The abnormal fetal abdomen:

Intra-abdominal cystic lesions

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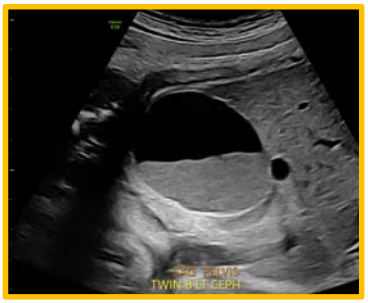
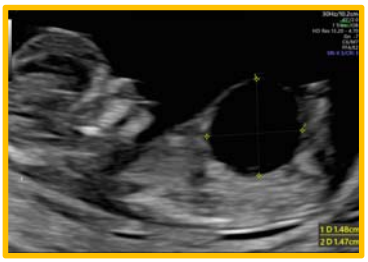
Objectives

- Overview of diagnosis and diagnostic challenges
- Differential diagnosis
- First trimester abdominal cysts
- Second/Third trimester abdominal cysts
- Role of fetal therapy
- Conclusions





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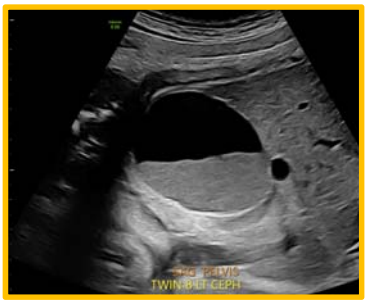
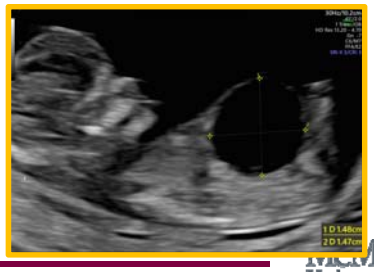
Introduction

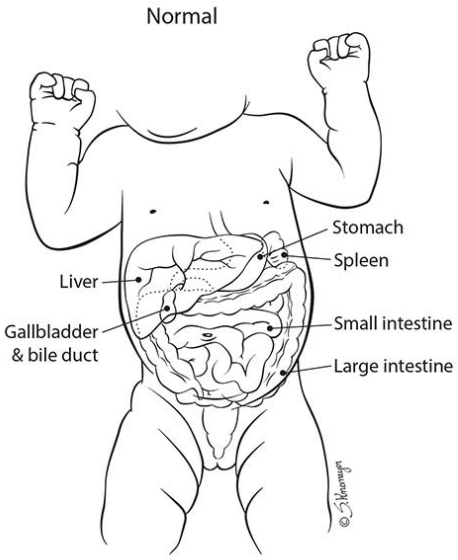
- Frequent findings at US examination.
- Easy to detect an obvious fetal abdominal cyst with US
- Outcome and prenatal counseling are dependent on:
  - Origin
  - Size
  - Associated anomalies





The challenge

- Detection rate of US in the identification of the origin of fetal abdominal cysts is still far below expectations.
- An abdominal cyst may originate from an intraabdominal structure, but an *extra-abdominal lesion*, such as pulmonary, spinal or retroperitoneal, may sometimes represent itself as an intra-abdominal cyst.
- Diagnosis is reported to be imprecise.
- Prediction of neonatal intervention is poor.
- Counseling and management remain challenging.



Fetal Diagnosis and Therapy

Original Paper

Fetal Diagn Ther 2010;28:153–159
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Perinatal Outcomes of Fetal Abdominal Cysts and Comparison of Prenatal and Postnatal Diagnoses

Ozgur Ozyuncu[a] Fuat Emre Canpolat[b] Arbay Ozden Ciftci[c] Murat Yurdakok[b]
Lutfu S. Onderoglu[a] Ozgur Deren[a]

- A database review was performed regarding the diagnosis of fetal abdominal cysts between 2002–2009.
- Structural characteristics and localizations of the cysts in the abdomen were recorded.
- Ante- and postnatal diagnoses were classified into systems according to the origin of the cyst and were compared.

