Fluid in too many spaces and places: Hydrops

Objectives
• Illustrate the appearance of fluid in different body cavities
• Discuss differential diagnosis of etiology hydrops
• Discuss use of MCA Doppler in assessment of fetal anemia

What is Hydrops?
Abnormal interstitial accumulation of fluid in body cavities
  Pleural
  Pericardial
  Peritoneal
Need 2 of these for diagnosis of hydrops
OR
1 body cavity fluid + anasarca

Placentomegaly and polyhydramnios are common, but not needed for diagnosis of hydrops

What causes Hydrops?
Late stage of many processes that lead to redistribution of body fluids among the intravascular and interstitial compartments

Etiology
Imbalance of interstitial fluid
  • Myocardial failure
  • High cardiac output failure
  • Decreased colloid oncotic plasma pressure (anemia)
  • Increased capillary permeability
  • Obstruction of venous and/or lymphatic flow

How do we diagnose fluid in the different body cavities/spaces?

Ascites is seen as fluid between bowel loops, along the abdominal flanks, around the liver, and/or outlining umbilical vessels
Care should be taken to not mistake the normal abdominal wall muscles or abdominal wall fat
For ascites (so called “pseudoascites”).
Isolated ascites can be an early sign of hydrops
But if truly isolated can be due to:
   - Urinary etiology such as obstructive uropathy
   - GI obstruction meconium peritonitis
More favorable prognosis than hydrops
Needs f/u to ensure that hydrops does not ensue

Small Pleural Effusion
Unilateral small pleural effusion does not shift the mediastinum
If marked mediastinal shift, assess for hernia or other chest mass
Prolonged large pleural effusions had lead to pulmonary hypoplasia

Primary Chylothorax Is the most frequent cause of an isolated pleural effusion leading to respiratory distress in the newborn. This is typically unilateral with mediastinal shift, and a flattened hemidiaphragm. Treatment is with drainage.

Pericardial Fluid
2 mm or less is normal
Large pericardial effusion can impact on venous return and lead to hydrops

Anasarca – diffuse body wall edema. It is important to appropriately measure the fetus with anasarca
   - BPD draw around the skull, not the skin
   - AC draw around the skin

What causes hydrops?
   - Immune
   - Non-immune
Prior to RhoGAM immune hydrops was >80% of all hydrops
Now, nonimmune hydrops represents 90% of hydrops

Immune Hydrops
Erythroblastosis Fetalis
Antibodies in maternal circulation destroy fetal RBCs
   - Anemia
   - Hepatosplenomegaly
   - Increased fetal erythropoiesis
   - Hypoproteinemia
   - CHF
Maternal Rh sensitization
Fetal maternal hemorrhage
   - Delivery
   - Abortion (spontaneous or therapeutic)
   - Amniocentesis
   - Abruption
   - Incompatible blood transfusions
Transplacental hemorrhage
- Occurs in 75% of pregnancies
- Less than 0.1 mL in 60% of cases
- 3% risk of sensitization of D-negative woman

RhoGAM (Rhesus immunoglobulin)
- 300 mg at 28 weeks
- Protects against 30 mL fetal blood
- If a greater degree of fetomaternal hemorrhage is suspected, a Kleihauer-Betke test can be utilized to quantify fetal blood in maternal circulation to determine dose needed

Immune hydrops – atypical antibodies
Develop in 1-2% of individuals after blood transfusion
2% hemolytic disease of fetus (Kell, RhC, E)

For obstetric /fetal procedures such as amniocentesis or other invasive procedure
- If patient is Rh – give RhoGAM within 48 hours
- If father of baby is Rh - some would omit this

Caucasian etiology of nonimmune hydrops
- 1/3000 live births
  - Cardiac 20-40%
  - Infection 5-10%
  - Chromosomes 16%
    - Turner syndrome
    - Trisomy 13, 18, 21
    - Triploidy

Pathophysiology

Increased hydrostatic pressure
- Primary myocardial failure
  - Arrhythmia
  - Anemia
  - TTS
  - Myocarditis
  - Cardiac malformation
- High output failure
  - AV shunt

Decreased plasma oncotic pressure
- Decreased albumin formation (cirrhosis, hepatitis)
- Increased albumin excretion (Finnish nephrosis)

Increased capillary permeability
- Anoxia
- Infection

Obstruction of lymph flow
- Turner syndrome
- Some combination of the above

Mirror syndrome is when there is maternal anasarca in a pregnancy complicated by fetal hydrops
Maternal pathology "mirrors" fetal pathology
- Massive edema
- Oliguria
- Hemodilution

MCA Doppler in hypoxemia
- Central redistribution of blood flow
- Increased blood flow to the brain
- Elevated PSV in cases of anemia

MCA Doppler technique
- Circle of Willis
- Transverse plane of the fetal head at the base of the skull
- Proximal MCA in longitudinal view, almost parallel to the ultrasound beam

Prognosis in hydrops is grim
- 70% mortality
- 100% if structural abnormality identified
- Transfusion of anemic fetuses can reverse hydrops

PUBS
- Check MCV > 100 to prove fetal
- Check hematocrit to determine amount of transfusion needed
- Hct < 30% is 2.5\textsuperscript{th} percentile beyond 20 weeks
- Use packed RBC (Hct >90%), O negative, irradiated
- Goal is Hct of 40

Diagnostic Approach
- Maternal studies
  - Blood group typing
  - Indirect Coombs test
  - CBC
  - Kleihauer-Betke
  - Syphilis, parvovirus, CMV, toxo, other
- Fetal studies
  - Ultrasound – no cause found in 15-30% of cases
  - Echocardiography
  - Amniocentesis
  - Karyotype
  - a-fetoprotein
  - Antigen tests by PCR and culture for syphilis, CMV, toxo, other
• MCA Doppler – if elevated then PUBS
• Follow-up
• Getting the best diagnosis is helpful in counseling regarding recurrence risks