Remenans et obstructio

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BVN – GBN 27/11

Content

01

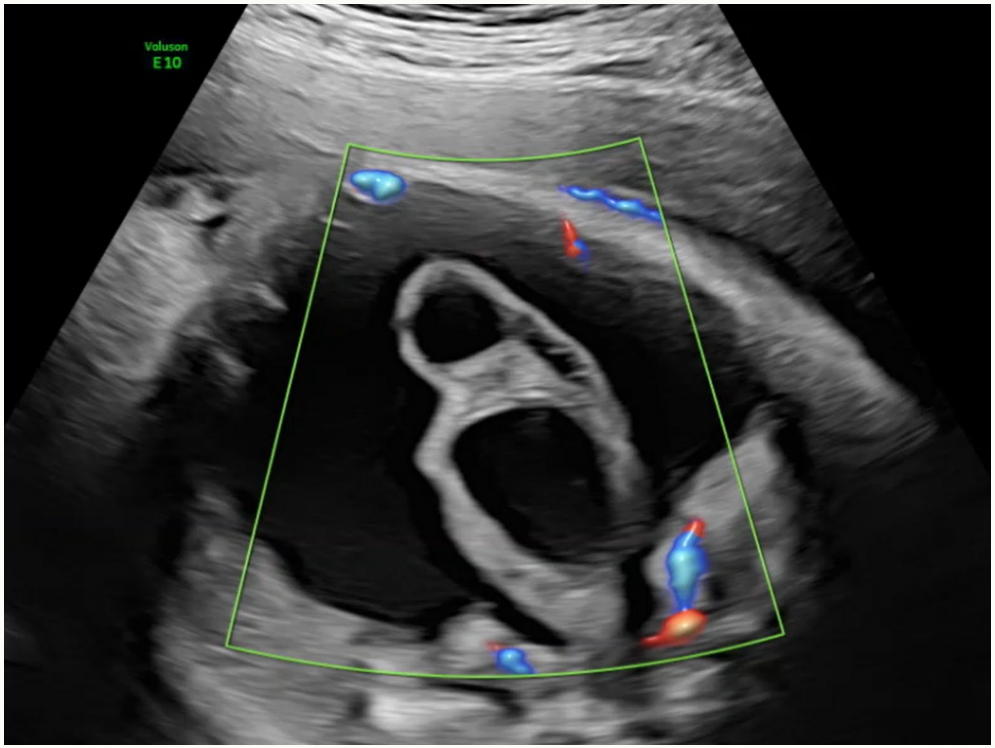
Pregnancy

Prenatal

Unproblematic pregnancy of a boy

PML 35w: referral to UZ Gent: intra-abdominal cyst

- Diagnosis of ascites
- No pleural effusion, no pericardial effusion, normal heart structure
- Impression of thickened bladder wall
- No hydronephrosis



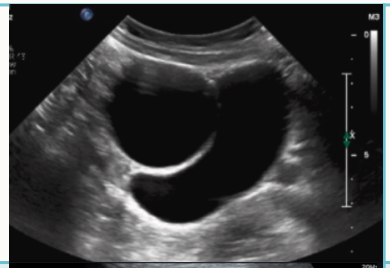
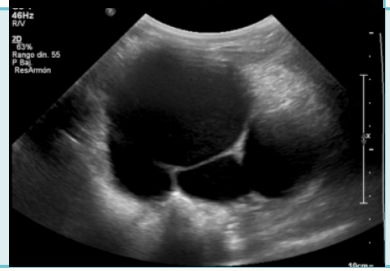
Differential diagnosis prenatal intra-abdominal cyst?

- Urachal cyst
- Kidney cyst
- Hydronephrosis

- Ovarian cyst

- Mesenteric cyst
- Choledochal cyst
- Meconium pseudocyst
- Remnant omphalomesenteric duct
- Duplication cyst
- Spleen/liver cyst

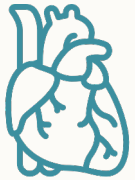
Prenatal intra-abdominal cyst

Urachal cyst		Meconium pseudocyst	
Hydronephrosis		Choledochal cyst	
Ovarian cyst		Duplication cyst	
Mesenteric cyst			

Differential diagnosis prenatal ascites?

