Ruptured Hydatid Cyst of the Liver with Biliary Obstruction: Presentation of a Case and Review of the Literature

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Abstract

The case of a 66 year old woman admitted with a picture of jaundice acute cholangitis is reported. Ultrasonography showed a dilatation of intrahepatic bile ducts, gallbladder hydrops with several stones, enlarged common bile duct (CBD) with hyperechoic material inside and a cystic tumor with hydatid features. With a strong suspicion of a hydatid cyst ruptured in the biliary tree with biliary obstruction, endoscopic cholangiopancreatography was performed. The diagnosis was confirmed by endoscopic retrograde cholangiopancreatography and the hydatid membranes were extracted from the CBD with subsequent clinical improvement. The second step of treatment comprised the surgical cure of the cyst and cholecystectomy. The data from the literature are finally presented with a special emphasis on the ultrasound diagnosis and the endoscopic treatment.

Key words

Biliary obstruction – hydatid liver cyst - ultrasonography– endoscopic treatment

Rezumat

Se prezintă observația clinică a unei paciente de 66 ani care s-a internat cu tabloul unui icter cu fenomene de angiocolită acută. Ecografia a evidențiat dilatarea căilor biliare intrahepatice, hidrops vezicular cu mai mulți calculi , dilatarea canalului coledoc cu imagini ecogene în interior şi o leziune chistică hepatică cu caractere de chist hidatic. Suspicionându-se o obstrucție biliară prin fragmente hidatice, s-a efectuat o colangiografie retrogradă endoscopică care a confirmat diagnosticul şi care a permis extragerea fragmentelor hidatice din coledoc cu ameliorarea tabloului clinico-biologic. În al doilea timp s-a efectuat cura chirurgicală a chistului şi colecistectomia . Se prezintă în continuare datele din literatură referitoare la chistul hidatic rupt în căile biliare, insistându-se asupra diagnosticului ecografic şi a tratamentului endoscopic.

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Introduction

One of the most common complications of hepatic hydatid cysts is the rupture of the cyst caused by raised pressure within it. The rupture in the bile ducts may cause jaundice due to biliary obstruction. Abdominal ultrasound (US) and other imaging modalities are able to diagnose the complications. Endoscopic cholangiopancreatography (ERCP) is used extensively nowadays both in the diagnosis and treatment of biliary complications of hepatic hydatid cysts (1-4). We report a case of obstructive jaundice caused by a ruptured hydatid cyst focusing on the US diagnosis and on various treatment modalities.

Case report

A 66 year old female patient was admitted as an emergency to the 3rd Medical Clinic for moderate epigastric pain irradiating to the right upper abdominal quadrant, jaundice, chills and fever (38°C). In addition, the patient presented nausea and vomiting.

The patient had no previous disease, did not drink alcohol and her family history was not significant.

A week before admission she had had some episodes of epigastric pain with nausea and vomiting, and mild fever. At US examination, cholelithiasis was diagnosed. She was treated with antibiotics, without an improvement in the clinical picture.

Physical examination at admission showed a patient with good nutritional status (60 kg weight at 160 cm height), mild jaundice and a tender hepatomegaly.

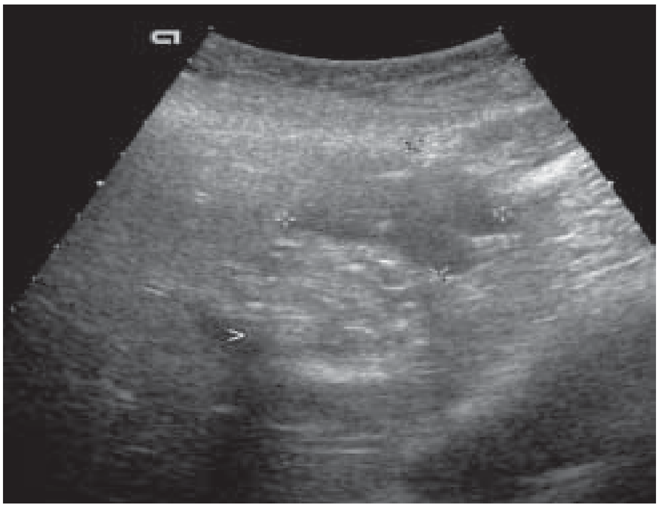

Fig.1 Hepatic US showing a 47/32 policyclic cystic lesion in the III segment adjacent to an echogenic lesion (>).

