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CLINICAL CASE

Choledochal cyst during pregnancy. Report of 3 cases and a literature review☆

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KEYWORDS
Choledochal cyst;
Pregnancy;
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Abstract
Background: Choledochal cysts are rare. They usually present during childhood in women, but it can also be seen during pregnancy. Clinical signs and symptoms are obscured during this time, thus it can complicate the diagnosis and represent a life threatening complication for both the mother and the child.
Objective: To communicate the case of 3 pregnant patients with choledochal cyst.
Clinical cases: Three pregnant women in which choledochal cyst were diagnosed. Two developed signs of cholangitis. The *first* one underwent a hepatic-jejunostomy, but had an abortion and died on postoperative day 10. The *second* one had a preterm caesarean operation due to foetal distress and underwent a hepatic-jejunostomy 4 weeks later; during her recovery she had a gastric perforation and died of septic complications. The *third* one did not develop cholangitis or jaundice. She had an uneventful pregnancy and had a hepatic-jejunostomy 4 weeks later with good results.
Conclusions: Management of choledochal cysts during pregnancy is related to the presence of cholangitis. When they do not respond to medical treatment, decompression of the biliary tree is indicated. Definitive treatment should be performed after resolution of the pregnancy.

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PALABRAS CLAVE
Quiste de colédoco;
Embarazo;
Complicaciones quirúrgicas

Quiste de colédoco y embarazo. Reporte de 3 casos y revisión de la bibliografía

Resumen
Antecedentes: Los quistes de colédoco son una enfermedad rara que afecta principalmente a mujeres en la infancia, pero que pueden presentarse durante el embarazo. Sus síntomas y signos están opacados por los cambios en esta fase, por lo que el diagnóstico puede ser tardío y sus complicaciones poner en peligro tanto a la madre como al producto.
Objetivo: Presentar 3 casos de quiste de colédoco en embarazadas, tratadas en un hospital de tercer nivel.
Casos clínicos: Presentamos 3 pacientes con diagnóstico de quiste de colédoco durante el embarazo. Dos desarrollaron datos de colangitis; la *primera* fue intervenida quirúrgicamente con anastomosis hepaticoyeyunal, pero abortó y falleció al décimo día del postoperatorio. La *segunda* tuvo una cesárea pretérmino por sufrimiento fetal; 4 semanas después fue intervenida con anastomosis hepaticoyeyunal, pero tuvo una perforación gástrica y falleció por complicaciones sépticas. La *tercera* no desarrolló colangitis, tuvo un embarazo sin complicaciones, se le realizó anastomosis hepaticoyeyunal 4 semanas después, con buenos resultados.
Conclusiones: El tratamiento de los quistes de colédoco durante el embarazo está relacionado con la presencia de colangitis. Cuando no responden al tratamiento médico, la descompresión de la vía biliar está indicada. El tratamiento definitivo debe realizarse una vez resuelto el embarazo.

Background

According to Angel and Nassar, et al. choledochal cysts were described by Vater in 1723.[1,2] Their aetiology is unknown, based on that published by Wu[3] and Gmijović et al.[4] in 1936 Yotsuyanagi posed the theory that they arise from inequality in the vacuolisation of the biliary tract in early embryonic life. Currently there are 2 theories; the *first* postulates that the distension or cyst forms secondary to congenital stenosis of the biliary tract; the *second*, proposed by Babbitt,[3,5] in 1969, is that they are the result of abnormal union between the bile duct and the pancreatic duct (greater than 15 mm), with pancreatic reflux, increasing the intraluminal pressure of the bile duct and its distension. The latter theory is the most widely accepted.

Martínez-Ordaz and Niño-Solís[6] contend that in 1959, Alonso-Lej proposed a classification that was modified by Todani in 1977. The symptoms are non-specific, principally jaundice without apparent cause during infancy and vague abdominal pain, and are even more evident in pregnant women. Pregnancy significantly alters the gallbladder, due to hormonal changes of oestrogen and progesterone levels. Oestrogens have been demonstrated to inhibit the motility of Oddi's sphincter. The gravid uterus can also accentuate or aggravate symptoms due to distal obstruction of the bile duct by compression.[2]

Objective

To present 3 cases of choledochal cysts in pregnant women, treated in a third level hospital and to compare them with cases reported in medical literature.