- Urinary tract obstruction
- Bladder rupture

- Volvulus
- Perforation
- Bile duct obstruction



- Congenital heart disease
- Foetal arrythmia

-> heart failure

- Cardiomyopathy

- Severe anaemia (e.g., parvovirus, alpha thalassemia major, Rhesus immunization)

- TORCHES
- Parvovirus

- Chromosomal abnormalities (Trisomy 21, 18, 13. Monosomy X)
- Inborn errors of metabolism

Prenatal

- TORCHES: negative
- Structurally normal heart, no arrhythmia
- No hydrops fetalis
- NIPT normal male
- No elevated peak systolic velocity

Differential diagnosis prenatal ascites?

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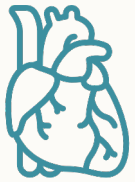

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02

Birth

Birth

Caesarean section PML 36w4d

No clear origin of cyst
Significant increase in ascites
After lung maturation treatment

Surprisingly good start

Spontaneous crying
APGAR 9/10/10
Urinates on delivery table
Large abdomen

Placement of ascites drain

And placement of bladder catheter

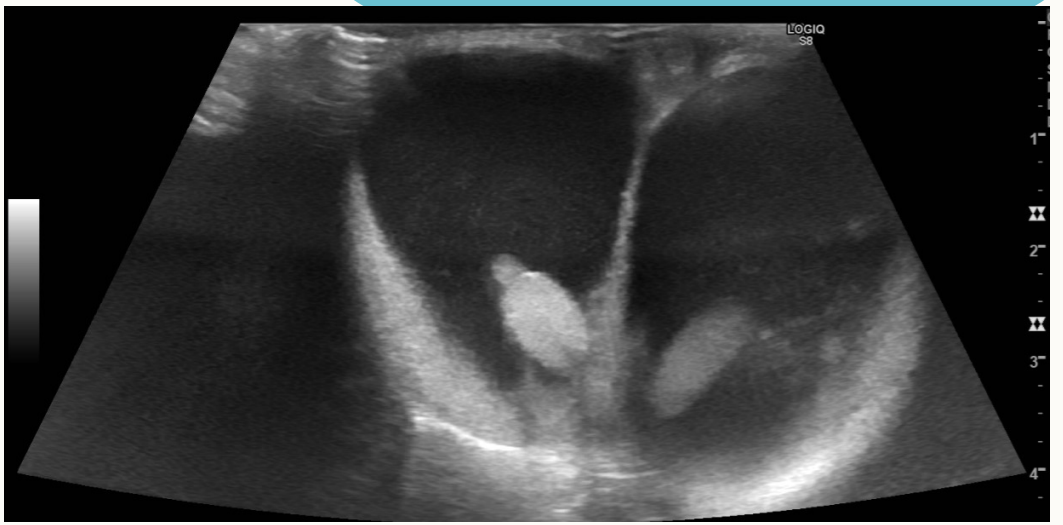
Further diagnostics

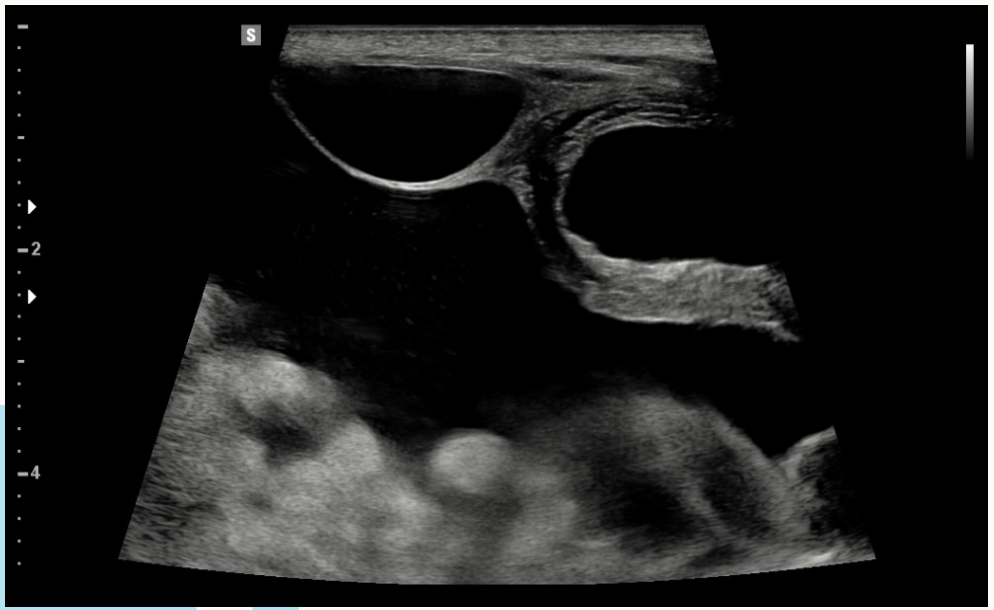