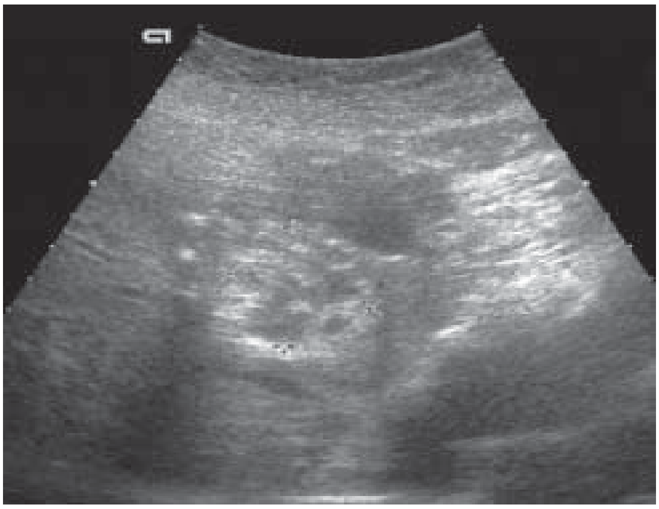

Fig.2 Detailed view of the echogenic lesion showing echogenic wall and cystic areas inside.

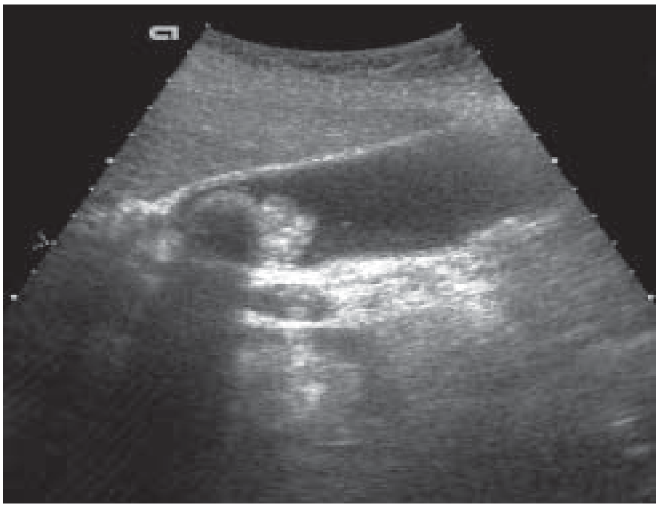

Fig.3 Gallbladder with stones and echogenic non shadowing material on US.

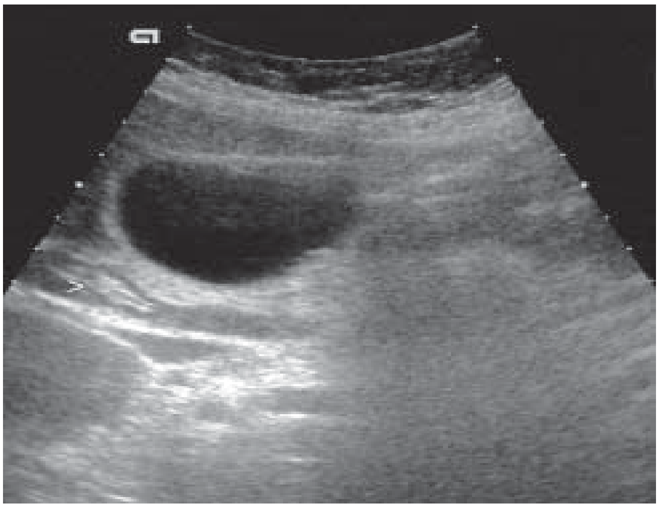

Fig.4 Linear echogenic images in the CBD (>) on US.