Clinical cases

Case 1

A 22 year-old primiparous woman, with an interuterine pregnancy of 9 weeks' gestation. She started to suffer abdominal pain that was treated with analgesics and antispasmodics. Then she experienced nausea and later vomiting, jaundice and choluria. On physical examination she was found to be conscious, cooperative and calm, adequately hydrated, with jaundiced skin and mucosa + cardiopulmonary examination showed no compromise; there was a palpable tumour in the hypochondrium and right flank of her abdomen, which was firm, mobile, painful, with no signs of peritoneal irritation. The laboratory results showed: direct bilirubin 3.54 mg/dl, indirect 0.22 mg/dl, total 3.76 mg/dl; with raised transaminases (alanine aminotransferase172 U/l, aspartate aminotransferase 89 U/l). An ultrasound was performed which reported an abdominal tumour dependent on the liver. There were no alterations obstetrically. Magnetic cholangioresonance reported a cystic lesion of 20 cm in cephalocaudal and 11 cm anteroposterior diameter, homogeneous, dependent on the extrahepatic bile duct (Fig. 1). The patient started with signs of cholangitis, which did not improve with medical treatment, and therefore underwent giant choledochal cyst resection, cholecystectomy and hepaticojejunal anastomosis. She was reoperated twice for a haemoperitoneum due to haemorrhage from the surgical site. As this developed absence of foetal heart rate presented, and therefore uterine curettage was performed. The outcome was poor, and the patient died due to multiple organ failure on the tenth day of the post-operative period.

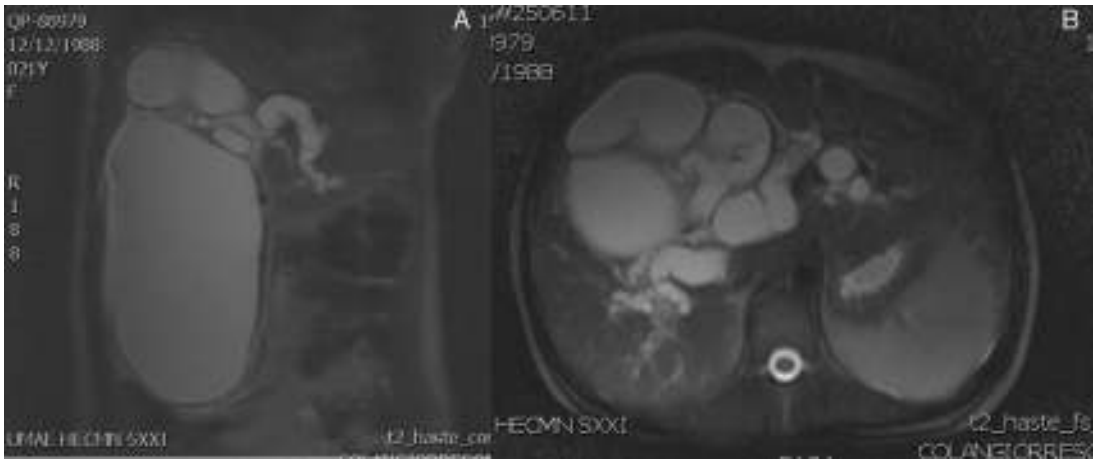

Figure 1 Coronal (A) and axial (B) section of nuclear magnetic cholangio resonance of a pregnant patient with choledochal cyst.