Sensitivity 88%, specificity 73%, FP ~5%, PPV ~75%

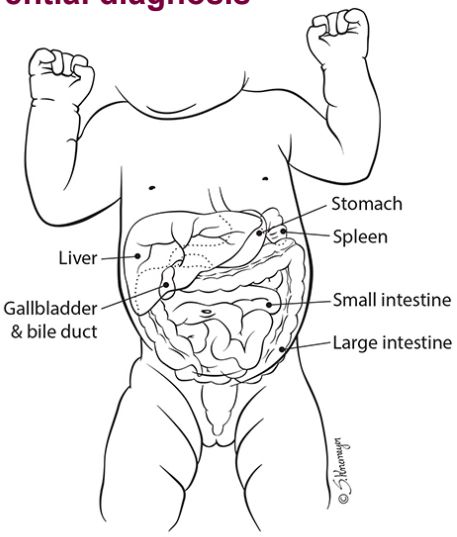
Table 3. Sensitivity, specificity, positive predictive value, false-positive rate, and accuracy of prenatal ultrasonography for the identification of the system of origin of fetal abdominal cysts

Possible origin	n	TD	FD	UC	Sensitivity	Specificity	PPV	FPR	Accuracy
Gastrointestinal	25	23	2	1	95.8	95.7	92.0	4.3	95.8
Mesenteric	2	1	1	3	25.0	98.5	50.0	1.5	94.4
Ovarian	14	8	6	2	80.0	90.2	57.1	9.8	88.7
Genitourinary	21	15	6	1	93.8	89.1	71.4	10.9	90.1
Hepatobiliary	7	3	4	0	100.0	94.1	42.9	5.9	94.4
Umbilical	1	1	0	0	100.0	100.0	100.0	0.0	100.0
Posterior abdominal wall	1	1	0	0	100.0	100.0	100.0	0.0	100.0
Total	71	52	19	7	88.1	95.7	73.2	4.3	94.8

TD = True diagnosis; FD = false diagnosis; UC = undiagnosed cases; PPV = positive predictive value; FPR = false-positive rate.

Fetal Diagn Ther 2010;28:153–159

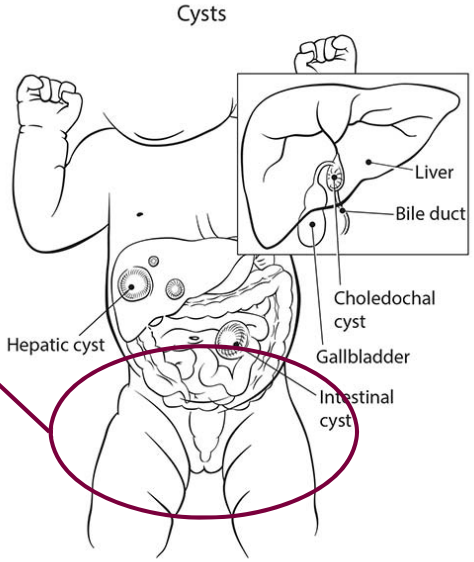
Differential diagnosis



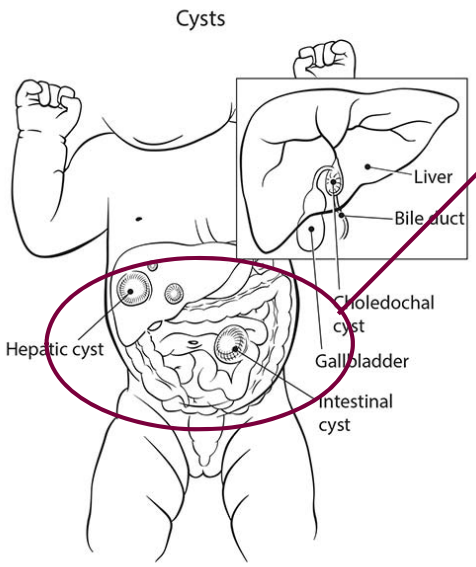
Differential diagnosis

Genitourinary tract

- Hydronephrosis
- Bladder
- Renal cyst
- Cystic renal dysplasia
- Collecting system duplication
- Ovarian cysts
- Hydrosalpinx
- Urachal cyst