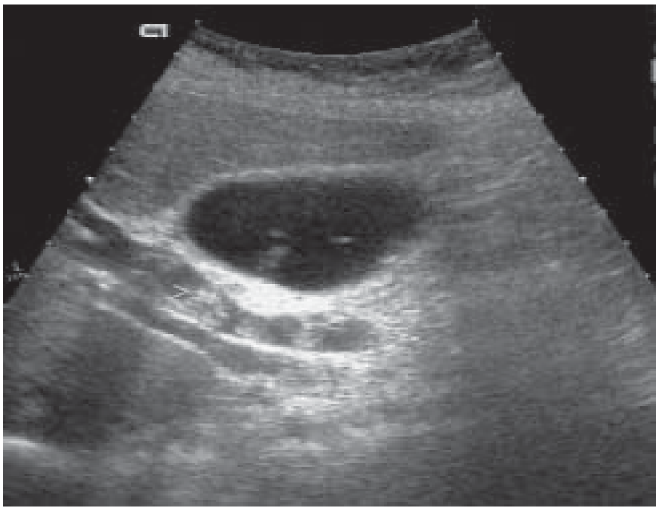

Fig.5 Intrabiliary masses with no acoustic shadow (>).

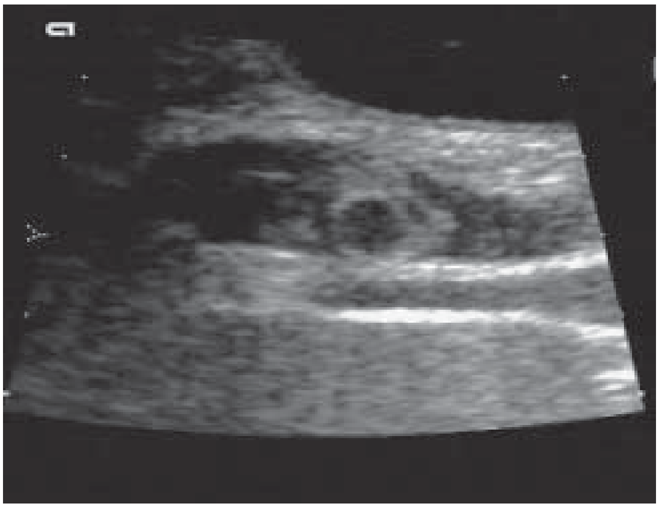

Fig.6 Intrabiliary masses with cystic areas inside.

Laboratory findings evidenced increased serum aspartate (76 U/L) and alanine aminotransferase levels (149 U/L)(normal up to 50UI/L), elevated bilirubin (3.5 mg/dl), alkaline phosphatase (587 U/L) and γ glutamyl trans-

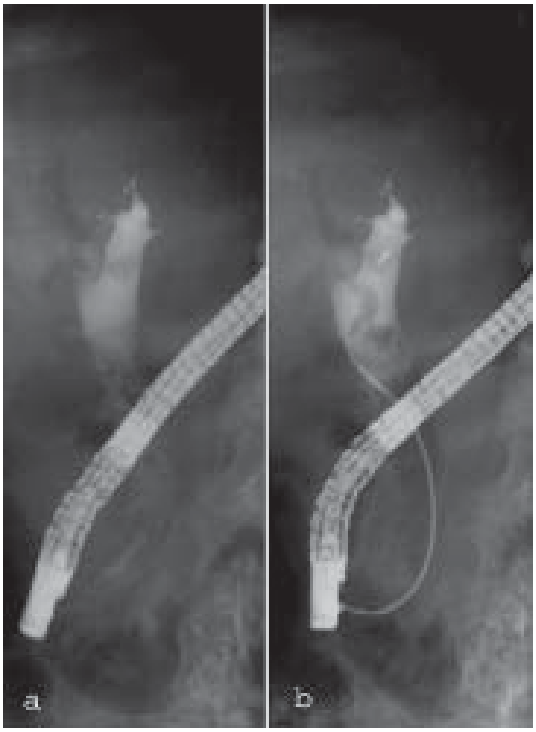

Fig.7a,b Dilated CBD with filling defects of varying size and shapes on ERCP.