03

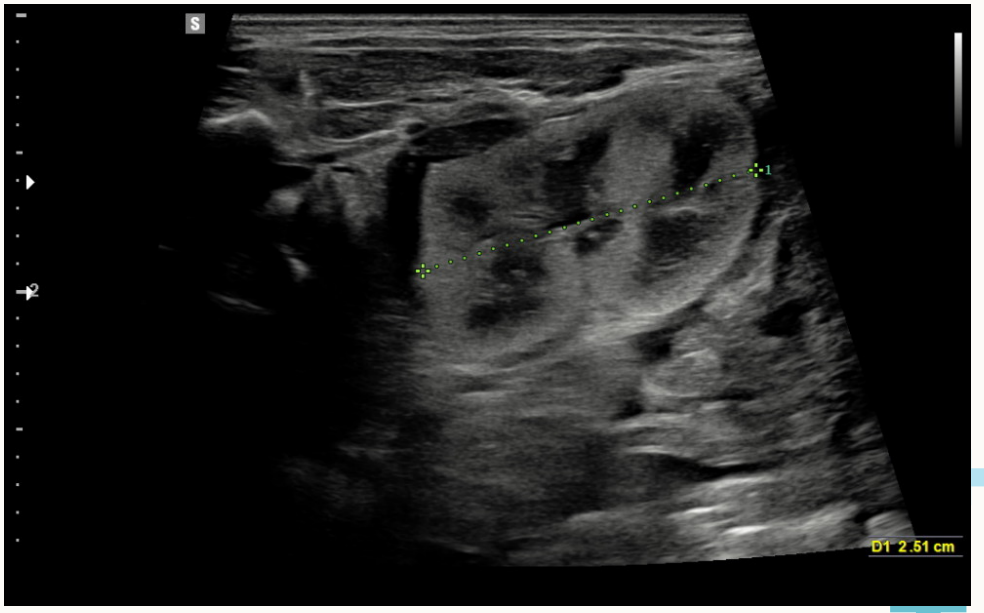
Diagnostics

Investigations

Abdominal ultrasound D1:







Investigations

Chest X-ray D1:

Echocardiogram D1:

- Structurally normal heart
- No pleural effusion, no pericardial effusion
- Signs of hypovolemia

Investigations

Blood analysis D1:

	Value	Unit	Normal value
Hb	20,8	g/dL	10 – 13.5
Platelets	351	10^3/µL	150 – 450
WBC	18.78	10^3/µL	7 – 15
CRP	< 0,6	mg/L	< 5
Na	136	mmol/l	136 – 145
K	4,8	mmol/L	3,6 – 4,8
Cl	105	mmol/L	98 – 106
Creatinine	0,91	mg/dL	0,17 – 0,42

	Value	Unit	Normal value
ALT	12	U/L	13 – 45
GGT	124	U/L	12 – 122
LDH	602	U/L	135 – 750
Total bili	2,1	mg/dL	3,4 – 11,5
Total protein	53	g/L	60 – 76
PT	20,4	sec	11,7 – 15,3
INR	1,5	/	0,9 – 1,1
APTT	48,4	sec	25,4 – 59,8
Fibrinogen	166	mg/dL	200 – 400

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Investigations

Urine D1:	Value	Unit	Normal value
Glucose	negative	/	negative
Total protein	negative	/	negative
Creatinine	20	mg/dL	20 – 400
WBC	104	/µL	0 – 25
RBC	12	/µL	0 – 25

Ascites D1:	Value	Unit
Glucose	84	mg/dL
Total protein	15	g/L
Creatinine	0,90	mg/dL
WBC	64	/µL
RBC	< 1000	/µL