Differential diagnosis



Gastrointestinal tract

- Enteric duplication
- Duodenal atresia
- Choledochal cyst
- Caroli"s disease
- Pancreatic pseudocyst
- Lymphatic or dermoid cyst within liver

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Differential diagnosis

Genitourinary tract

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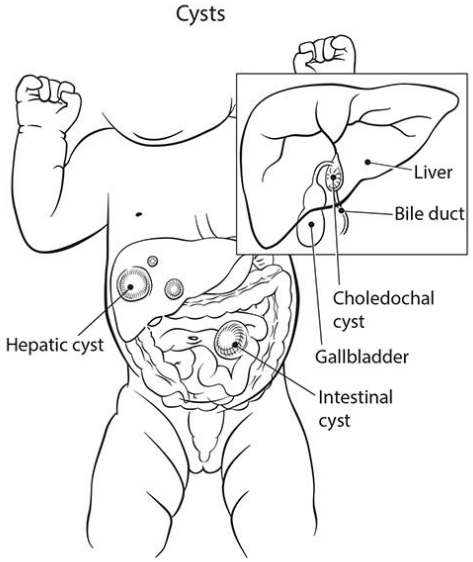

Gastrointestinal tract

- Enteric duplication
- Duodenal atresia
- Choledochal cyst
- Caroli"s disease
- Pancreatic pseudocyst
- Lymphatic or dermoid cyst within liver

Others

- Adrenal cyst
- Mesenteric or omental cyst

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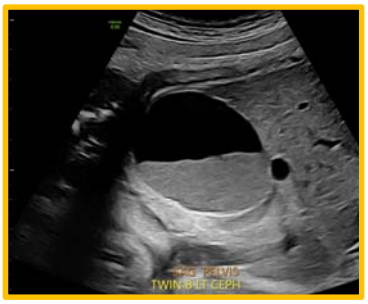
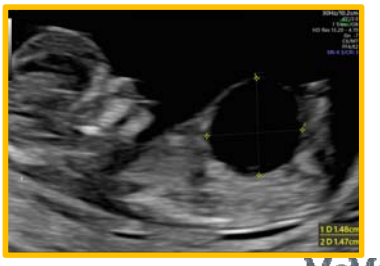
General diagnostic approach

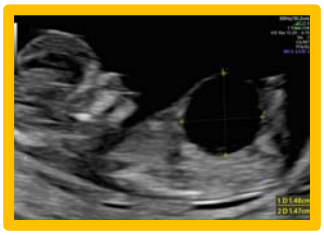
- Gestational age at diagnosis and evolution over time
  - First trimester
  - Second/third trimester
  - Persistent cystic lesions
- Location of the mass
- Echogenicity
- Type of calcification, if present (focal or scattered)
- Arterial and venous supply, if any, to the suspected abnormality
- Presence of associated abnormalities

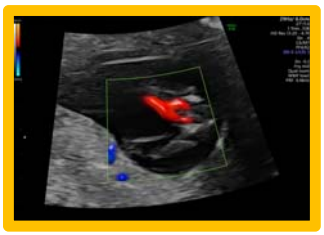




1st trimester abdominal cysts

- Role of the 1st trimester scan has expanded to include detection of many fetal structural malformations.
  - Abdominal/pelvic cysts are more frequently encountered.
- Difficult to diagnose accurately the nature of the cyst and its postnatal outcome.
- Case reports of abdominal cysts in the 1st trimester are rare, prognosis is usually good.