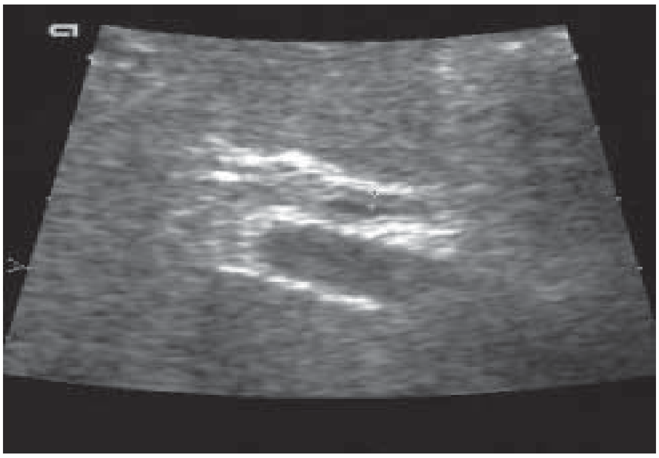

Fig.8 US 2 days after endoscopic removal of hydatid membranes: CBD of normal size with thick, double layered walls.

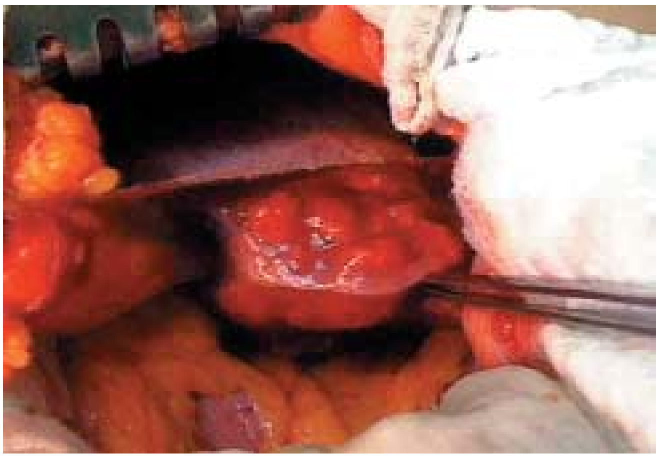

Fig.9 Surgical exploration revealed a hepatic hydatid cyst in the III-IV and II segment.

Fig.10 Surgical opening of the cyst.

Fig.11 Daughter cysts and hydatid membranes from the hydatic cyst.

Fig.12 Gallbladder lumen after surgical removal and opening : stones and hydatid membranes.

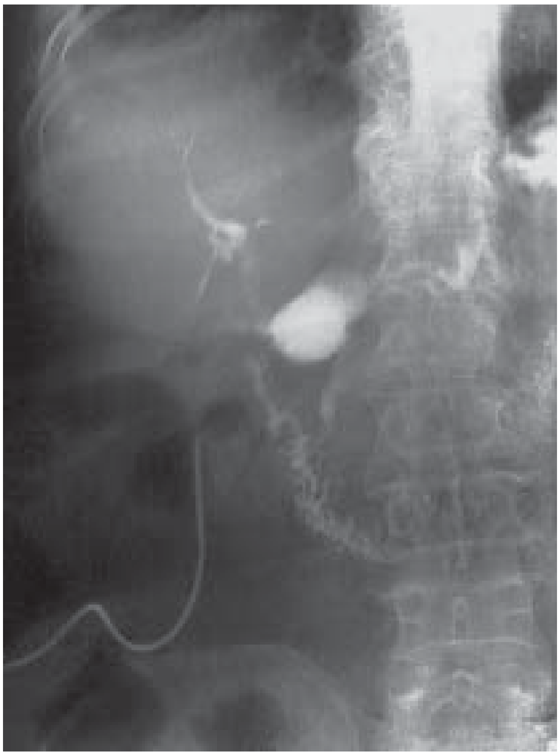

Fig.13 T- tube cholangiography: normal CBD with no strictures or filling defects.

peptidase (592 U/L) and ESR 33-70 mm/h. There was a slight eosinophilia (8%) but a normal leukocyte count. All the other examinations including hemoglobin, urea, blood glucose, coagulation tests, calcium, triglyceride level, were normal.

Abdominal US revealed moderate hepatomegaly with slightly dilated intrahepatic bile ducts, and two adjacent hepatic lesions in the II and III segment: one of cystic type (47/36mm) with thick echogenic walls and the other (33/33mm in size) with echogenic walls and echogenic irregular areas in interior (Figs.1,2). The gallbladder was enlarged, containing stones and echogenic material (Fig.3)

The common bile duct (CBD) was dilated (10.6 mm in its middle part and 13 mm distally) and contained echogenic material inside without an acoustic shadow.. The material had various shapes and aspects (linear - Fig 4, mass-like with and without cystic images inside - Figs.5,6). The pancreas, spleen and kidneys were normal.

