HYDATID CYSTS OF THE BRAIN
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E ver since the beginning of this century hydatid (Echinococcus) cerebra l cysts have drawn the attention of River Plate authors. In 1901 Morquio published the first case report in Uruguay. Since then, a large number of publications have dealt with the subject. Argentine literature has also often been concerned with brain hydatids from 1893 onwards.

Case reports in Uruguay number over 100. On the other hand, Phillips, in Australia, reported 29 cases spread over the last 60 years.

General Notions on Echinococcosis with Special Reference to Cerebral Localization. According to Mackie hydatid disease is most widespread in Algeria, Egypt, Cape Colony, Palestine and Syria, South Australia, Tasmania, New Zealand, Central Europe, Argentina, Uruguay and Paraguay. It is also frequently met with in Spain and Italy. These are all countries noted for the raising of cattle and sheep.

It is interesting to note the distribution of cysticercosis in Latin America. It is very common in Mexico, Peru and Chile, where hydatid disease is a rare occurrence. On the other hand, cysticercosis is hardly ever encountered in Uruguay or Argentina.

Two per cent of all hydatid cysts are found in the brain. The cyst is nearly always solitary in the brain (primary localization); but multiple cysts may develop within the brain if a cyst that is attached to the wall of the left lateral ventricle of the heart ruptures.

According to Dew they are seven times more common in children than in adults. Schroeder and Medoe found 9 adults in 42 cases.

The hydatid cyst grows readily in the brain, a soft, easily displaced tissue; hence, it may attain a large size, keeping its normal hyaline cyst capsule with no daughter vesicles. Nearly every complicated case is the result of rupturing a single cyst. Among our 13 cases we found one suppurating cyst, with daughter vesicles, a very rare occurrence.

Infestation with the disease occurs through the ingestion of dogs' feces containing Taenia echinoceclus rings. Children may suffer contamination directly from such feces, as do cattle, pigs, and sheep. Contamination of water and foods, principally salads, by such canine feces is probably a more important source of contagion. Contamination by way of the air is still controversial. It could account for pulmonary infection.

The rings deliver eggs, which upon reaching the digestive tract detach from their membrane, freeing a hexacanth embryo 0.025 mm. in diameter. This embryo then passes through the intestinal wall into the blood stream.
After this, the hexacanth embryo has to go through two capillary filters, i.e., the liver and the lung, before it can reach other parts of the body. Hence its high occurrence in liver and lung and rareness in the brain.

In a previous report with Asenjo, the senior author already called attention to the fact that although the hexacanth embryos of the Taenia solium and the Taenia saginata (the latter having the same size as the Taenia echinococcus), penetrate the organism in the same way and have to pass through the same capillary networks, they present very different localizations (nervous system, skin, muscular tissue, eye, but very rarely the liver and lung). It seems logical to conclude that a special histotropism accounts for these differences.

It is impossible to be certain how long it will require for cysts to attain the enormous sizes met with—628 gm. in one of our cases (Fig. 9). Experimentally it has been shown that a cyst takes from $3^{14}$ to $16^{23}$ months to grow to 1 cm. in diameter. We suspect, therefore, that cerebral hydatids are at least 3 to 4 years old when they come to the attention of the surgeon.

Hydatid cysts are found most commonly in the parieto-temporo-occipital region. In our 13 cases, 3 involved the frontal lobes, 1 was parietal, and the remainder were temporo-occipito-parietal. In 27 of the 42 cases reported by Schroeder and Medoc, the cysts were found in the temporo-occipito-parietal region. Reggiardo reported an intraventricular cyst but at the Neurological Institute in Montevideo neither Schroeder nor we have had a similar experience. Carrau described a cyst in the sella turcica which he believed to be meningeal. Devé, who examined this specimen later, believed it to be of osseous origin. We shall not deal in this paper with hydatidosis of the cranial bones, which is of rare occurrence.

Microscopically, the hydatid cyst consists of the parasite, a hyaline membrane limited by an adventitia and the germinative membrane, and the reaction of the tissue surrounding the cyst. It should be borne in mind that calcified hydatids have also been observed. The brain is displaced by the cyst, and shows a striking tolerance to it. According to Schroeder and Medoc, an area of demyelinization is formed around the cyst, the axis-cylinders remaining intact for a long time. Macrophages and compound granular corpuscles appear then. The vessels and cells assume an arrangement parallel to the surface of the cyst. A moderate fibrillary and cellular hypertrophy of the micro-, oligodendro- and macroglia takes place about the cyst, both close to the cyst wall and at some distance from it. An adventitia surrounds the cyst wall. It can be perfectly isolated during the removal of the cyst. It is made up of a highly vascular connective tissue.