Case 2

A 31 year-old woman, with an interuterine pregnancy of 22 weeks' gestation. She had a history of one normally progressing pregnancy, with an uncomplicated labour 8 years before her current condition. A 3 week history of jaundice, choluria, acholia, colicky transflictive abdominal pain located in the epigastrium, increasing in intensity to 10/10, diaphoresis and nausea. On physical examination the patient was found to be conscious, cooperative and calm, jaundice +++, soft abdomen, rounded due to adipose panniculus and gravid uterus, with mild pain on deep palpation, with no signs of peritoneal irritation. Laboratory tests showed: albumin 2.5 g/dl, total bilirubin 4 mg/dl (indirect bilirubin 0.54 mg/dl, direct bilirubin 3.46 mg/dl) and haemoglobin 10.7 g/dl. The ultrasound reported a live singleton of 22.5 weeks' gestation from the femoral length, hydrocephaly, intrabdominal calcification indicative of foetus-in-fetu, amniotic fluid echoes, which is evidence of bleeding, placenta maturity grade II, cephalic presentation. Magnetic cholangioresonance reported dilatation of the intrahepatic biliary duct and the presence of a voluminous Todani I choledochal cyst compressing the adjacent structures (Fig. 2). An elective caesarean section was performed due to chronic foetal distress, and a live girl was delivered at 26 weeks by Capurro. Because the patient's hyperbilirubinaemia (direct bilirubin 25 mg/dl) and signs of cholagitis, 4 weeks after the caesarean, an elective hepaticojejunal Roux en Y anastomosis was formed finding a findings of choledochal cyst 25 cm × 12 cm, with firm adherences to the portal vein. The patient progressed satisfactorily over the 8 weeks post surgery. However, she presented haemorrhage of the upper digestive tract with a drop in haemoglobin and haemodynamic deterioration, and therefore an exploratory laparotomy was performed and a haemoperitoneum of 1300 cc was found with the presence of ischaemic perforation in the gastric fundus. The outcome was poor; the patient presented abdominal sepsis, septic shock and disseminated intravascular coagulation, and died on the 21st postoperative day.

Case 3

A 22 year-old primiparous patient, with no relevant history. At 20 weeks' gestation she started with signs of mild abdominal pain in the right hypochondrium in relation to cholecystokinetics. Laboratory tests were performed which reported aspartate aminotransferase 201 U/l and alanine aminotransferase 343 U/l. An ultrasound was performed, reporting chronic lithiasic cholecystitis and probable choledochal cyst. Magnetic cholangioresonance was performed, which reported dilatation of the common bile duct of 5 cm × 6 cm compatible with a choledochal cyst (Fig. 3). The patient was managed solely symptomatically for the remainder of her pregnancy. The pregnancy proceeded normally with an uncomplicated labour. One month after the patient was operated, a cholecystectomy was performed and resection of the choledochal cyst with hepaticojejunal anastomosis. Postoperative recovery was satisfactory and the patient was discharged with no complications.

Discussion

Babbit's theory postulates that abnormality in the biliopancreatic union with the formation of a long and abnormal common channel out of the control of the Boyden sphincters, allows reflux of pancreatic enzymes into the common bile duct and these cause inflammation, de-epithelialisation and thinning, with distal obstructions, which eventually causes the formation of the cyst. Incidence varies, but one case in 13,000 to 2 million live births, and is more common in women (4:1) and Asians. They are principally diagnosed during infancy (60%).[3–5,7,8] Type I cysts (50–85%) and type IV (30–40%) are the most common.[3,5,9]

In general, the symptoms are vague; patients present jaundice and imprecise abdominal pain. The classic presentation triad is: abdominal pain (87%), jaundice (57%) and palpable abdominal tumour in the right hypochondrium (17%); it presents only in 20% of cases.[6] Alterations are expected in liver function tests.

The most frequent complications are cholangitis and pancreatitis.[8,10,11] Other complications include biliary

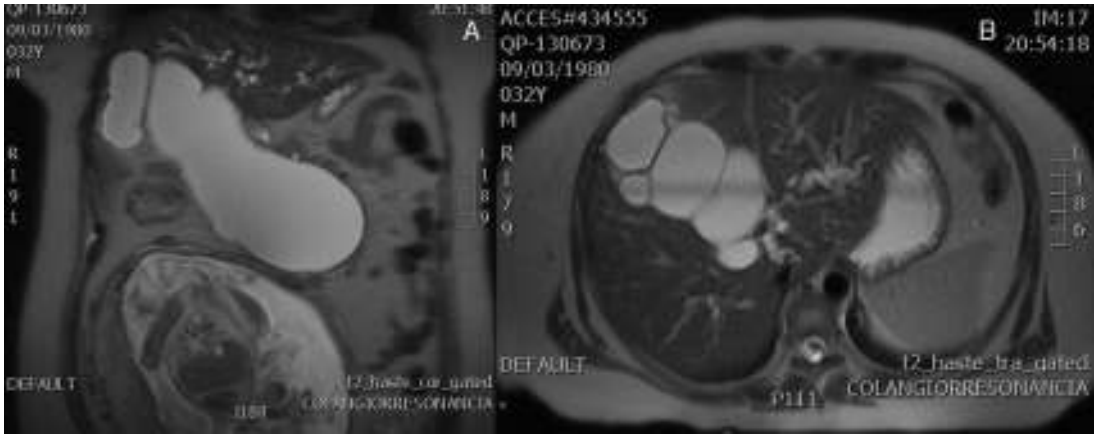

Figure 2 Coronal (A) and axial (B) section of nuclear magnetic cholangio resonance of a pregnant patient with choledochal cyst. The foetus can be seen in the lower part.

