in shape and are often found in clusters. Through the semi-transparent cyst wall, the cloudy contents can be seen, which are calcified bodies and not scolices. Clusters of scolices can be made out on the inner wall, and remain characteristically attached to the wall of the cyst.

MATERIAL

The present study is based on a series of 20 cases of hydatids in the central nervous system, of which 14 had the cysts in the brain and 6 in the spinal column.

HYDATID DISEASE OF THE BRAIN

1. J.G. (E.M.) Age 18: Three months before admission he complained of weakness of the right limbs, followed by pain in the neck, nausea and vomiting two months later, and double vision ten days before admission. On examination, papilloedema and right hemiparesis. Blood picture normal. X-rays of skull: signs of increased intracranial pressure. Ventriculogram abnormal. Site: Left fronto-parietal. A single cyst, 5 cm. in diameter was removed. Complete recovery with occasional fits. No recurrence.

2. H. de K. (E.F.) Age 13: 7 months before admission she developed a right hemiparesis, followed by headaches and double vision 4 months later. Jacksonian seizures set in 4 days before admission. On examination, bilateral papilloedema and right hemiparesis. Blood picture normal. X-rays of skull: normal. Electro-encephalogram: abnormal. Ventriculogram showed left parietal mass. A single cyst, 6 cm. in diameter, was removed. She made a partial recovery, with an occasional seizure. Three years later she had only a slight residual hemiparesis and the last seizure had occurred 8 months previously.

She presented a year later with increasing weakness of the right limbs and dysphasia. Angiography revealed a large cyst in the left parietal region. At operation a large cyst was removed, which proved to be a porencephalic cyst and not a recurrence of the hydatid disease.

3. P.K. (E.M.) Age 23: 5 months before admission he experienced headache and difficulty to concentrate. Two months later he noticed a right temporal field defect and tremors of the right hand. Two weeks before admission he experienced spells of diplopia and a mild dysphasia became noticeable, superimposed on his previous tendency to stutter. On examination, bilateral papilloedema; right lower temporal quadrantic field defect; slight dysphasia and weakness of the left side of the face, of a central type. X-rays of skull: normal. Electro-encephalogram: abnormal. Blood picture normal. Angiography revealed a cyst in the left temporo-parietal region. At operation a large, left temporo-parietal cyst, 7 cm. in diameter, was removed. Complete recovery followed. A year later a moderate-sized hydatid cyst was removed from the left trapezius muscle.

4. D. v. Z. (E.M.) Age 12: 3 months before admission he experienced right-sided Jacksonian fits, a month later, right hemiparesis and 2 months later, headache and nausea. On examination he had a right hemiparesis and papilloedema. Skull X-ray showed springing of sutures. Normal leucocyte count. Electro-encephalogram abnormal and angiogram suggestive of a cyst. Two large hydatid cysts were removed from the left parietal region. Complete recovery.

5. A.J. (C.F.) Age 5: Progressive weakness of lower limbs and right upper limb, and difficulty with micturition for six months. On examination the patient was found to have bilateral papilloedema and a right hemiparesis, including the face. Skull X-rays showed springing of the sutures. Blood count within normal limits. Ventriculogram demonstrated a large fronto-parietal mass on the left side, extending to the midline; and marked dilatation of the lateral ventricles. At operation a large, single hydatid cyst was removed from the left fronto-parietal region. The child died three days later.

6. J.A. (C.M.) Age 17: Weakness of the right leg for 2 years, followed by weakness of the right arm 18 months later. Head-

aches and deterioration of vision for 3 weeks. On examination, bilateral papilloedema with secondary optic atrophy and old haemorrhages. Right spastic hemiparesis. X-rays of the skull abnormal, showing prolonged pressure effect. Blood count within normal limits. Angiogram showed a large left fronto-parietal avascular space-occupying mass. At operation a large hydatid cyst was removed. The hydatid skin test was negative and the flocculation test positive when these were done post-operatively. When discharged, 3 weeks later, he had some residual weakness of the right foot.

7. D.K. (C.F.) Age 19: A few days before admission, the patient became aware of weakness of the right limbs, headache and diplopia. On examination, bilateral papilloedema with exudates, and paresis of the right limbs, including the face. Reflexes brisk on the right side and a right extensor plantar response. X-ray of the skull showed a small area of calcification in the left cerebral hemisphere. Angiography confirmed an avascular mass in the left parietal region. At operation a single, large hydatid cyst was removed from the left parietal region. She made a complete recovery.