Schroeder and Medoc believe that the changes occurring in the nervous tissue about the cyst are entirely mechanical in origin.

CLINICAL STUDY

The clinical symptomatology attending cerebral hydatid cysts depends on whether we are dealing with a single cyst or multiple ones. We are particu-
HYDATID CYSTS OF THE BRAIN

particularly interested in the single ones, as multiple cysts are rarely found.

Our clinical study includes both children and adults. Of our 13 patients, 10 were children, 15 years of age or younger, and 3 were adults. In one of the latter the cyst was infected.

1. CEREBRAL HYDATID DISEASE IN CHILDREN

The symptomatology usually includes headache, vomiting, lowering of visual acuity, hemiparesis, somnolence and convulsive fits. The objective signs commonly are impairment of general health, somnolence, Macewen’s sign, papilloedema, optic atrophy, hemianopsia, hemiparesis, signs of cerebellar dysfunction (tremor and hypotonia), astereognosis and stiffness of the neck.

(a) Subjective symptoms:

**Headache** is observed consistently, and was the earliest symptom in 8 out of 10 cases. Localization becomes difficult because of the age of the patients, the more so when somnolence is present. The area of maximum pain corresponds in some cases to the actual location of the cyst. The headache follows the pattern of headaches caused by intracranial hypertension. It may subside together with the intracranial hypertension, when the latter presents a two-stage evolution as so often happens in children.

**Vomiting** was present in 9 of our 10 cases. It often is associated with the headaches and improves when the intracranial hypertension subsides.

**Lowering of visual acuity.** Five children had diminished visual acuity. This symptom may be more frequent but it is sometimes difficult to ascertain in children.

**Hemiparesis.** In 5 cases the children complained that they were unable to move one half of their body. This disturbance was never found to be intense, not even in one case in which it appeared as the initial symptom.

**Somnolence, mental dullness.** In 3 instances the parents complained that the children showed mental dullness and were inclined to sleep a great deal.

**Convulsions.** These were observed in 2 patients. Some writers state that they are rare.

(b) Objective signs:

**General condition.** It was poor in at least 3 of our cases, probably related to the huge size of the cysts. It is usually remarked that cerebral hydatid disease is well tolerated; yet, some writers have observed children whose general health was impaired.

**Macewen’s sign.** This symptom, also called the “cracked-pot” sound, was observed in 3 patients. Sometimes it was found in the area corresponding to the location of the cyst, but in other instances, it was more readily demonstrated in the opposite hemiceranium. We also observed one instance of craniotabes.

**Papilloedema.** It was consistently observed and nearly always was of great intensity.

**Optic atrophy.** Two patients showed definite optic atrophy.
Hemianopsia. The writers observed homonymous contralateral hemianopsia in half of the patients.

Hemiparesis. In agreement with subjective symptoms, 5 patients exhibited a moderate objective hemiparesis of the pyramidal type. In one case Babinski's sign was elicited bilaterally.

Cerebellar syndrome. This was seen in 5 of the 10 children. In 3 it was associated with hemiparesis. Of these latter, 2 presented both hemiparesis and neocerebellar hemisyndrome on the same side; in the third they were contralateral.

Tremor. This is a striking feature. It is usually bilateral and most clearly seen in the upper limbs. It was observed, with varying intensity, in 6 of the patients.

Hypotonia. In 5 cases it was observed either in two or four limbs.

Astereognosis. This was observed in 2 children.

Stiffness of the neck and Kernig's sign. They were observed clearly in 2 patients. One child showed a palsy of the third cranial nerve, surely caused by a temporal pressure cone; another experienced pains in the cervical region radiating toward the upper limbs. Such spinal pains were also reported by Morquio.26

The most important signs observed in our 10 cases may be placed in two groups: those caused by intracranial hypertension, and those of localizing significance.

In children it is not uncommon for the intracranial hypertension to present a two-stage evolution, i.e., after attaining a certain intensity, it improves suddenly and sharply, coinciding with the separation of the cranial sutures. Later the headaches and vomiting return.