cirrhosis, portal hypertension, rupture, peritonitis, vesicular or choledochal lithiasis, and the major complication is the risk of malignancy with increases with age (0.7% in minors under 10, 7% between 10 and 20, 14% in people over 20 and 50% in people over 50).[2,5,12,13]

Choledochal cysts in pregnant women are rare, but they represent a threat to the mother and the foetus. The main expected complications are loss of the foetus or preterm labour.[12,13] Diagnosis during pregnancy is difficult, as it requires its suspected diagnosis and diagnostic aids. The most common is ultrasound, however, it is difficult during pregnancy due to the distortion of the normal anatomy of the abdomen and due to the gravid uterus.[12,14] Ionising radiation or contrast studies can be performed, such as computed tomography or retrograde endoscopic cholangiopancreatography, but with extreme precaution. Cholangiography (percutaneous, endoscopic or nuclear) demonstrates the type of cyst, intra- or extra-hepatic compromise and abnormality of the biliopancreatic union. Magnetic resonance imaging can provide adequate visualisation and relationships between the choledochal cyst (size and extension) and the biliary tree; however, it has limitations for diagnosing small cysts or duct abnormalities.[4,7,15,16]

Once the diagnosis has been made, the patients should be referred to a specialist unit, as inappropriate treatment might be catastrophic to mother and foetus. Pregnant patients constitute a special situation. The presence of the cyst in the case of very large lesions, or complications developing from these lesions is associated with a rise in maternal and foetal morbimortality. Experience in the management of this disease in pregnant women was reviewed and a summary is presented in Table 1 [1,7–21]; it also includes (at the end) the 3 patients from our study; of these, in 29 diagnosis was made during pregnancy and in 2 cases in the immediate postnatal period. Eighteen patients (69%) out of the 26 where the number of pregnancies was mentioned were primiparous. The most frequent symptoms were associated with cholangitis, with abdominal pain in 28 patients (90%), jaundice in 17 (57%) and fever in 12 (40%). A palpable tumour was only mentioned in the right hypochondrium in 6 patients (19%), although the presence of a gravid uterus should be taken into consideration, which probably affected the assessment during the physical examination. In 12 patients the pregnancy was resolved through labour (39%), in 13 through caesarean section (42%) and 4 abortions (13%). It was not mentioned in 2 cases. In 15 patients (48%) pregnancy reached 36 weeks or more, in 10 it reached 35 weeks or less and 4 (13%) cases ended in abortion. In 2 cases the weeks of pregnancy were not mentioned.

Treatment of pregnant women should be conservative when the cysts are asymptomatic.[1,3,14,19] Fifteen patients were treated conservatively. The pregnancy of nine of these

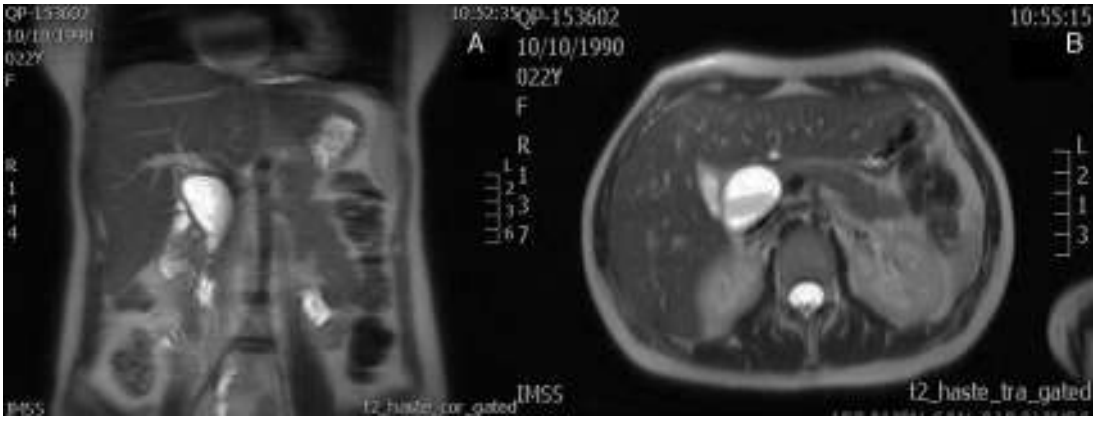

Figure 3 Coronal (A) and axial (B) section of nuclear magnetic cholangio resonance of a pregnant patient with choledochal cyst.