Khalil A. et al. Outcome of first-trimester fetal abdominal cysts: cohort study and review of the literature. Ultrasound Obstet Gynecol 2014; 43: 413–419

Table 1 Fetal abdominal cyst in the first trimester: clinical cases

Case	MA (years)	GA (weeks)	CRL (mm)	NT (mm)	Abdominal cyst (mm)	Remarks
1	27	13 + 2	77	2.3	8 × 7	Resolution. Term delivery of male infant with intestinal malrotation, midgut volvulus, necrotizing enterocolitis, mesenteric thrombosis, intestinal resection, short bowel syndrome
2	20	13 + 1	72	1.1	5 × 4	Term delivery of female neonate. Surgery for choledochal cyst at 7 weeks of postnatal life
3	17	12 + 1	60	0.7	10 × 8	Cyst aspiration at 19 weeks with subsequent resolution. Preterm rupture of membranes and delivery at 33 weeks. Normal female neonate discharged on day 17
4	31	11 + 5	53	1.0	10 × 9	Resolution. Term delivery of normal female neonate
5	28	10 + 4	36	ND	11 × 11	Resolution. Term delivery of normal male neonate

CRL, crown–rump length; GA, gestational age; MA, maternal age; ND, no data available; NT, nuchal translucency thickness.

Spontaneous resolution (n=3)	2 healthy 1 had intestinal pseudo-obstruction in neonatal life.
Persistent (n=2)	1 aspirated, resolved 1 choledochal cyst

Sepulveda W et al. Fetal abdominal cysts in the first trimester: prenatal detection and clinical significance. Ultrasound Obstet Gynecol 2008; 32: 860–864

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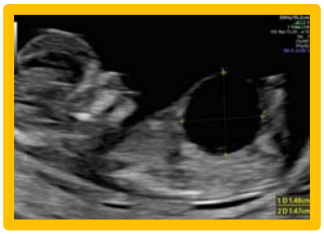
1st trimester abdominal cysts

- **Approach:** exlude megacystis.
- When megacystis is excluded differential diagnosis must be aimed at GI tract.

No other genitourinary cysts have been reported in the 1st trimester



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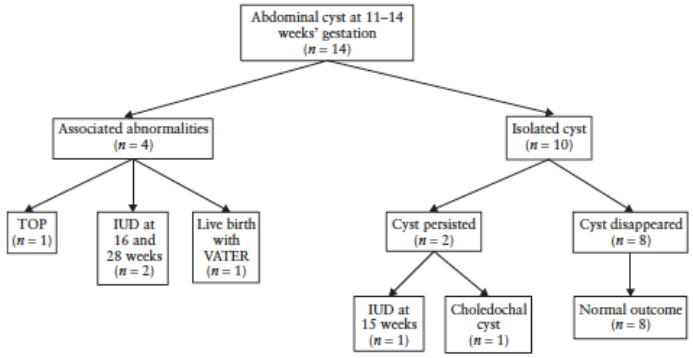
Outcome of first-trimester fetal abdominal cysts: cohort study and review of the literature

A. KHALIL, P. C. COOKE, E. MANTOVANI, A. BHIDE, A. T. PAPAGEORGHIOU and B. THILAGANATHAN

Fetal Medicine Unit, Academic Department of Obstetrics and Gynaecology, St George's University of London, London, UK



If isolated 80% disappear

All with normal outcome

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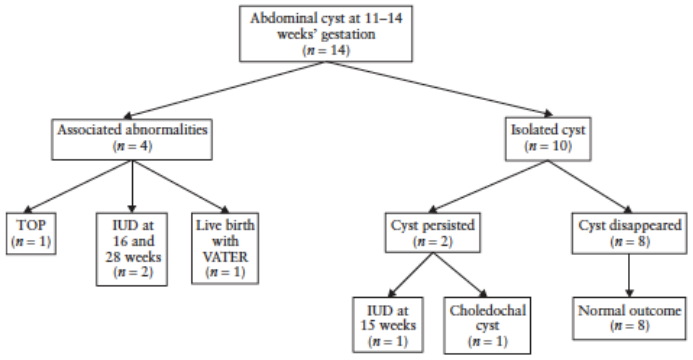