8. P.K. (E.M.) Age 5: The patient developed weakness of the right side 6 months before admission, followed by headaches and fits. On examination, he presented with a right hemiplegia, including the face; bilateral papilloedema. Ventriculography demonstrated a mass in the left lateral ventricle. At operation a large hydatid cyst was removed from the left lateral ventricle. Complete recovery resulted.

9. (E.M.) Age 46: Headaches and fits for one year. On examination no abnormal neurological signs could be demonstrated. X-ray of the skull showed a calcified mass in the left frontal region. A large calcified hydatid cyst was removed from the left frontal region. Complete recovery.

10. M.C. (E.F.) Age 9: A week before admission, the child developed peri-orbital swelling of the left eye, followed by headache, vomiting and unsteadiness on the feet a few days later. On examination, bilateral papilloedema; partial third cranial nerve palsy on the left; and a right facial paresis. Blood picture normal. X-rays of skull, abnormal. Ventriculogram demonstrated a space-occupying lesion in the left fronto-temporal region. At operation a large extradural cyst was removed, followed by complete recovery.

11. D.J. (C.F.) Age 13: One year previously she had a generalized epileptic seizure, followed by a left hemiparesis 6 months later. Headache and vomiting shortly before admission. On examination, bilateral papilloedema and a left hemiplegia, including the face. Electro-encephalogram abnormal. Blood count within normal limits. Angiography demonstrated an avascular mass in the right temporo-parietal region. At operation a large, single hydatid cyst, 8 cm. in diameter, was removed. She made a full recovery.

12. P.N. (C.M.) Age 18: 15 months before admission, the patient felt dizzy on standing or walking. One month later he experienced weakness of the left limbs. On examination, a left spastic hemiplegia, including the face, was found. Blood picture normal. Lumbar puncture—normal pressure and normal constituents. Ventriculography demonstrated a large, right temporo-parietal tumour which, at operation, was found to be a large, single hydatid cyst. Complete recovery took place.

13. C.H. (C.F.) Age 50: Patient complained of weakness of the left limbs for four weeks before admission. On examination she had bilateral papilloedema with haemorrhages; and a left hemiplegia. Ventriculography demonstrated a right parietal mass. She died 6 hours after ventriculography. At autopsy, a hydatid cyst of the right parietal region was found.

14. C.W. (C.F.) Age 34: 4 months before admission the patient experienced pain in the frontal region of her head, accompanied by vomiting; 3 months later her vision deteriorated. On examination there was bilateral papilloedema and moderate visual field constriction. Paresis of the right rectus oculi muscle. Electro-encephalogram—abnormal. Ventriculography demonstrated a large filling defect of the posterior part of the third ventricle. A right frontal craniotomy was performed and the third ventricle explored through the right lateral ventricle, but no tumour was encountered. 1 c.c. of Myodil was instilled into the third ventricle, which outlined the whole of the third ventricle. The patient died

and at autopsy, a small cyst, 3 cm. in diameter, was found, which proved to be a hydatid cyst on histology.

HYDATID DISEASE OF THE SPINAL COLUMN

1. G.N. (C.M.) Age 42: 3 years before admission the patient experienced backache. Two and a half years later he experienced pain in both legs, aggravated by coughing; followed by weakness of both legs and difficulty in initiating micturition; and a band of numbness of the left groin. On examination, weakness of both legs; depression of vital sensation in the sacral dermatomes and in the T 12 to L 3 dermatomes on the left side; and patchy depression of sensation on the right side. The abdominal and cremasteric reflexes were absent on the left side—knee and ankle jerks absent on both sides. X-rays of the spine showed erosion of the left pedicle of Lumbar 1 vertebra. Lumbar puncture showed a complete block, with 1,600 mgm.% protein, globulin +++ and 2 lymphocytes. Myelography showed a complete block at L 1. At operation a large number of small hydatid cysts were removed from the extradural space at L 1 and 2. On discharge, a month later, there was minimal residual weakness of the legs and only slight depression of sacral sensation.