Investigations

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Composition is different

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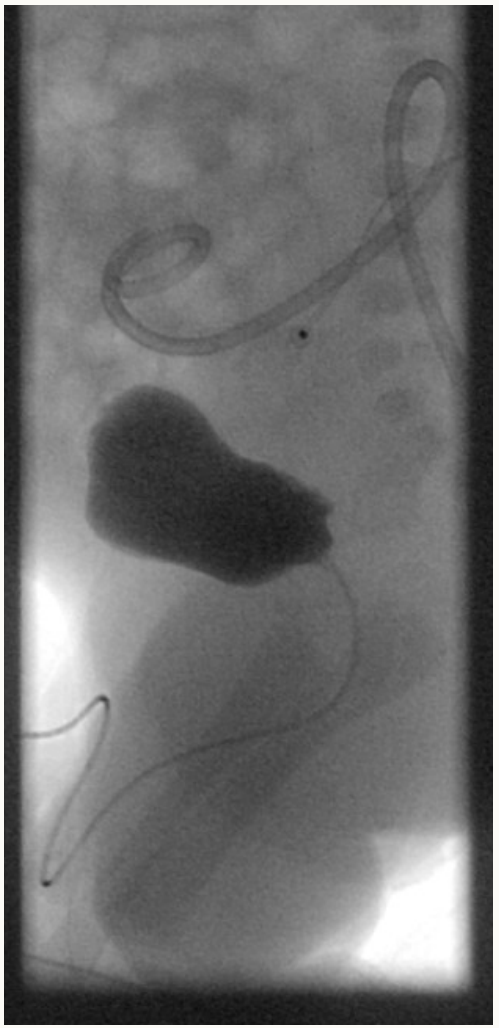
Total protein	15	g/L
Creatinine	**0,90**	mg/dL
WBC	**64**	/µL
RBC	**< 1000**	/µL

Investigations

RX cystography D1

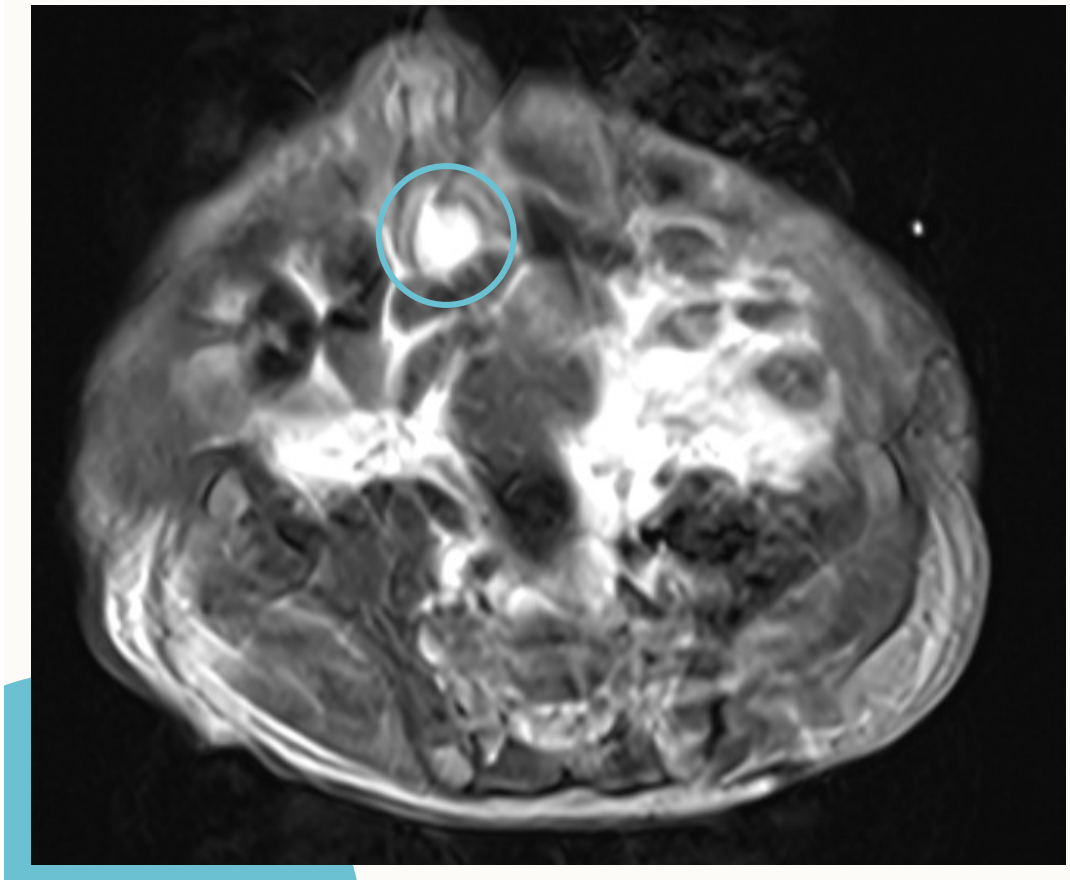
- Good filling of the bladder
- No signs of perforation/leakage or filling of a urachal anomaly