All cases of associated anomalies resulted in poor outcome

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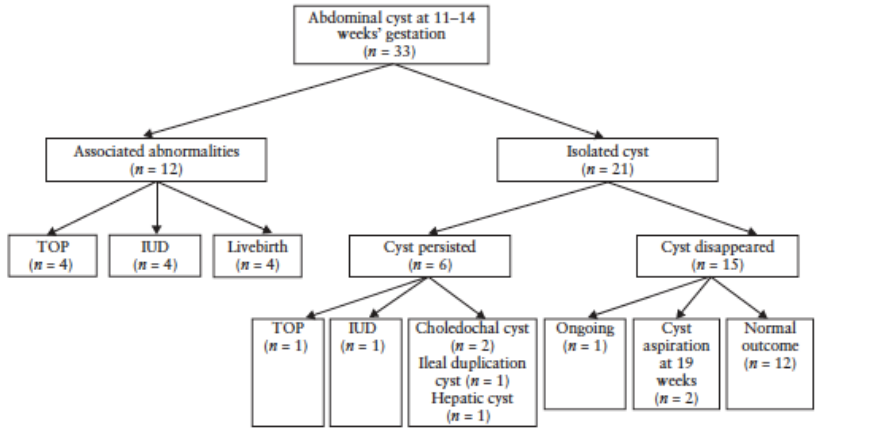

Non-isolated (37%)	Live births All had anorectal malformations
Isolated (63%)	71% disappearance rate. Persistent cysts related to GI tract anomalies

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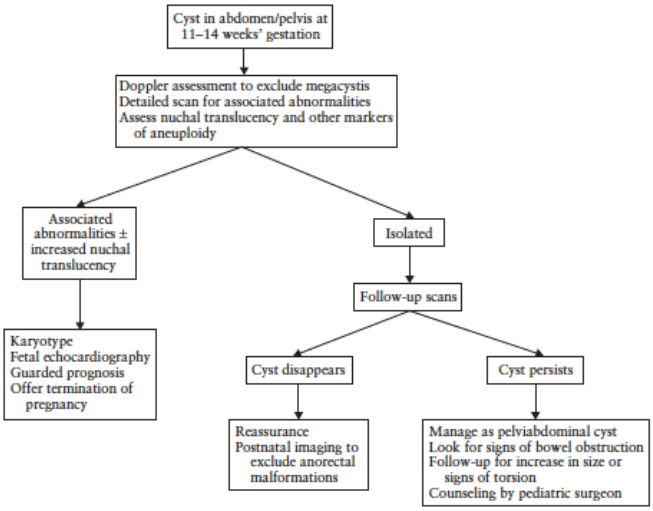
Step-wise approach to diagnosis



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Differential diagnosis → 2nd trimester

Gastrointestinal tract	Mesenteric, omental, intestinal duplication, hepatic, choledochal cysts.
Genitourinary tract	Ovarian, renal, urachal and adrenal cysts

Khalil A. et al. Outcome of first-trimester fetal abdominal cysts: cohort study and review of the literature. Ultrasound Obstet Gynecol 2014; 43: 413–419

Duodenal atresia

- The congenital absence or complete closure of a portion of the lumen of the duodenum.
- "Double bubble"
- Prevalence:
  - 1 in 5,000 – 10,000 births.
  - Males > females
- Often associated with other anomalies (50%)
- Ddx: abdominal cysts, compression (annular pancreas)

Important to demonstrate a connection between the two fluid-filled structures.

