Hydrops Fetalis

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Patient History

- 16 YO G1P0, 17 weeks gestation by LMP
- Referred for full fetal ultrasound evaluation by physician for abnormally low maternal AFP
- Some causes of low maternal serum AFP include incorrect dates, Down syndrome (trisomy 21), Edward’s syndrome (trisomy 18), and fetal demise.
17w Fetus Sagittal Body

Findings consistent with hydrops fetalis
Hydrops fetalis

• First described in 1892
• Latin for “edema of the fetus”
• Incidence of 1/1500 to 1/4000 deliveries
• Characterized by generalized edema and fluid collections in serous cavities
• Peripheral edema may be significant (anasarca)
Diagnosis of Hydrops Fetalis

- Fluid accumulations must involve more than 1 site for the term “hydrops” to be used.

- If fluid collection in only one cavity (i.e. isolated ascites) - can have a better prognosis, and narrows differential diagnosis.
Sonographic Findings

- Subcutaneous edema (thickness 5mm)
- Pleural Effusion
- Ascites
- Pericardial Effusion
- Polyhydramnios
- Placental Thickening (more than 6 cm)
All abnormal images are from our patient’s 17 week fetus.

All comparisons are from a normal 17 week fetus.
Sonographic Findings

• Subcutaneous edema (thickness 5mm)
• Pleural Effusion
• Ascites
• Pericardial Effusion
• Polyhydramnios
• Placental Thickening (more than 6 cm)
Hydropic versus Normal Fetal Scalp

Hydrops

Normal
Hydrops, Scalp Edema

- Placenta
- Cranium
- Skin
- Falx Cerebri
Sonographic Findings

- Subcutaneous edema (thickness 5mm)
- Pleural Effusion
- Ascites
- Pericardial Effusion
- Polyhydramnios
- Placental Thickening (more than 6 cm)
Pleural Effusion & Ascites

- Abdominal Contents
- Heart/Lungs
- Ribs
- Placenta
- Pleural Effusion
- Diaphragm
- Ascites
- Abdominal Contents

BIDMC

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Hydrops Versus Normal Abdomen

Hydrops

Normal
Normal

- Placenta
- Spine
- Liver
- Abdominal Wall
- Stomach
Hydrops

- Spine
- Placenta
- Ascites
- Abdominal Wall
- Bowel
Sonographic Findings

- Subcutaneous edema (thickness 5mm)
- Pleural Effusion
- Ascites
- Pericardial Effusion
- Polyhydramnios
- Placental Thickening (more than 6 cm)
Pericardial Effusion

- Ribs
- Heart
- Lungs
- Pericardial Effusion
Sonographic Findings

• Subcutaneous edema (thickness 5mm)
• Pleural Effusion
• Ascites
• Pericardial Effusion
• Polyhydramnios
• Placental Thickening (more than 6 cm)
Gross Placental Pathology

http://www.echt.chm.msu.edu/courseware/blockII/Pathology/Gest_14.html
Microscopic Placental Pathology

Normal Placenta

Hydropic Placenta

http://www-medlib.med.utah.edu/WebPath/PLACHTML/PLAC076.html
http://www-medlib.med.utah.edu/WebPath/PLACHTML/PLAC097.html
Careful sonographic analysis of the fetus revealed further abnormalities:

1. Congenital Heart Defect
2. Cystic Hygroma
Normal versus Abnormal Fetal Heart

Hydrops

Normal
Normal Heart

- Ribs
- Vertebra
- RV
- LV
- LVOT
- LA
- RA

BIDMC
Abnormal Heart

There is a single AV valve. Other views (not included) showed a single ventricular outflow tract, consistent with complex congenital heart disease.
Can you diagnose the abnormality on this sagittal fetal ultrasound?
Cystic Hygroma

- Cranium
- Skin
- Spine
- Placenta
- Cystic Hygroma
Cystic Hygroma

• Incidence 1/12,000 births
• Develops around 40d gestation - failure of embryonic lymphatics to connect with the venous system, causing dilation.
• Half have Turner’s syndrome (XO), 10-15% have trisomy 18, 21, 13 (total 60-75% aneuploid)
• Usually associated with elevated AFP
Differential Diagnosis

• Immune Hydrops - alloimmune hemolytic disease/Rh isoimmunization (etiology for the majority of cases prior to 1960s)

• Nonimmune Hydrops - LARGE number of causes! Currently comprises approximately 75% of cases
Some Causes of Non-Immune Hydrops

- **Hematologic** - homozygous α-thalassemia
- **Congenital Infections** - Secondary syphilis, parvovirus, CMV, HSV type I
- **Cardiovascular** - Structural anomalies, tachyarrhythmias, bradyarrhythmias
- **Lymphatic Abnormalities**
- **Pulmonary Malformations** - Pulmonary sequestration, CCAM
Some Causes of Non-Immune Hydrops (cont.)

- Chromosomal Abnormalities - Trisomies (13, 21, 18), triploidy
- Neoplasms - Sacrococcygeal teratoma
- Placental Abnormalities - Chorioangioma
- Metabolic/Storage Disorders - Gaucher’s disease, mucopolysaccharidosis
Prenatal Evaluation of Hydrops

• Complete ultrasound evaluation +/- fetal echo
• Exclusion of isoimmunization - Rh incompatibility
• Amniotic fluid analysis with cultures/PCR
• Chromosomal evaluation
• Maternal serology
Prognosis of Non-Immune Hydrops Fetalis

• 50% of all cases diagnosed in utero result in fetal death.
• 50% of all live born infants with non-immune hydrops die.
• Exact prognosis depends on the underlying condition - some fetal infections (i.e. parvovirus B19) can have remission of hydrops.
Poor Prognostic Indicators

- Identification prior to 24w gestation
- Structural Malformations, esp. Cardiac
- Chromosomal Disorder
- Pleural Effusions/Pulmonary Hypoplasia
- Severe Hydrops
- Prematurity
References

Books:

Articles:

Web Sites:
Merck Manual Online - Chapters 18 & 19
www.merck.com/pubs/mmanual
University of Minnesota - Neonatology Department Teaching Files
www.peds.umn.edu/divisions/neonatology/tfiles/hydrops.html
Brigham & Women’s Radiology cases (BrighamRad)
http://brighamrad.harvard.edu/cases/bwh/hcache/36.full.html
Pathology Web Sites:
http://www.echt.chm.msu.edu/courseware/blockII/Pathology/Gest_14.html
http://www-medlib.med.utah.edu/WebPath/PLACHTML/PLAC076.html
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