Hemianopsia, hemiparesis, astereognosis, and the cerebellar symptoms are of localizing value. The convulsions apparently have only minimal localizing significance. In some cases manifestations that the patient's family refer to as convulsions are in reality generalized, bilateral tremors. These are usually associated with the larger intracranial lesions. Morquio26 pointed out this possible confusion between tremor and convulsion.

Hypotonia and the cerebellar hemisyndrome were encountered both with occipito-parietal and frontal lesions.

A number of symptoms described in the literature on the subject were not observed by the writers. For instance, we did not observe any psychic disturbances.

We do not believe there are any symptoms typical or diagnostic of cerebral hydatid cysts as some workers do.

The signs of MacEwen and the craniotabes, although associated with cerebral hydatid cysts, are a consequence of the great increase of intracranial pressure, so we prefer to deal with them under hypertension symptoms.

2. CEREBRAL HYDATID DISEASE IN ADULTS

We observed 2 adults, 38 and 48 years of age, with cerebral hydatid cysts. The symptomatology corresponded to that of brain tumors. There was
nothing characteristic of this particular condition. In one of these 2 cases there were signs of localized cerebral disease but no papilloedema.

*Suppurating hydatid cyst* (Fig. 1). In our only case of this type symptoms suggestive of a benign cerebral tumor had been developing for 3 years. The patient’s illness began with convulsions. We thought he had a frontal tumor. The only unusual finding was a fever during the 4 days of hospitalization prior to operation, coupled with a 10 per cent eosinophilia in the blood. This was unlike Obrador’s findings with another suppurating hydatid cyst.

**DIAGNOSIS**

Morquio came to the conclusion that 50 per cent of cerebral tumors in children are hydatid cysts. During the last two years we have operated upon 18 brain tumors and 8 cerebral hydatid cysts. The clinical characteristics likely to lead to the positive diagnosis of a cerebral hydatid cyst are: 1) symptoms of increased intracranial pressure; 2) diffuse neurological symptoms, not indicative of an exact localization, but revealing a lesion involving a large portion of one cerebral hemisphere or even of both hemispheres; 3) the patients live in rural areas. We have never found a case in Montevideo. 4) The presence of a hydatid cyst elsewhere in the body. According to Dew 80 per cent of cerebral hydatid cysts are associated with cysts in the liver. But of the 13 intracranial cysts reported here only 2 were associated with pulmonary cysts and no hepatic cysts were observed. Furthermore, the
presence of cysts elsewhere in the body is far from conclusive. Schroeder reported the case of a child with a hydatid cyst in the lung and another in the peritoneum. The child had a glioma of the cerebellar vermis.

The general condition of the patient is often only fair. He commonly seeks consultation when the cyst has undergone several years' evolution and attained a large size, giving rise to somnolence, mental dullness and generalized tremor which hinders the correct utilization of the limbs. Of the 3 adult patients, one exhibited obvious mental deterioration. The good general health shown by some patients does not help to differentiate hydatid cyst from other intracranial tumors. Even patients with malignant or metastatic tumors may be in excellent general health.

Pérez Fontana, Rodríguez and Rodríguez Barrios pointed out the occurrence of headache localized at the site of the cyst a few hours after the performance of the Casoni reaction.

**DIFFERENTIAL DIAGNOSIS**

Since intracranial hypertension is the prevailing manifestation of cerebral hydatid cysts, differential diagnosis is primarily concerned with brain tumors. Tumors of the cerebellum must be given special consideration as they too are more common in children. The erroneous diagnosis of a tumor in the posterior fossa is likely to lead to a serious complication. If ventricular puncture or ventriculography is attempted the hydatid cyst in the parieto-occipito-temporal region is likely to be punctured, scattering the hydatids and giving rise to secondary or daughter cysts. In regions where hydatid disease is a possibility cerebral angiography, not ventriculography, should be used to establish the diagnosis and to differentiate cerebral hydatid cysts from cerebellar tumors. In our 13 cases we generally approached surgical intervention bearing in mind the possibility of cerebral hydatid cysts. Notwithstanding, in one instance we mistook the condition for a posterior fossa tumor and punctured an occipital hydatid cyst while making a ventriculogram.