PRENATAL DIAGNOSIS
Prenat Diagn 2002; **22**: 616–623.
Published online in Wiley InterScience (www.interscience.wiley.com). **DOI:** 10.1002/pd.341

Prenatal ultrasonographic detection of gastrointestinal obstruction: results from 18 European congenital anomaly registries

Table 2—Detection rate and gestational age at prenatal diagnosis within subgroups

Gut subgroups (BPA code)	Total # (% of 349)	# PD (% total #)	95% CI	PD ≤24 WG (% of # PD)[a]	PD ≥25 WG (% of # PD)[a]
Isolated	192 (55)	60 (31)	24.7–37.9	12 (20)	45 (75)
Associated	157 (45)	59 (38)	30.0–45.2	36 (61)	20 (34)
Chromosomal	31 (9)	16		10 (62)	5 (31)
Syndromic	28 (8)	8		6 (75)	2 (25)
Multiple	98 (28)	35		20 (57)	13 (37)
Esophagus (750.3)	122	31 (25)	17.7–33.1	9 (29)	17 (55)
Isolated	73 (60)	17		3	12
Associated	49 (40)	14		6	5
Duodenum (751.10)	64	33 (52)	39.4–63.8	12 (36)	21 (64)
Isolated	38 (60)	18		3	14
Associated	26 (40)	15		9	7
Small intestine (751.11–19)	68	27 (40)	28.1–51.3	8 (30)	17 (63)
Isolated	49 (72)	21		5	15
Associated	19 (23)	6		3	2
Large intestine (751.2)	95	28 (29)	20.3–38.7	19 (68)	10 (36)
Isolated	30 (32)	4		1	4
Associated	65 (68)	24		18	6
Totals (%)[a]	349 (100)	119 (34)	29.1–39.1	48 (40)	65 (55)

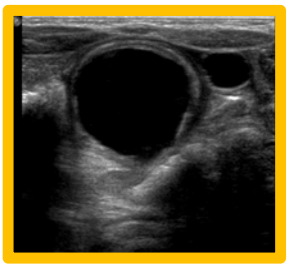
Mesenteric

- Can occur anywhere in the mesentery of GIT
- Etiologic theory - benign proliferation of ectopic lymphatics in the mesentery that lack communication with the remainder of the lymphatic system
  - Represents obstructed lymphatic drainage.
- The fluid contents may be serous, chylous or hemorrhagic.

Multi-septate or unilocular
Usually midline
Variable size – mm to cm
Solid appearance may be secondary to hemorrhage

Intestinal duplication cysts

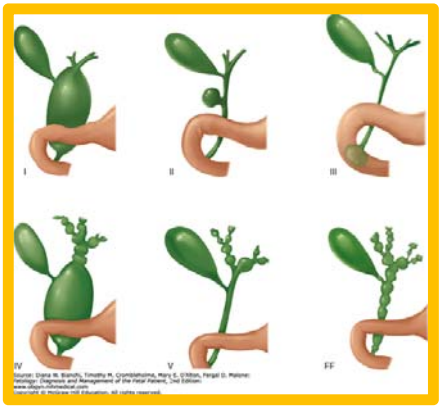
- Benign, rare anomalies that arise during early embryonic development.
- Most frequently found in the proximal small intestine, but can also occur anywhere in the GI tract.
- *Two types*:
  - Adjacent to the lumen (no communication to the gastrointestinal wall)
  - Tubular and communicate directly with the lumen
- May be isolated or associated with other malformations (spine or GI).



Peristalsis involving the cyst/mass
Cystic Non-communicating structure located on the greater curve of the stomach near the pylorus
"Double-wall" sign