Table 1 Characteristics, presentation, evolution and treatment of 30 patients with a choledochal cyst diagnosed during pregnancy or the immediate post natal period.

Patient/ reference	Patient and age	Gestational age	Signs and symptoms	Type and size of cyst	Treatment given until resolution of pregnancy	Week of term of gestation	Treatment after gestation term	Evolution of patient/foetus
1/Angel et al.[1]	Primiparous, 25 years old	20 weeks, twins	Abdominal pain, nausea, choluria, acholia, jaundice, abdominal pain, nausea, jaundice	Type I, 11 cm × 8 cm	Percutaneous drainage 21.5 WG	Labour 35 WG	HJA at 6 weeks	Good/good
2/Nassar et al.[2]	Primiparous, 18 years old	18 weeks	Abdominal pain, nausea, jaundice, vomiting, tumour in right hypochondrium	Not mentioned 14.5 cm × 10 cm × 15 cm	Percutaneous and then HJA on sixth day (WG 19)	Labour 39 WG		Good/good
3/Wu et al.[3]	Primiparous, 27 years of age	20 weeks	Abdominal pain, nausea, anorexia, jaundice, vomiting, choluria, weight loss, fever, tumour in right hypochondrium	Type IV, 12 cm × 9 cm × 10 cm	Cystojejunal anastomosis and T-tube	20 WG (abortion)		Poor; death after 25 days due to septic shock/abortion
4/Gmijović et al.[3]	Primiparous, 23 years of age	36 weeks	Abdominal pain, nausea, anorexia vomiting, jaundice	Type IV, 15 cm × 3 cm × 16 cm	Percutaneous drainage	Labour 40 WG	Cystojejunal anastomosis at 4 weeks	Good/good
5/Wu et al.[3]	Multiparous, 26 years of age	28 weeks	Abdominal pain, jaundice, tumour in right hypochondrium	Not mentioned, 20 cm × 20 cm × 20 cm		Labour 28 WG	Cystojejunal anastomosis after the first week	Good/not mentioned
6/Gmijović et al.[4]	Primiparous, 28 years of age	32 weeks	Abdominal pain, fever, jaundice	Type I, 16 cm	Percutaneous drainage by US	Caesarean 39 WG	HJA at 8 weeks post caesarean	Good/not mentioned
7/Prochazka et al.[5]	Not mentioned, 29 years of age	35 weeks	Abdominal pain, nausea, hyporexia, vomiting	Type I, 12 cm × 11 cm × 10 cm		Caesarean 35 WG	HJA one week afterwards	Good/good

Table 1 (*Continued*)

Patient/ reference	Patient and age	Gestational age	Signs and symptoms	Type and size of cyst	Treatment given until resolution of pregnancy	Week of term of gestation	Treatment after gestation term	Evolution of patient/foetus
8/Prochazka et al.[5]	Not mentioned, 23 years of age	32 weeks	Abdominal pain, nausea, vomiting, jaundice, palpable hepatomegaly	Type I, 15 cm × 15 cm × 10 cm		Labour 32 WG	HJA at 2 weeks	Good/not mentioned
9/Fok et al.[7]	Second gestation, 18 years of age	18 weeks	After Caesarean abdominal pain, nausea. Tumour in right hypochondrium	Type I, 12 cm × 9 cm × 10 cm		Caesarean 38 WG	HJA 3 days afterwards	Good/not mentioned
10/Nasu et al.[8]	Primiparous, 26 years of age	18 weeks	Abdominal pain, fever	Type I, 12 cm	Percutaneous drainage	Caesarean 37 WG	HJA at 6 weeks	Good/good
11/Jabbour et al.[9]	Not mentioned, 19 years of age	28 weeks	Abdominal pain, fever, jaundice	Type I, 7.5 cm	HJA at 29 WG	Labour at term		Good/good
12/Jabbour et al.[9]	Not mentioned, 32 years of age	15 weeks	Abdominal pain	Not mentioned, 3 cm × 5 cm × 5 cm	HJA at 16 WG	Labour at term		Good/good
13/Beattie et al.[10]	Primiparous, 20 years of age	24 weeks	Abdominal pain, vomiting	Type I, 4.5 cm × 4.6 cm	Pseudocyst and pancreatic drainage at 24 WG	Caesarean 35 WG	HJA weeks later	Good/good
14/Diamond y Panesar[11]	Primiparous, 23 years of age	35 weeks	Abdominal pain, peritoneal irritation, fever	Type I, with no size		Caesarean 35 WG	Cholecystoduodenal anastomosis 4 days post caesarean	Good/good
15/Hewitt et al.[12]	Primiparous, 19 years of age	36 weeks	Jaundice	Type I, 15 cm		Caesarean 36 WG	HJA 5 weeks afterwards	Good/not mentioned
16/Hewitt et al.[12]	Primiparous, 17 years of age	16 weeks	Abdominal pain, vomiting, fever	Type I, with no size	Laparotomy due to rupture of the cyst with placement of T-tube	16 WG	HJA 6 months afterwards	Good/spontaneous abortion

