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# PULMONARY CYSTIC ECHINOCOCCOSIS IN A CHILD PRESENTING IN THE UNITED KINGDOM WITH FEVER AND CHEST PAIN: A BRIEF REPORT AND DISCUSSION ON MANAGEMENT

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**Abstract:** Cystic echinococcosis is a zoonosis caused by the larvae of *Echinococcus granulosus*. Pulmonary disease may be asymptomatic until the cyst ruptures or becomes secondarily infected. We report a case of pulmonary cystic echinococcosis presenting in the United Kingdom, with discussion on management: optimum antihelminthic agent, length of treatment and type of operative intervention. Treatment should be individualized to the clinical scenario.

**Key Words:** cyst, hydatid, pulmonary

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Hydatid disease, or human echinococcosis results from infection with the larval stage of the tapeworm *Echinococcus,* with *Echinococcus granulosus* causing cystic echinococcosis (CE) and *Echinococcus multilocularis* causing alveolar echinococcosis. Hydatid disease is found most in the Mediterranean region, South America, north/east Africa, southern/central Russia and central Asia). The definitive hosts for *E. granulosus* are dogs, with livestock (eg, sheep and goats) acting as intermediate hosts.[1]

Eggs released into the feces of the definitive host are ingested by the intermediate host, hatching in the small intestine and releasing hooked oncospheres which are able to penetrate the intestinal wall and move through the circulation to end organs, usually liver (65%) and lungs (25%).[1] Once in the end organ, the oncosphere develops into an echinococcal (hydatid) cyst containing many protoscoleces. The definitive host ingests the cyst-filled organs of the intermediate host and becomes infected, with protoscoleces attaching to their intestinal mucosa and maturing into adult worms in 32–80 days.[1] Humans are an aberrant intermediate host, with infection occurring after ingestion of eggs via the fecal-oral route.

## CASE DESCRIPTION

A previously healthy 12-year-old boy presented to his local hospital with 24 hours history of fever and worsening right-sided lower chest pain. His oxygen saturations were 92% on room air, with tachypnoea of 36 breaths per minute at rest. His initial white cell count was $10.7 \times 10^9$/L; with neutrophils $9 \times 10^9$/L, lymphocytes $0.6 \times 10^9$/L, eosinophils $0.38 \times 10^9$/L and C-reactive protein (CRP) 68 mg/L. Chest radiograph (CXR) (Fig. 1A) findings were suggestive of a lung abscess. He was commenced on intravenous ceftriaxone and oral clindamycin to cover likely local pathogens for primary lung abscess (*Streptococcus aureus*, *Streptococcus pneumoniae* and Group A Streptococcus*)*. By day 5 of admission, the child remained febrile. Additional history included that the child had moved to the United Kingdom from Botswana at 7 years of age. He had lived in a rural area of Botswana and had helped to look after goats at his family's property.

Further investigations demonstrated CRP 270 mg/L, Erythrocyte sedimentation rate 117 mm/h, and alanine transaminase 143 IU/L. Hepatitis B, C and HIV serologies and interferon-gamma release assay (QuantiFERON-TB Gold, Quiagen, Germantown, MD) were negative. Computed tomography scan of the thorax (Fig. 1B) demonstrated a cystic/cavitating lesion in the right lower lobe with an associated area of nodularity of the pleura at the base of the lesion. There were calcifications noted within the cyst fluid, suggestive of a nonacute lesion. There was no marked hilar lymphadenopathy. Abdominal ultrasound did not demonstrate any other cysts.

Primary differentials included a congenital cystic adenomatoid malformation or CE (cystic hydatid disease) with superadded bacterial infection, or pulmonary amoebiasis. Amoebic and hydatid serologies were sent and metronidazole was added to cover for amoebic disease.

## MEDICAL AND SURGICAL MANAGEMENT

Hydatid serology was positive (*Echinococcus* IgG enzyme-linked immunosorbent assay (ELISA) positive at optical density of 0.517). This was confirmed with species-specific immunoblot testing, which was positive for *E. granulosus*. Albendazole (15 mg/kg/day) and praziquantel (40 mg/kg/day) were commenced, and plans were made for surgical removal of the lesion. Prednisolone and chlorphenamine were given respectively at 24 and 12 hours preoperatively to reduce the risk of anaphylaxis, which can occur with operative spillage of hydatid contents.[2]

Right lower lobectomy was completed 3 weeks after the initial presentation, allowing the child to have several days' treatment with praziquantel preoperatively. Intraoperatively, significant adhesions and inflammatory changes of the pleura were noted, and cyst contents were described as "gelatinous/caseous" material. Macroscopic appearance of the excised lobe is shown in Figure 1C. Histology demonstrated markedly thickened pleura and laminated eosinophilic retractile layer of *Echinococcus* in varying stages of development (Fig. 1D).