**SUPPLEMENTARY DIAGNOSTIC PROCEDURES**

*Laboratory Findings.* Eosinophilia in the blood, the Casoni reaction and the Weinberg reaction are not of great value. In our 13 cases eosinophilia was looked for in 10 and found in only 3; the Casoni reaction was carried out in 8 and was weakly positive in 1; the only 2 Weinberg tests which were made were negative.

*Electroencephalography.* According to Fúster, Castells and Gastaut, the typical electroencephalographic findings with a cerebral hydatid cyst are an area of electrical silence surrounded by delta waves. They look upon such a finding as pathognomonic.

*Roentgenography.* Ordinary roentgenograms may yield highly interesting data, above all as regards the thinning of the vault of the skull, as described by Herrera Vegas and Phillips. Carrea considers it the stage preceding
the development of parchment-like diploe. In the rare cases of calcified hydatid cysts, regular roentgenography may be very valuable.

Cerebral Angiography. The development of this technique has greatly facilitated the diagnosis of cerebral hydatid cysts. In a previous paper\textsuperscript{2,5} we dealt with this subject at length. We concluded that there are several angiographic characteristics of cerebral hydatid cyst (Figs. 2, 3, 4 and 5).

![Angiograms showing the displacement of cerebral vessels and the peculiar aspect of the sylvian group.](image1)

![Angiograms showing the displacement of cerebral vessels.](image2)

**Fig. 2.** Angiography showing the enormous displacement of the cerebral vessels and the peculiar aspect of the sylvian group (G.S.).

**Fig. 3.** The vessels of the sylvian group (G.S.) are all crumpled up but even so they keep their peculiarly distended, thick appearance.

1) Complete absence of blood vessels within the lesion.

2) The vessels surrounding the mass are not abnormal as with some intracerebral tumors. They are arranged parallel along the surface of the cyst. Because of the large size of the cyst they are not tortuous and in this respect are similar to those observed in hydrocephalus. The arteries, which Moniz\textsuperscript{24} described as "spider legs," have an even diameter throughout their course.

3) There is an enormous displacement of the vessels. Hydatid cysts attain a volume hardly ever exhibited by tumors, although supratentorial tumors in children may attain a huge size, too.\textsuperscript{7}
4) The circumferential arrangement of the vessels about the spherical mass is strongly diagnostic of a hydatid cyst.

The above characteristics enabled us to arrive at the diagnosis of hydatid cyst in 6 of our 10 cases. In the 4 remaining cases the technique led us to a well-grounded suspicion of the disease.

Cerebral angiography is a particularly ideal diagnostic method in these cases because it does not entail the risk of puncturing the cyst; as the cysts are usually supratentorial it is well adapted to these cases; and in most cases it discloses the nature of the condition as well as localizing it.

Although ventriculography is of localizing value and may even disclose some peculiar features as described by Schroeder it is contra-indicated because of the danger of puncturing the cyst.

**TREATMENT**

The treatment of a cerebral hydatid cyst is surgical. As noted above these cysts are nearly always solitary and when they are removed without ruptur-
HYDATID CYSTS OF THE BRAIN

In Uruguay, the Dowling\textsuperscript{17} or Schroeder\textsuperscript{25} techniques were applied until a few years ago. In our opinion, the former is the more advantageous because it enables one to remove the cyst intact, through the technique described in detail by Viale,\textsuperscript{42} a disciple of Dowling. A large bone flap is reflected, the dura mater is opened, and then cerebral cortical incisions, that Dowling graphically termed episiotomies, are made. Finally, through gentle maneuvers, placing cotton pledgets between the brain and the cyst, the latter is induced to come out spontaneously. We have removed two cysts in this ingenious manner, which fulfills all the conditions of ideal treatment.

Prof. Schroeder\textsuperscript{28} worked out a technique by which the cyst is punctured after thoroughly walling-off the exposed surface. One per cent formalin is then injected into the cyst and left 5 minutes in the cavity. Then the fluid is evacuated and the cystic membrane is easily removed with the aspirator, to which it adheres.

We applied this technique in 3 cases in which the true condition was not recognized early. It is a useful procedure enabling the surgeon to remove the membrane expeditiously whenever the cyst is ruptured during an operation. However, Schroeder himself admitted the hazards of contamination resulting from the puncture of the cyst.