Table 1 (*Continued*)

Patient/ reference	Patient and age	Gestational age	Signs and symptoms	Type and size of cyst	Treatment given until resolution of pregnancy	Week of term of gestation	Treatment after gestation term	Evolution of patient/foetus
17/Hewitt et al.[12]	Second gestation, 25 years of age	Puerpera (4 days)	Tumour in right hypochondrium	Type IV a, with no size		Term	HJA	Good/not mentioned
18/Wig et al.[13]	Not mentioned, 20 years of age	Immediate postnatal period	Abdominal pain, fever, jaundice, hepatomegaly, dehydration	Type I, 10 cm		Term	HJA	Good/not mentioned
19/Wig et al.[13]	Nulliparous, 24 years of age	20 weeks	Abdominal pain, fever, jaundice	Type I, with no size	Cholecystectomy and placement of T-tube	Abortion (24 WG)	HJA 2 months after abortion	Good/abortion
20/Binstock et al.[14]	Primiparous, 39 years of age	27 weeks	Choluria, abdominal pain, vomiting, weight loss, jaundice	Type I, 20 cm		Caesarean 35 WG	HJA during the caesarean	Good/good
21/Wu et al.[15]	Primiparous, 28 years of age	37 weeks	Abdominal pain	Type I, 13 cm		Caesarean 37 WG	Percutaneous drainage and subsequently HJA 6 weeks afterwards	Good/not mentioned
22/Wu et al.[15]	Primiparous, 30 years of age	38 weeks	Abdominal pain	Type I, 12 cm × 6 cm		Caesarean 38 WG	HJA 5 weeks afterwards	Good/not mentioned
23/Conway et al.[16]	Primiparous, 19 years of age	22 weeks	Abdominal pain, fever, jaundice	Type I, with no size	HJA at 22 WG	Caesarean 40 WG		Good/good
24/Hurtado Dayz et al.[17]	Primiparous, 20 years of age	22 weeks	Nausea, abdominal pain, vomiting, fever	Type I, with no size	External drainage (cholecys-tostomy)	36 WG	HJA 4 weeks afterwards	Good/not mentioned
25/Aszodi et al.[18]	Second gestation, 20 years of age	20 weeks	Nausea, vomiting, abdominal pain	Type I, 10 cm × 7.5 cm × 7.5 cm	Resection of cyst, Roux-en-Y HJA	Caesarean 40 WG		Good/good

Table 1 (*Continued*)