2. J.J. (C.M.) Age 12: 4 months before admission he developed a sudden pain in the back, followed by paraplegia a few days later. He had experienced difficulty with micturition during the latter 2 months. On examination, the patient presented with a spastic paraparesis, with only slight movement in the feet: diminution of vital sensation below L 1; sense of position and passive movements defective in knees, ankles and toes. Reflexes very brisk and bilateral ankle and knee clonus; bilateral extensor plantar responses. Lumbar puncture was performed and only a few drops of cerebrospinal fluid obtained. X-rays of the spine showed a paravertebral shadow and some destruction of the body of T 11. A small, single hydatid cyst was removed at T 11. He made an incomplete recovery, only in so far as sensation and joint sensation were concerned.

3. S.B. (E.F.) Age 18: This patient experienced numbness of the left leg 7 months before admission, followed by numbness of the right leg a month later. Weakness of the legs set in 3 months later, and she was unable to walk for the 2 weeks immediately before admission. On examination there was a spastic paresis, with only minimal movement in the right foot. Vital sensation was lost below the level of L 1 and joint sensation absent in all the joints below the knees. Reflexes were increased. Lumbar puncture revealed cerebrospinal fluid that clotted on standing, and Queckenstedt's test was positive. Myelography demonstrated a complete block at T 10. At operation, 3 small hydatid cysts were removed at T 10. She made a complete recovery.

4. M.T. (E.F.) Age 70: One month before admission she experienced girdle pains, aggravated by movements. Two weeks later, she woke up one morning to find her legs completely paralysed. She was numb below the umbilicus and had difficulty in passing urine. (Coughed up a hydatid cyst two years previously.) On examination, there was a flaccid paralysis of both legs; vital

sensation depressed below T 9, and sense of position and passive movements lost in the legs. Abdominal and deep reflexes of the legs abolished. Blood picture normal. Casoni test negative. Myelography demonstrated a complete block at T 10. Several small hydatid cysts were removed from the extradural space at T 10. Incomplete recovery resulted.

5. D.B. (E.F.) Age 10: Clumsiness of the legs was noticed 6 weeks before admission, and patient unable to walk for 5 days before admission. On examination, a spastic paraparesis; depressed sensation to pin-prick below T 8; position and motion sense not affected; knee and ankle jerks brisk; abdominal reflexes absent and bilateral extensor plantar responses. X-rays of the spine showed erosion of the pedicle of T 6 on the left side. Lumbar puncture revealed a low pressure and an impeded rise and fall on jugular compression. Protein 450 mgm.% and 4 lymphocytes. Myelography demonstrated a complete block at T 5/6, and several hydatid cysts were removed from the extradural space. Recovery was incomplete.

Improvement continued for 2 years, when she developed weakness of the legs and difficulty with micturition. On examination a spastic paraparesis was present and loss of deep sensation in both legs. Myelography demonstrated a partial block at T 4/5 and a complete block at T 12. A laminectomy at this level revealed adhesions due to arachnoiditis. No improvement followed and she was again explored at T 4/5 and 20 small and one large hydatid cysts were removed. Recovery was rapid and virtually complete.

6. M.L. (E.F.) Age 26: The patient developed weakness and numbness of both legs 3 months before admission. On examination, spastic paraparesis of both legs and vital sensation depressed below T 7. Myelography demonstrated a complete block at T 4. Cerebrospinal fluid showed protein value of 127 mg.%. At operation several hydatid cysts were removed from the extradural space at the level of T 4. Complete recovery resulted.

DISCUSSION

In our series, 6 of the 14 cerebral cases were children. There were 7 females and 7 males. The age does not affect the manner in which the disease manifests itself. It is commoner in the young adult—only 3 cases were older than 23 years of age. All of them had resided on farms for the major part of their lives.

Ten of the 14 cerebral hydatids presented with hemiplegia. 6 had focal or generalized epilepsy at one or other stage of the disease. 10 experienced headache due to raised intracranial pressure. 12 of the 14 cases had papilloedema. All but one case had single cysts—average 7–13 cms. in diameter. All the cysts were supratentorial, 1 in the extradural space, 8 in the left hemisphere, 1 in the left ventricle, 3 in the right hemisphere and 1 in the third ventricle. All the cases were