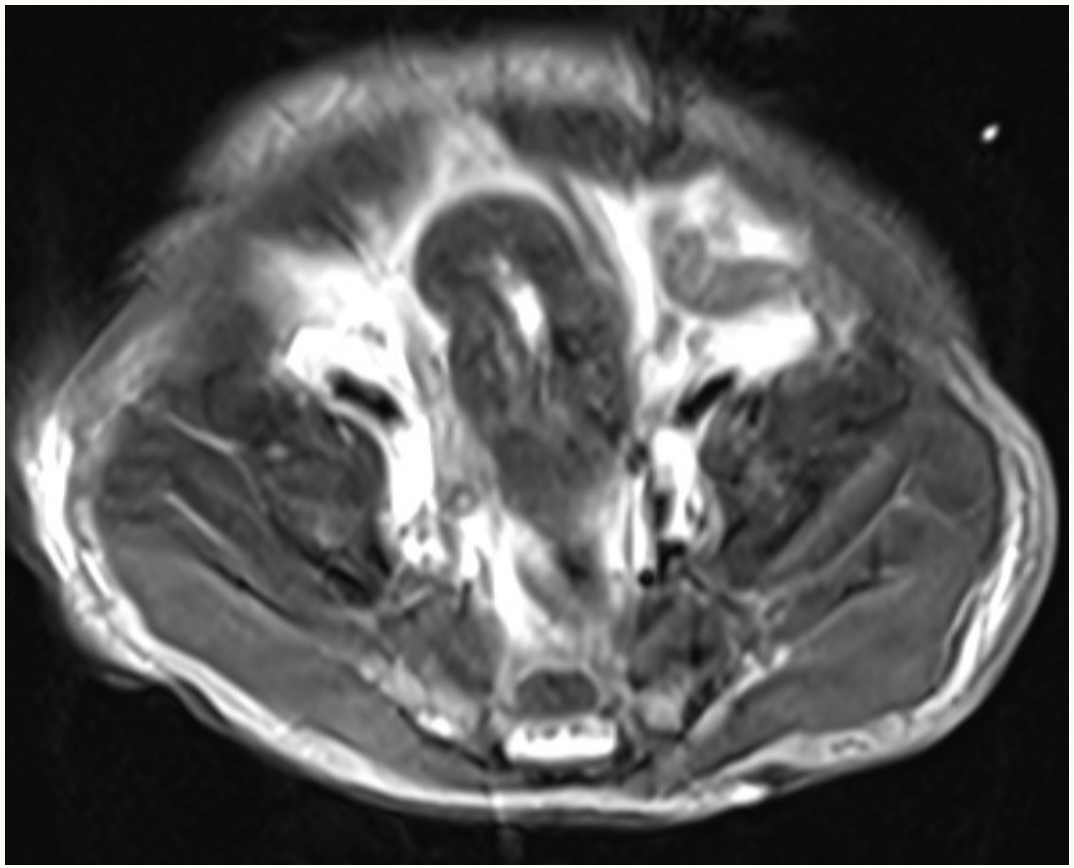


Investigations

= Urachal cyst

MRI D5

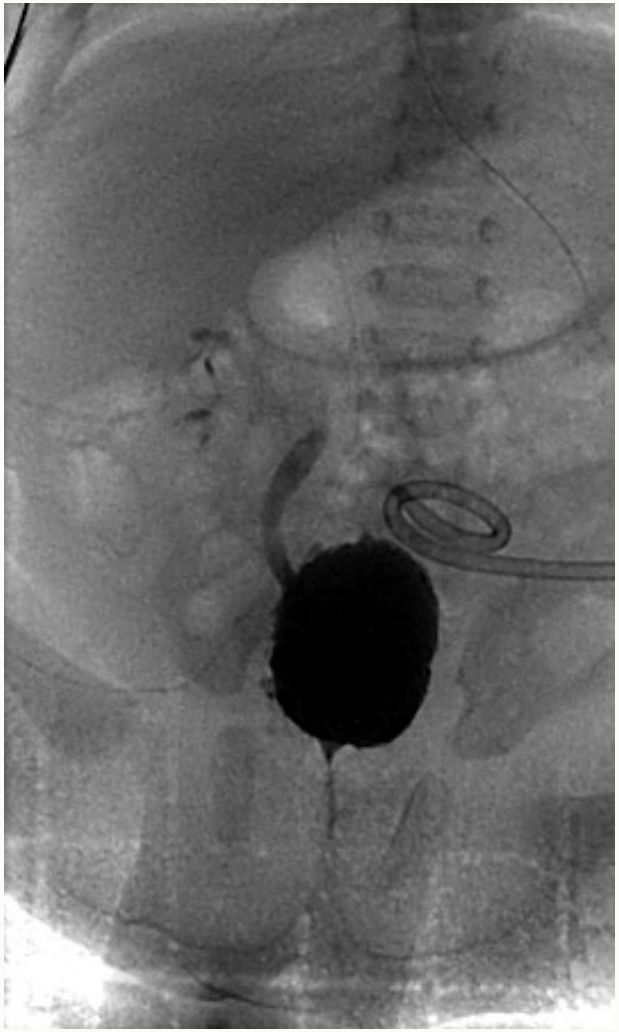




Investigations

RX cystography D7

- Active reflux grade 1
- Bladder trabeculation
- Permictional narrowing at the posterior urethra, suggestive of posterior urethral valves



Diagnosis

Urachal cyst

+

Posterior urethral valves

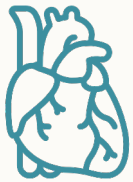
04

Course

Course

Extubation D3, HFNB D3

Coagulopathy D1 requiring 1x plasma



Vascular depletion requiring fluid bolus and compensation for ascites
Control echocardiogram D12: normal

Subfebrile and CRP 35 mg/L on D3:
Vanco-Amika D3-10. Cultures negative
Afterwards: uroprophylaxis with trimethoprim

Ascites drain D1-3, clamped D3-7
Abdominal circumference stable – no increase in ascites fluid

Start of enteral feeding D4
Fully enteral D8

Cystoscopy: confirmation of posterior urethral valves.
Valve resection not possible; meatus too narrow

Course

- Discharge home with transurethral bladder catheter in situ
- New cystoscopy for valve resection -> successful
- No resection of urachal cyst to date