The child was well postoperatively and defervesced. He received 2 further weeks of praziquantel and 3 months of albendazole. He had full blood count and liver function test monitoring every 2 weeks while on albendazole, with no neutropenia or



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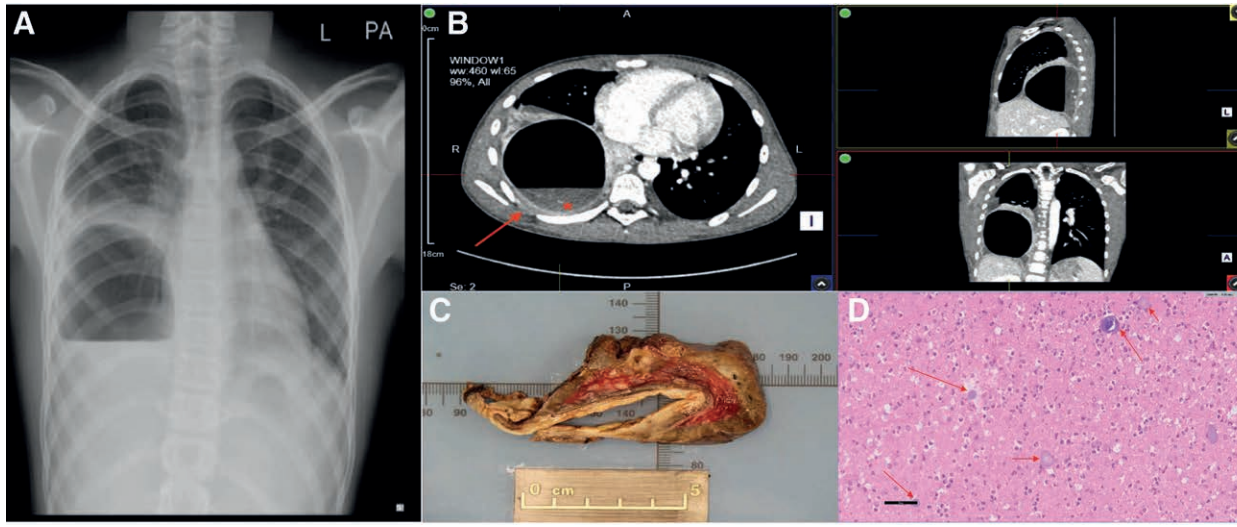


**FIGURE 1.** Radiology and histopathology findings. A: Plain CXR demonstrating large right-sided pulmonary cyst containing an air-fluid level. B: CT thorax with multi-angle views demonstrating cystic/cavitating lesion of right lower lobe containing dependent fluid. The lesion is thin-walled, with an associated area of nodularity of the pleura (arrow). Calcifications are visible within the fluid (asterisk) and there is no marked hilar lymphadenopathy. C: Macroscopic appearance of resected lobe of lung. D: Hematoxylin and eosin-stained section demonstrating laminated layers of *Echinococcus* at varying stages of development (arrows). CT, computed tomography; CXR, chest radiograph.

disturbance of his liver enzymes. Postoperative CXR showed good expansion of the right lung and almost complete resolution of the pleural thickening. One year postoperatively, the child remained well and had been able to resume playing football.

## DISCUSSION

### Diagnosis of Cystic Echinococcosis

Pulmonary CE appears to be more frequent in children than in adults, most commonly presenting with chest pain, cough and/or dyspnea, with hemoptysis or urticarial/anaphylactic symptoms if rupture into the bronchial tree occurs.[3] Spillage of cyst material into the bronchial tree or pleural cavity can cause pneumothorax, pleural effusion, empyema or pulmonary abscess with secondary infection.[3] The growth of the cyst is slow enough (around 1–5 cm/year)1 that most children and adolescents with lung lesions remain asymptomatic despite large lesions, which may be picked up incidentally on CXR.

A diagnosis of hydatid disease would be supported by eosinophilia, associated with leakage of cyst contents, but this is only present in up to 15% of cases.[3] Serologic tests are the mainstay of diagnosis in the absence of histopathology and are also useful for monitoring posttreatment. A positive screening test can be followed by immunoblot testing using species-specific antigens to confirm a diagnosis of CE.[4] Serology is more likely to be positive in liver disease (with 80%–90% sensitivity reported for IgG ELISA) than lung disease (60%–85% sensitivity for IgG ELISA),[4] therefore negative serology does not necessarily rule out disease.

## IMAGING IN CYSTIC ECHINOCOCCOSIS

Pulmonary CE is likely to be unilateral in children, occurring most frequently in the right lower lobe, as in this case.[5] Uncomplicated cysts are classically described as being well-circumscribed, fluid-filled lesions with homogenous content and "hyperdense" walls and calcifications are noted to be rare.[6] Cysts that have ruptured into the bronchial tree can develop an air-fluid level or "Cumbo's sign." Superinfected cysts may also have the appearance of an air-fluid level,[6] and 1 retrospective pediatric study noted that CXR findings of children with pulmonary echinococcosis appeared very similar to those of a lung abscess in 65% of cases.[5]

## MANAGEMENT OF CYSTIC ECHINOCOCCOSIS

Given the wide range of presentations with pulmonary CE, there is no uniform treatment recommendation, rather the location, size and stage of the cyst and available resources dictate disease management.