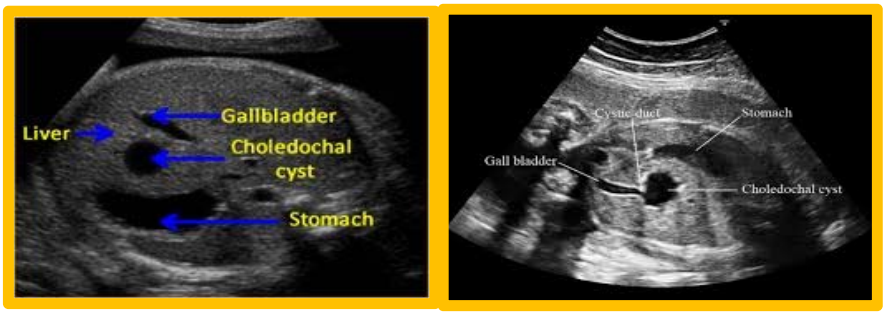
Biliary cysts/choledochal cysts

- Cystic dilations that may occur singly or in multiples throughout the biliary tree.
- Types:
  - Intrahepatic
  - Extrahepatic
- 0.005:10,000 live births



Early postnatal intervention may prevent biliary cirrhosis, portal hypertension, calculi & adenocarcinoma.
Operative mortality ~10%

Biliary cysts/choledochal cysts



Look for communication with biliary tree (not always possible by US) and distinguish from hepatic cysts (which are intraparenchymal and non-communicating)

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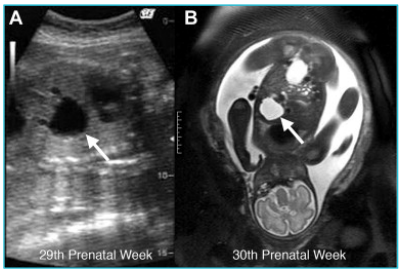
Hepatic cysts

- Very rare.
- Typically arise from Rt lobe of the liver.
- Completely intraparenchymal with no conection to biliary tree
- Result from an obstruction of the hepatic biliary system.



Unilocular
Intrahepatic cysts
Asymptomatic (rarely bleed or infection)

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Ultrasound Obstet Gynecol 2017; **50**: 167–174
Published online 9 July 2017 in Wiley Online Library (wileyonlinelibrary.com). **DOI:** 10.1002/uog.17244

Outcomes associated with fetal hepatobiliary cysts: systematic review and meta-analysis

M. LEOMBRONI[1], D. BUCA[1], C. CELENTANO[1], M. LIBERATI[1], F. BASCIETTO[1], S. GUSTAPANE[1], L. MARRONE[1], L. MANZOLI[2], G. RIZZO[3] and F. D'ANTONIO[4]

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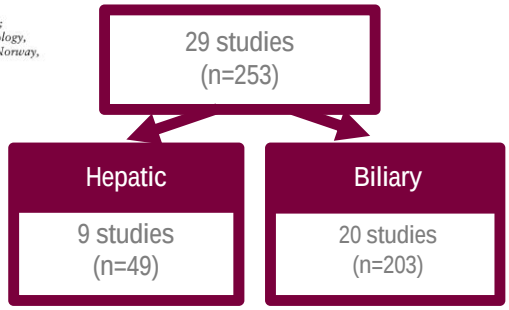
KEYWORDS: abdominal cysts; biliary cysts; hepatic cysts; outcome; prenatal diagnosis



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Biliary/choledocal cysts	▪ Clinical symptoms occurred in 55% of fetal biliary cysts and were related to bile obstruction in the large majority of cases. ▪ About half the affected children showed impaired liver function after birth. ▪ Biliary atresia was present in 22% of fetuses diagnosed as having biliary cysts on prenatal ultrasound
Hepatic cysts	▪ Good outcomes: Symptoms in 13%, postnatal surgery in 18% ▪ 4% (2/49) had another associated liver anomaly with no biliary tree anomalies.