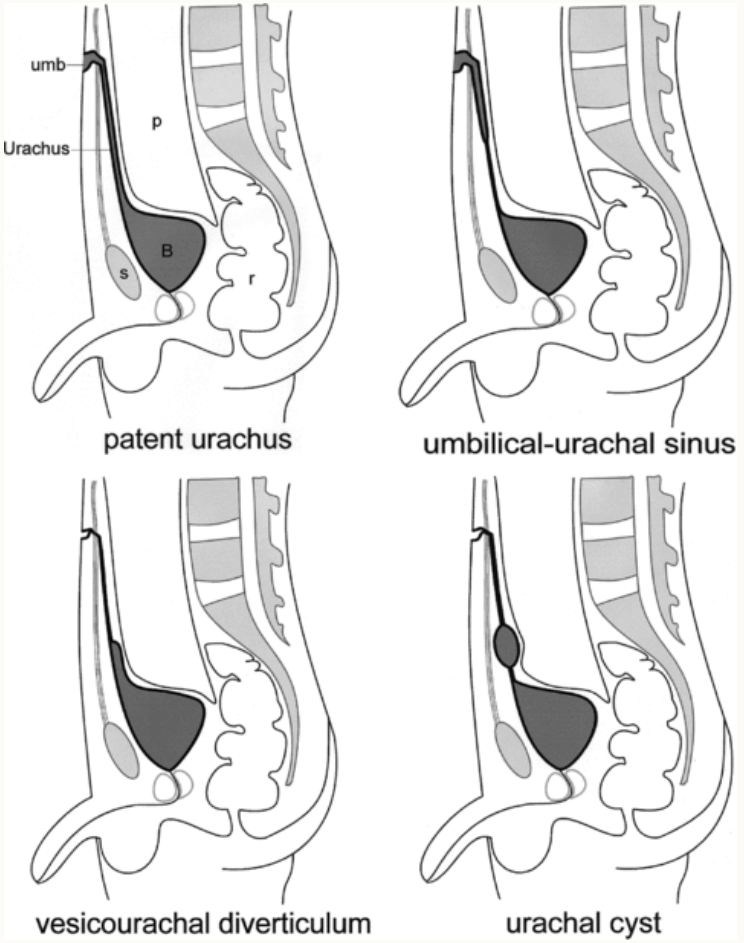
05

Discussion

Urachal cyst/anomaly

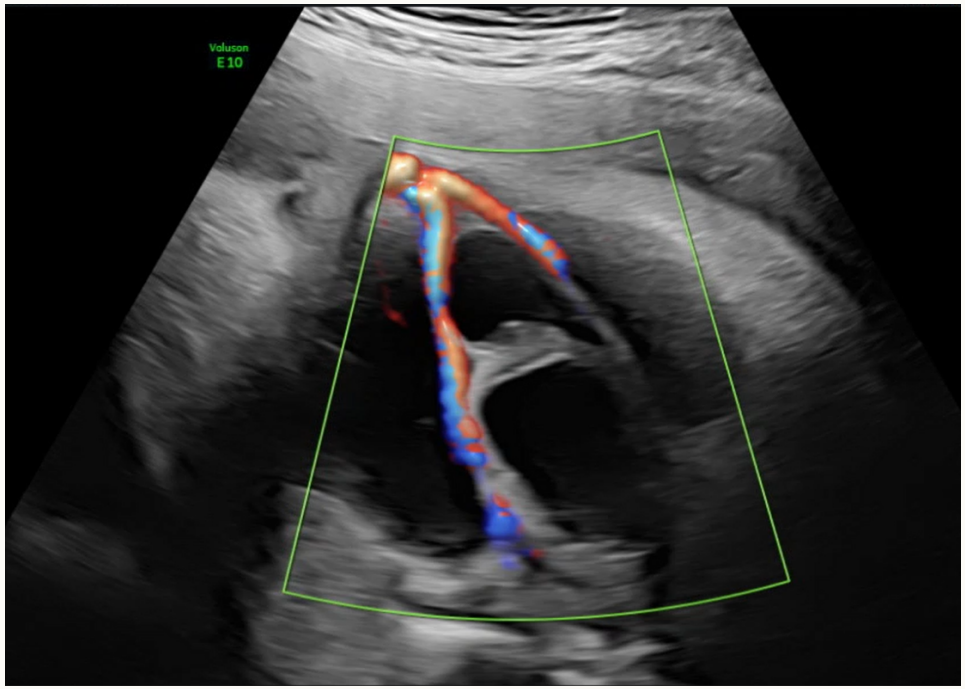
- Embryonic canal connecting the navel to the bladder, that does not close completely
- True incidence = unknown
- Often incidental finding
- Clinical findings
  - Umbilical drainage
  - Lower abdominal pain
  - Abnormal appearance of umbilicus
  - Infection
- May rupture or abcedatie