In 1 of the 3 cases in which we used this method, we had suspected a tumor of the posterior fossa and had punctured the cyst during ventriculography. A recurrence resulted which required a second operation 1 year later. Because of this, we consider the techniques reported by da Gama Imaginario,\textsuperscript{19} Dew,\textsuperscript{16} and Philipov\textsuperscript{30} as ill advised.

We propose two techniques for removal of the cyst\textsuperscript{2,4,37}

1. The technique we proposed in 1951 together with Rodríguez Barrios\textsuperscript{3,4} (Fig. 6) may be summed up as follows: 1) diagnosis and localization of the cyst by cerebral angiography;\textsuperscript{2,7} 2) a very broad osteoplastic flap; 3) contralateral ventricular puncture; 4) exposure of the cyst by radiating incisions of the cortex; 5) placing of the head with the cyst dependent so that gravity will aid in removing the cyst; 6) a gentle injection of air into the ventricle of the opposite cerebral hemisphere; 7) gentle irrigation of saline between the cyst and the cerebral substance.

\begin{figure}[h]
\centering
\includegraphics[width=\textwidth]{fig6.png}
\caption{Diagram of technique proposed by Arana, Rodríguez Barrios and San Julián.\textsuperscript{3,4} The cyst is expelled by air injected into the opposite ventricle.}
\end{figure}
2. Observation of the above technique convinced us that the injection of saline solution between the cyst and the brain could not only facilitate the removal of the cyst but provoke it. Accordingly we have managed our 4 most recent cases as follows, with highly satisfactory results\(^\text{37}\) (Figs. 7, 8 and 9).

This technique is comprised of the following stages:
1) Localizing diagnosis by cerebral angiography; 2) a very broad osteo-

![Diagram of technique proposed by San Julián and Arana.\(^\text{37}\) Saline solution is injected between the cyst and the brain.](image)

plastic flap; 3) incisions in the brain substance, only when necessary, and taking care not to make them too extensive but attempting to keep the cavity between the cyst and the brain relatively closed so that the injected saline may collect within this cavity beneath the cyst; 4) placing of the patient with the cyst dependent so as to derive the greatest benefits from gravity; 5) irrigation of saline between the cyst and the brain substance (Fig. 8). Even without the help of gravity the injection of saline solution is highly effective, undoubtedly more so than the injection of air into the
opposite ventricle. Furthermore this technique does not require a contralateral opening for ventricular puncture.

While the previous technique presented the advantage of avoiding any contact with the cyst membrane during surgical maneuvers, in this second method only the saline solution gets in touch with it, thus ensuring the delicacy of the maneuvers, for there is no contact with solid instruments or cotton pledgets. Since the liquid is injected between the cyst and the nervous substance surrounding it, the slight hypertension that develops is less likely to increase intracranial pressure than air injected into the opposite ventricle. Although the number of cases is still low, this latter technique apparently results in a more benign postoperative course than that following the techniques of Dowling or Schroeder, or our first procedure. In the case of very deep cysts, the first procedure may still prove very useful.

Before closing this paper we cannot help recalling the following remarks, uttered by the Argentinian, Cranwell in 1908.

Hydatid cysts are usually operated upon after attaining such a huge size that the brain has been irreparably damaged. Operation in such cases sometimes only hastens the patient's death. Even those patients who survive are often not actually cured. In fact, in many cases, only the immediate danger is averted and somber complications still await them. Hence, with the long term view we conclude that cerebral hydatid disease is a particularly serious, nearly always fatal condition.

Fifty years later, thanks to the cerebral angiography of Moniz and the present-day neurosurgical technique initiated by Harvey Cushing, and the pioneer work of Dowling in Argentina and Schroeder in Uruguay, we may
state that the dismal outlook of Cranwell has given way to the hope of success in every single case.

SUMMARY

On the basis of 13 observed cases of cerebral hydatid cyst, we have discussed their clinical characteristics and emphasized the importance of angiography for the diagnosis of the disease. Current operative methods have been discussed, and we have proposed two techniques involving: 1) injection of air into the contralateral ventricle; and 2) injection of saline solution between brain tissue and the cyst. We favor this latter procedure because of its simplicity and effectiveness.

The above 13 patients were operated upon and all are living at present. In one case the cyst was punctured during ventriculography when we suspected a tumor in the posterior fossa. This patient was re-operated upon one year later. There were three cysts in the bed of the primary cyst.

REFERENCES


HYDATID CYSTS OF THE BRAIN