Patient/ reference	Patient and age	Gestational age	Signs and symptoms	Type and size of cyst	Treatment given until resolution of pregnancy	Week of term of gestation	Treatment after gestation term	Evolution of patient/foetus
26/Shanley et al.[19]	Multiparous, 34 years of age	34 weeks	Abdominal pain, jaundice, choluria	Type I, 10 cm × 8 cm × 14 cm		Labour 34 WG	HJA one week afterwards	Good/good
27/Arce-Sánchez et al.[20]	Primiparous, 15 years of age	31 weeks	Abdominal pain, fever, jaundice	Type I, with no size	RECP and endoprothesis 31 WG	Labour at term	Not performed	Good/not mentioned
28	Primiparous	32 weeks	Abdominal pain, Acute pancreatitis	Type I, with no size		Caesarean 34 WG	HYA at 2 weeks	Good/good
29 (case 1 of this report)	Primiparous, 22 years of age	9 weeks	Abdominal pain, fever, jaundice, Tumour in right hypochondrium	Type I, 20 cm × 11 cm	HJA at 11 WG	Abortion 11 WG		Poor; abdominal sepsis and death/abortion
30 (case 2 of this report)	Second gestation, 31 years of age	22 weeks	Abdominal pain, fever, jaundice	Type I, 25 cm × 12 cm		Caesarean 26 WG	HJA 3 weeks afterwards	Poor; gastric perforation and death/poor
31 (case 3 of this report)	Primiparous, 22 years of age	20 weeks	Abdominal pain	Type I, 6 cm × 4 cm		Labour at term	HJA one month afterwards	Good/good

HJA, hepaticojejunal anastomosis; RECP, retrograde endoscopic cholangiopancreatography; WG, weeks of gestation; US, ultrasound.

patients (60%) resolved before week 36; subsequently, 13 were reoperated for resection of the cyst with hepaticojejunal anastomosis, and 2 patients underwent cystoduodenal and cystojejunal anastomosis, respectively. One of the patients (case 2 of this report) died during the late postoperative period. Seven percent mortality is reported with this treatment.

In symptomatic cases which do not respond to treatment with antibiotics the cyst should be decompressed via the percutaneous transhepatic, endoscopic or surgical route.[3,5,8,14] In these patients 9 drainages were performed, 5 percutaneous, 3 surgical with placement of a tube (T tube in 2 cases and another with cholecystostomy) and one endoscopic retrograde cholangiopancreatography. Progress after drainage was good in 7 patients (in 5 cases the pregnancy was resolved in week 36 or later), and in 2 cases the mother progressed well but lost the foetus (both were patients operated with placement of a T-tube due to rupture of the cyst and grave cholangitis, respectively); none of the patients in this group died. After resolution of the pregnancy, 7 patients underwent resection of the cyst with hepaticojejunal anastomosis, another patient underwent cystojejunal anastomosis, and in one patient no other subsequent surgical treatment was reported.

The definitive treatment is surgical, and therefore the decision to operate should take into account the risk to the mother–child binomial, and the complications of the disease itself. If possible, surgical treatment should be performed up until the second trimester, as surgery carries a high risk of morbimortality in pregnant women.[2,12,18] Surgery is justified in the first trimester when the life of the mother is at risk, and induction of labour or caesarean section during the third trimester if the foetus is sufficiently mature.[3,9] Definitive surgical treatment was performed on 7 patients during pregnancy. In 5 cases (71%) both the mother and foetus made good progress, and in all cases the pregnancy went to term. Two cases (28%) made poor progress, with maternal death secondary to the complications of surgery.

Definitive surgery should be based on the classification of the cyst and the general condition of the patient. The surgery of choice is hepaticojejunal Roux-en-Y anastomosis owing to its low incidence of cholangitis and surgical reintervention. Resection of the cyst is widely accepted for types I, II and IV. Type III cysts require surgical drainage generally by the endoscopic route with sphincterotomy. There is controversy for type IV, the options being resection of the extrahepatic cyst alone or total resection of the cyst with hepatectomy. In type V cysts some authors recommend hepatic resection. It is recommended that resection is performed over the derivation of the cyst due to the greater incidence of carcinoma of the common bile duct in the cyst remnant.[2,3,18] At 10 years' follow-up one of the cases on whom this procedure was performed had developed a carcinoma.

Conclusions

Choledochal cyst is a rare disorder which can be found in patients during pregnancy. A successful prognosis in pregnant patients is based on the time that the disease is diagnosed, the patient's general condition, her clinical progress during monitoring and when the final treatment is given. Patients should be referred to specialist units for integral management. Conservative treatment is indicated for asymptomatic patients. Those with cholangitis should be treated with antibiotics and if necessary decompression of the bile duct through the percutaneous transhepatic route or retrograde endoscopic cholangiopancreatography, which is a temporary form of treatment. The definitive procedure (resection with hepaticojejunal anastomosis) should be based on the patient's general condition, the support of diagnostic aids and surgical experience as these are patients with a high risk of morbimortality.

Conflict of interests

The authors have no conflict of interests to declare.

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