Albendazole acts by inhibiting the assembly of microtubules, preventing the germinal layer of the cyst from absorbing glucose. Some studies have shown efficacy of albendazole alone on small and uncomplicated cysts, including 1 solely pediatric study, but for more complicated lesions (eg, larger or infected cysts, or lesions causing compression of the parenchyma),[5] a conservative surgical approach has been recommended with adjunctive albendazole treatment. This latter approach was supported by a systematic review and meta-analysis, which noted greater treatment success with the combination of surgery and albendazole over albendazole alone.[7]

Praziquantel is thought to have both protoscolecidal action and reduce viability of early cysts, with suggestion that preoperative treatment may reduce the risk of recurrence and seeding with spillage of cyst contents. This was supported by a study of 22 patients with pulmonary CE; in the group treated with praziquantel for 5–6 days preoperatively, 43%–64% protoscoleces were nonviable at surgery, versus 9% in untreated patients.[8]

A number of pharmacological studies also noted increased bioavailability of albendazole when co-administered with praziquantel, and a small safety study recorded only mild, infrequent and reversible side effects such as nausea and loose stools. There are no randomized controlled trials of longer-term praziquantel treatment in cases not amenable to surgery, or in severe disease.

Recurrence is one of the major complications of hydatid cysts, with reported rates of postoperative recurrence as high as


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18% in some studies, postulated to be because of spillage of cyst contents and dissemination, or additional small cysts being missed intraoperatively.[3] Conservative surgical methods, such as cystotomy and capitonnage (emptying the cyst and closing its cavity with sutures) have been recommended where possible, as they conserve the maximum amount of lung parenchyma, with reduced morbidity, shorter hospital stay and low risk of recurrence. The largest case series of 643 children who had surgical treatment of lung hydatid demonstrated good outcomes with 3% postoperative morbidity and 0.5% mortality.[9]

There is debate about the optimum length of perioperative therapy; an extensive review of data on albendazole use from multiple sources suggested that initial treatment should be for 3 months, with some consideration of continuation beyond this being balanced with the possibility of side effects.[10] The most common side effects of albendazole treatment are reversible hepatotoxicity, cytopenias and alopecia, hence the need for regular full blood count and liver function monitoring while on treatment. Timing of relapse in CE is very variable and has been reported between 3 months and 20 years posttreatment,[1] therefore patients are followed up for 10 years with repeated imaging and serology to check for signs of recurrence.

## SUMMARY

This case demonstrates the importance of exposure history in a child with fever and pulmonary cyst; this brought hydatid disease into the differential diagnosis, allowing preoperative antiparasitic therapy to be initiated. The review of the literature demonstrates some of the issues and uncertainties surrounding diagnosis and treatment of echinococcal disease, the lack of randomized controlled trials on treatment and the need to individualize management to the clinical scenario.

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# *MYCOBACTERIUM CANETTII* TUBERCULOSIS LYMPHADENOPATHY IN A 3-YEAR-OLD CHILD

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**Abstract:** A 3-year-old male originating from Djibouti presented with a cervical mass evolving for 2 months. Tuberculous lymphadenopathy was suspected based on biopsy results, and he improved quickly on standard antituberculous quadritherapy. Subsequently some features of the mycobacterium that grew in culture were unusual. The isolate was eventually identified as *Mycobacterium canettii*, a peculiar species of the *Mycobacterium tuberculosis* complex.

**Key Words:** *Mycobacterium canettii*, tuberculosis, lymphadenopathy

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Cervical lymphadenopathy is a frequent cause for consultation in pediatrics. Although most are benign self-limited and reactive, it can be an early sign of malignancy, immunodeficiency or severe infection. Here we report a case of a tuberculous lymphadenopathy due to *Mycobacterium canettii* and propose a brief review on the specific features of this very rare pathogen.

## Case Presentation

A 3-year-old male presented to the emergency department with a right cervical swelling that enlarged over 2 months. He had no significant medical nor surgical history, and he had up to date vaccinations, including the Bacille Calmette-Guerin vaccine. He did not travel outside Djibouti, where he lived with his parents and his older brother who were in good health. His family had no pet. In August 2021, his parents consulted a pediatrician in Djibouti for a cervical mass that appeared 1 month before and was increasing in size since. The child was generally unwell with weight loss, poor oral intake, and night sweats, prompting his referral to a French hospital for biopsy to rule out a malignancy.

He was admitted in a French tertiary referral center in September 2021 and was found to have a large noninflammatory cervical swelling. The rest of his clinical examination was normal, without respiratory symptoms, other adenopathy or hepatosplenomegaly. Initial work-up included a complete blood count with a white blood cell count of $15.19 \times 10^9$/L and an absolute neutrophil count of $6.27 \times 10^9$/L, hemoglobin was at 128 g/L. The c reactive protein was at 7 mg/L (normal level: <5 mg/L), lactate dehydrogenase was normal at 317 U/L as was the uric acid (236 µmol/L). A computed tomography (CT) scan confirmed the

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