The case was interpreted as a ruptured hydatid cyst with biliary obstruction, acute cholangitis and choledocholithiasis. In order to certify the diagnosis and to remove the material from the CBD, an ERCP was performed. ERCP revealed a dilated CBD with filling defects of varying size and shapes (Fig.7a,b) and after a sphincterotomy (ES), hydatid fragments (membranes) were removed. The cystobiliary fistula was not observed.

After ES and removal of the hydatid membranes an improvement in both clinical and laboratory signs (decrease of bilirubin to 0.76mg/dl, alkaline phosphatase 355 UI/L and γ glutamyl transpeptidase 261 UI/L and normalization of transaminases) were noted. An US control showed a normal CBD (6.6mm), with thick walls (Fig.8) but without echogenic material inside. The gallbladder size returned to normal, with walls attached to the stones and no visible lumen .

One week after the ES and removal of the hydatid material a surgical exploration revealed a hepatic hydatid cyst in the III-IV and II segment (Fig.9,10) with corticalisation on both anterior and posterior surface of the liver, a strong inflammation around the CBD and cholecystolithiasis. Partial cystectomy with double drainage and removal of the daughter cysts and hydatid membranes (Fig.11), CBD exploration with T tube drainage and cholecystectomy were performed. The cystobiliary fistula was not found. Inside the gallbladder, stones and dark fragments resembling those found in the hidatid cyst were seen (Fig.12). After the surgical intervention the patient was free of symptoms, with only slightly increased cholestasis enzymes. A T- tube cholangiography showed a normal CBD with no strictures or hydatid membranes inside (Fig.13). There were no complications and after one week the patient was discharged.

Discussion

The hydatid cyst is a parasitic disease mainly caused by Echinococcus granulosum. Hydatid cysts are common in the Mediterranean countries, the Middle and Far East, Europe, Asia, South America and Australia (1-3). In humans, the most commonly infested organs are the liver and lung. Most patients have a solitary cyst, localized in about two thirds of cases in the liver.

A nonruptured hydatid cyst is surrounded by the pericyst, a layer derived from compressed host tissue and chronic inflammatory cells (5,6). The thickness and strength of the pericyst depends on the host organ, in the liver being well defined and tough (5). Blood vessels do not pass through the pericyst but bronchioles and biliary ducts usually do. The true cyst wall has two layers, the ectocyst and the endocyst; both structures are usually referred to collectively as the endocysts. The endocyst is fragile, being protected from the rupture by the tougher pericyst.

An enlarging cyst may compress the surrounding parenchyma causing atrophy and fibrosis (4). An increase in pressure inside the cyst precipitated by trauma, effort or violent cough will cause rupture (2,4). The rupture may be classified as: *contained rupture* when the endocyst is torn but the cyst content is confined within the pericyst; *communicating rupture* consists of a tear of the endocyst with loss of the cyst content via small biliary ducts and *direct rupture* when a tear of both endocyst and pericyst occurs, allowing the cyst content to spill into the peritoneal or pleural spaces (5).

Compression and displacement of the biliary ducts are frequent. At the point of contact with a biliary duct, a rupture may occur. The communicating intrabiliary rupture may be: occult (seen in 10-37% of patients) or frank (3-17%)(4,5,7,8). The occult rupture is usually silent and may be accompanied by suppuration or can evolve towards a frank rupture.

In the frank rupture daughter vesicles and fragmented membranes escape into the biliary tree causing obstruction, cholangitis or septicemia (5). Moreover, acute pancreatitis and acute cholecystitis caused by hydatid material were described in the literature (5, 9,10).

Before the introduction of US or computed tomography, preoperative diagnosis of the complications of hepatic hydatidosis was difficult and was based on clinical manifestations and results of laboratory studies. US may suggest the diagnosis of a frank intrabiliary rupture in most of the cases (5). There are several ultrasound criteria that allow the diagnosis of a intrabiliary rupture of a hydatic cyst (2,5,6,11,12).

Complicated hydatid cyst of the liver. The ruptured cyst is often multivesicular, has a polycyclic shape, septa or irregular echogenic areas in it. In our case, the cyst was composed of a polycyclic echofree part and an echogenic part with small cystic areas inside (daughter vesicles).