  -> Rare in neonates



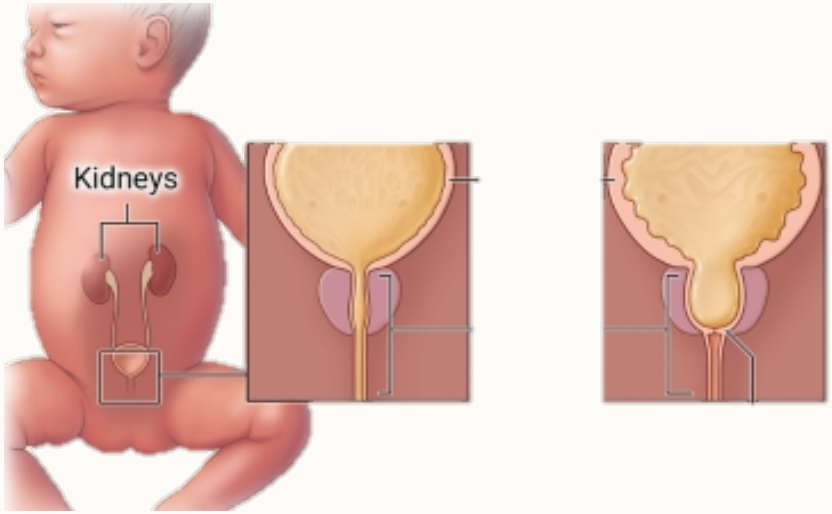
Urachal cyst/anomaly

- Diagnostics
  - Ultrasound
  - RX cystography
- Conservative treatment and follow-up or surgical excision



Posterior urethral valves

- Obstructing membranous folds within the lumen of the posterior urethra
- Often leads to bladder trabeculation, vesicoureteral reflux, and hydronephrosis
- Clinical findings
  - Urinary tract symptoms: poor urinary stream, pyelonephritis, urosepsis
  - Abdominal distension (overdistended bladder of ascites)
  - Respiratory distress (due to lung hypoplasia in oligohydramnios)



Posterior urethral valves

- Diagnostics
  - Ultrasound: hydronephrosis, dilated and thickened bladder, dilated posterior urethra (keyhole sign)
  - RX cystography
- Treatment: valve resection

1) No hydronephrosis

Presumably, pressure caused by posterior urethral valves could “escape” through the urachal cyst, rather than rising to the kidneys

2) Ascites

2 proposed aetiologies:

- Rupture – open communication
- Transudation

1) No hydronephrosis

Presumably, pressure caused by posterior urethral valves could “escape” through the urachal cyst, rather than rising to the kidneys

2) Ascites

2 proposed aetiologies:

- Rupture – open communication
- **Transudation**

3) Composition urine – ascites

If fluid in the abdomen is urine, the composition should match and thus contain a high creatinine and urea.

Presumably, different composition due to partial resorption of ascites by the peritoneum

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Total protein	15	g/L
Creatinine	**0,90**	mg/dL
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Questions?

Thank you for your attention

06

Sources

- Fetal intra-abdominal cysts: accuracy and predictive value of prenatal ultrasound, Vincenzo Davide Catania et al.
- Differences in Origin and Outcome of Intra-Abdominal Cysts in Male and Female Fetuses, Marjolein Husen et al.
- Reabsorption of ascites and the factors that affect this process in cirrhosis, Sinan Akay et al.
- Fetal Abdominal Cysts: Prenatal Diagnosis and Management, Abdullah Serdar Açıkgöz et al.
- Management of symptomatic urachal cysts in Children, E. Basuguy et al.
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- Urachal anomalies: ultrasonography and management. Ueno T et al.
- Current diagnosis and management of urachal remnants. Naiditch JA et al.
- Urachal anomalies: a longitudinal study of urachal remnants in children and adults. Ashley RA et al.
- Neonatal Ascites Associated with Urinary Outlet Obstruction (Urine Ascites). Rogelio Moncada et al.
- Diagnostic Aspects of Neonatal Ascites: Report of 27. Thorne Grisco et al.

Aetiology ascites

- **Urinary tract obstruction**: Rupture fornix/calix kidney or transudation
- **Bladder rupture**: leakage of urine
- **Volvulus**: mostly chylous –> blockage and damage to lymphatic vessels
- **Bowel perforation**: leak of intestinal contents and inflammatory response
- **Bile duct obstruction**: indirectly through liver damage (portal hypertension), directly due to leakage
- **Congenital heart disease/arrhythmia/cardiomyopathy**: right sided heart failure
- **Severe anaemia**: cardiac failure
- **Infection**: not always clear pathogenesis. Parvovirus: attacks red blood cell progenitors, hepatocytes, and myocardial cells causing transient aplastic crisis, hepatitis, and myocarditis
- **Aneuploidy**: obstruction or incomplete formation of the lymphatic system, cardiac failure
- **Inborn errors of metabolism**: congestion of abdominal viscera from accumulation of these metabolites