TABLE I. DATA IN 14 CASES OF HYDATID OF BRAIN

Case	Age in Years	Duration of Symptoms before Operation (months)	Headache	Fits	Hemi-plegia	Papill-oedema	Site (Hemisphere)	Result
1	18	3	—	+	+	+	L	Cured
2	13	7	+	+	+	+	L	Improved
3	23	5	+	—	—	+	L	Cured
4	12	3	+	—	+	+	L	Cured
5	5	6	—	—	+	+	L	Died
6	17	24	+	—	+	+	L	Improved
7	19	1 week	+	—	+	+	L	Cured
8	5	6	+	+	+	+	L	Cured
9	46	12	+	+	—	+	L	Cured
10	9	1 week	+	—	—	+	L	Cured
11	13	12	+	+	+	+	R	Cured
12	18	15	—	—	+	—	R	Cured
13	50	1	—	—	+	+	R	Died
14	34	4	+	—	—	+	IIIrd ventricle	Died

operated on and histologically confirmed. 3 cases died—1 after investigation by ventriculogram.

None of the cases had demonstrable hydatid disease elsewhere in the body. 1 developed hydatid cysts in the trapezius one year after the original operation—1 had coughed up a hydatid cyst 2 years before. No case had a recurrence of the disease in the brain.

The condition can be strongly suspected on clinical grounds when a young adult or child presents with headaches, hemiparesis, fits and papilloedema of gradual onset, and further investigations should be restricted to angiography rather than ventriculography for fear of puncturing a posteriorly situated cyst. Roma-Arana-Iniguez *et al.* drew attention to the characteristically fine, bilateral tremor, not accentuated by closing the eyes, and disappearing shortly after operation. The aetiology is obscure, but it may be a toxic element or the effect of generalised pressure on the basal nuclei.

We have long ago discarded the hope that eosinophilia, skin reactions or complement fixation tests are of any help. Some workers have tried the antigen obtained from human hydatids, with better results. Plain X-rays of the skull are not helpful in differentiating it from other space-occupying lesions. They may show signs of raised intracranial pressure, such as erosion of the clinoids, springing of the sutures, silver-beaten appearance of the skull and displacement of the calcified pineal. When calcified, it cannot be differentiated from calcifying tumours, arteriovenous anomalies, or tuberculomata.

Ventriculography should be avoided when there is associated hemiplegia, or when hydatid disease is suspected on clinical grounds. This is in sharp contrast to *Cysticercus cellulosae* and *Coenurus cerebralis*, which very commonly block the cerebrospinal fluid pathways, in which case ventriculography, supplemented by Myodil ventriculography is the chief means of investigation.

Angiography is the ideal method of investigation and, more often than not, the condition can be correctly diagnosed. There is usually gross displacement of the vessels, circumscribing a regular avascular area, typical of a cystic mass. The size of the 'tumour' is usually surprisingly big and not in keeping with the extent of physical disability produced.

Electro-encephalography is not very helpful and there is nothing pathognomonic in those performed on our cases. None of our cases demonstrated an area of electrical silence surrounded by delta waves—as described by Foster, Castells and Gastant. In our cases diffuse abnormality throughout

the brain, suggestive of hydrocephalus, and delta wave activity in the region of the cyst, surrounded by an area of delta wave activity of high voltage, are more usual.

TREATMENT

The treatment of hydatid disease of the central nervous system, is the removal of cysts *in toto*, without rupturing them. Many methods have been described, but we have found the method described by San Julian *et al.* to be the easiest and most efficacious. It involves a liberal craniotomy, the location of the cyst by means of a brain cannula, used very gently to avoid puncturing the cyst wall, followed by an incision through the cortex in a radial fashion. The line of cleavage between brain and cyst is defined, and further isolation of the cyst is effected by a gentle jet of water through a catheter, the tip of which is inserted between cyst and brain, assisted by gravity by tilting the head, so that the cyst is lowermost.

The mortality rate is low, the morbidity rate low and, in most cases, the recovery is complete. In the spinal cases, all improved and 3 made complete recoveries.

SUMMARY

The pathology of hydatid disease has been discussed, with special reference to the differentiation of hydatid disease caused by the larval stage of *Taenia echinococcus* from *Taenia solium* and *Multiceps multiceps*.

The clinical diagnosis, or suspicion, is important in order that further investigations be restricted to angiography, rather than ventriculography. The left hemisphere is by far the commonest site. Treatment is by means of enucleation assisted by a jet of water, and gravity. The prognosis is good.

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