Communication. A loss of continuity of the cyst wall adjacent to the bile duct representing the site of communication is a pathognomonic sign of cyst perforation in the biliary tree, but it is very rarely seen, as was in our case.

Dilatation of the biliary tree allows the diagnosis of an obstructive jaundice.

Echogenic material inside the CBD. The presence of small cystic images in CBD which may represent daughter cysts is a very specific sign of rupture, but not very sensitive. Linear hiperechoic images may be found in the CBD and are the expression of hydatid membranes. Nonshadowing material in the CBD is more often seen and is most probably the expression of hydatid sand. However, this sign is not very specific being present in case of biliary sludge, blood clot or inflammatory sludge.

Thickened, double layered bile ducts wall as an expression of cholangitis was present in our case but seen only after endoscopic removal of hydatid fragments. All these sonographic signs were evidenced in our patient, thus making the diagnosis very probable. In a series of 15 cases the sensitivity of US for the diagnosis of intrabiliary rupture of a hydatid cyst was 66.7% (4).

Magnetic resonance cholangiography may show a characteristic intense rim, daughter cysts and detachment of the membranes (4,13).

ERCP provides more comprehensive information in patients with biliary hydatid disease. Daughter cysts may be seen in the duodenum, impacted in the ampulla of Vater or obstructing any part of the biliary tree. On ERCP, the intrabiliary parasitic material appears as non-homogeneous, irregularly shaped and mobile filling defects. A communication between the cyst cavity and the biliary tree can be demonstrated. Other findings at ERCP are displacement and distortion of intrahepatic bile ducts by the hepatic cysts, intrahepatic duct stricture and a mild dilatation of the pancreatic duct. ERCP also helps to differentiate cholestatic jaundice due to ruptured hydatid liver cyst from other causes of obstruction such as choledocholithiasis and carcinoma (14-17). In our case ERCP revealed the intrabiliary material but failed to demonstrate the cystobiliary fistula.

ERCP is very useful in the diagnosis of biliary complications after surgery. The ERCP findings are: external biliary fistula, hydatid material within the bile duct, bile duct stenosis, cystobiliary fistula and hydatid membranes in the gallbladder, extrinsic compression to bile ducts, and a normal cholangiogram (15).

The major role of ERCP is a therapeutic one. Endoscopic sphincterotomy has proved to be an alternative treatment for patients with biliary hydatid disease. In 1985, Al Karawi and Hanid published the first report on the use of ERCP with endoscopic sphincterotomy and extraction of retained echinococcal daughter cysts from the CBD. The success rate of endoscopic treatment is 90-100% in patients without a history of surgery. In patients who have previously undergone surgery for hepatic hydatid disease the endoscopic treatment modalities are as follows: endoscopic sphincterotomy (ES) and nasobiliary drainage in patients with biliary fistula; balloon and or bougie dilation and stenting in patients with biliary stricture; and ES and balloon extraction in patients with residual hydatid material within the bile duct (15,16). In such cases the overall success rate is 70-86% with a rate of fistula closure of 81 % in 10 to 20 days (15,16). In our patient ES and removal of the hydatid remnants resulted in the disappearance of the clinical picture and an improvement in biochemical tests.

In case of cystobiliary fistula a surgical cure of the cyst is mandatory. The most common types of cure are: partial cystectomy with primary closure, partial cystectomy with drainage, cystotomy with drainage, hepatic resection (atypic, segmentary or lobar) and omentoplasty. Suture of the cystobiliary fistula, if visible common bile duct exploration using intraoperative cholangiography are also required. If the biliary tract is cleaned of all cystic content, T-tube drainage should be sufficient. In cases with Oddi stenosis a transduodenal sphincterotomy or choledochoduodenostomy should be considered (18-20). In our patient a partial cystectomy with double drainage and removal of the daughter cysts and hydatid membranes, a CBD exploration with T-tube drainage and cholecystectomy were performed but the cystobiliary fistula was not identified.

It has been suggested that presurgical use of albendazole or mebendazole in echinococcus infestations facilitates surgery by reducing intracystic pressure and by reducing the risk of recurrence after surgery or endoscopic evacuation of a ruptured cyst into the biliary tree (21).

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