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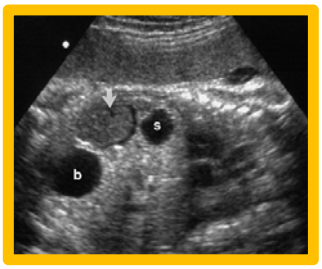
Ovarian

- Among live births, the best estimate of the incidence of clinically significant ovarian cysts is 1:2,500
- Increased frequency with advancing gestational age
- Associated with:
  - DM
  - PET
  - Rhesus isoimmunization



Non-midline regular cystic structure
Mostly unilateral
Normal urinary tract and GIT

Simple cysts <2 cm in are considered normal and physiologic. Larger and complex cysts are more likely to be non-physiologic. Associated anomalies are rare since the cysts usually result from hormonal stimulation of fetal and maternal gonadotropins

Original Paper

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Although the sensitivity, specificity and accuracy of ultrasonography in identifying the origin of an abdominal mass seem to be high, the PPV was approximately 75% = 25% the cyst may involve a different system

Incorporation of different disciplines, such as neonatology, genetics and pediatric surgery, in the counseling, management and postpartum follow-up is crucial.

What about cyst aspiration?

- Most experience & evidence from ovarian cysts.
- Antenatal aspiration of other abdominal cysts are case reports.
- Antenatal aspiration may be considered in cases of massive cysts resulting in thoracic compression.

Ultrasound Obstet Gynecol 2018; **52**: 159–164
Published online 4 July 2018 in Wiley Online Library (wileyonlinelibrary.com). **DOI:** 10.1002/uog.18973

In-utero aspiration *vs* expectant management of anechoic fetal ovarian cysts: open randomized controlled trial

C. DIGUISTO[1,2], N. WINER[3], G. BENOIST[4], H. LAURICHESSE-DELMAS[5], J. POTIN[1,2], A. BINET[6,2], H. LARDY[6,2], B. MOREL[7,2] and F. PERROTIN[1,2]

Table 3 Antenatal, delivery and neonatal outcomes of 61 pregnancies diagnosed with anechoic fetal ovarian cyst, randomized to *in-utero* aspiration or expectant management

Outcome	In-utero *aspiration* (n = 34)	*Expectant management* (n = 27)	P*	*Relative risk (95% CI)*
Neonatal intervention	7 (20.6)	10 (37.0)	0.18	0.55 (0.24–1.27)
In-utero involution of cyst	16 (47.1)	5 (18.5)	0.02	2.54 (1.07–6.05)
Gestational age at delivery (weeks)†	40 (39–40)	39 (38–39)	0.88	
Preterm delivery†	1 (3.0)	3 (11.1)	0.21 (F)	0.27 (0.30–2.47)
Neonatal or *in-utero* involution of cyst	23/32 (71.9)	15/24 (62.5)	0.45	1.21 (0.81–1.83)
Oophorectomy	1 (2.9)	6 (22.2)	0.03 (F)	0.13 (0.02–1.03)

Data are given as *n* (%), *n/n* (%) or median (interquartile range). **P* calculated using Student's *t*-test, chi-square test or Fisher's exact test (F). †Data missing for one patient from *in-utero* aspiration group.

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A systematic review and meta-analysis on fetal ovarian cysts: impact of size, appearance and prenatal aspiration

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92 studies (n=380)

Expectant (n= 324) | Aspiration (n= 56)

- Resolution 46%
- Decreased with increasing size
- Risk of torsion increased in cysts 30 – 59 mm (15-34%)

McMaster University

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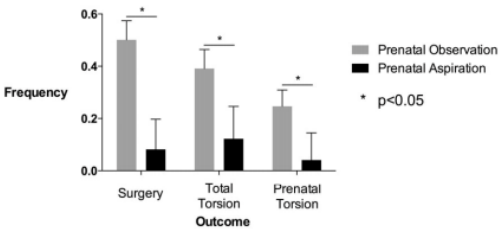

40 mm

Less torsion rate (39% vs 12%)
Less prenatal torsion (25% vs 4%)
Less postnatal surgery (63% vs 8%)

Big ovarian cysts that are aspirated prenatally have less risk of overall torsion, antenatal torsion and need for post-natal surgery.

Less risk of oophorectomy

McMaster University

Conclusions

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