

Nonimmune Hydrops Fetalis



This guideline was updated in April 2012 by Saul Snowwise with input from members of the New Zealand Maternal Fetal Medicine Network.



Background

Nonimmune Hydrops Fetalis (NIHF) is now responsible for 90% of all hydrops. It has an estimated incidence of 1/1500 to 1/3800 births. The pathogenesis of NIHF is not clear, but it is associated with numerous potential mechanisms and underlying disorders. NIHF has an obvious poor prognosis for the fetus, but can also have maternal consequences as well with a 10% incidence of Mirror syndrome.

Objective

To provide guidance and a consistent approach for the accurate diagnosis and management of mothers and fetuses presenting with nonimmune hydrops fetalis at any point in gestation.

Definition

NIHF is established by the ultrasound findings of at least two of the following:

- Ascites
- Pleural effusion: *ANY* fluid abnormal;
- Pericardial effusion: > than 2mm
- Skin edema: > than 5mm on chest and scalp
- Polyhydramnios: Max pocket > 8cm, or AFI > 24cm
- Placentomegaly: Thickness >4cm



Differential Diagnosis

- Immune hydrops
- Isolated fluid collection
 - Scites
 - Pleural effusion
 - Pericardial effusion
- Skin edema
- Polyhydramnios or placentomegaly

Underlying Conditions

- Aneuploidy
 - Most common cause of NIHF under 24/40
- Cardiovascular abnormalities
 - Structural
 - Most common cause of NIH over 24/40
 - Functional
 - Arrhythmias
 - Tachyarrhythmia
 - Bradyarrhythmia
- Vascular
 - Shunt or Thrombosis
- Thoracic abnormalities
 - CCAM



- Diaphragmatic hernia
- Masses
- Pulmonary sequestration
- Chylothorax
- Airway obstruction
- Non cardiac/thoracic anomalies
 - Lymphatic
 - Gastrointestinal
 - Genitourinary
 - Neurological/Decreased movement
- Anaemia
 - Decreased production
 - Parvovirus
 - Infiltration/Storage diseases
 - Myeloproliferative/ congenital leukaemia
 - Increased loss
 - Intrinsic cell abnormality (alpha-thalassemia, G6PD, membrane...)
 - Hemangioma
 - Haemorrhage/abruption
- Infectious disease
 - toxo, syphilis, varicella, adenovirus, coxsackie
- Metabolic storage disease



- placental
 - TTTS/TRAP
 - Trauma
 - Cord anomalies
 - Chorio-angioma
- Idiopathic
 - Decreasing significance with knowledge

Clinical Presentation

- 35% incidental finding
- Size > dates
- Decreased fetal movement
- Mirror Syndrome in mother

Important History

- Personal and family history to look for inheritable disorders associated with
 - Alpha thalassemia
 - Metabolic disorders
 - Genetic syndromes
- Infectious exposures
 - Parvovirus
- Consanguinity



Evaluation

- Detailed ultrasound
 - anatomy
 - MCA PSV
 - echocardiogram
- Laboratory
 - Mother
 - Blood group and RBC antibody screen
 - Baseline BP and urinalysis
 - FBC and film screen for thalassemia
 - Parvo, toxo, rubella, syphilis,
 - HSV(if recent primary infection)
 - Kleihauer-Betke
 - Infant
 - Amniocentesis
 - Karyotype
 - PCR for TORCH pathogens
 - Storage for further testing
 - Cordocentesis
 - RBC for anaemia
 - metabolic/genetic tests



Prognosis

The overall perinatal mortality rate is 50 – 98%. There has been no significant change over past 15 years. Mortality rates will vary according to

- Gestation (earlier has worse prognosis)
- Pleural effusions (worse prognosis, esp. if 20/40)
- Underlying etiology

Management options include;

- Termination
- Selective therapeutic intervention if possible
- Ongoing pregnancy with hydrops
 - Monitor for PET/Mirror syndrome
 - Consider neonatal palliation
 - If for active intervention
 1. Monitor with CTG's or BPP's
 2. Delivery at tertiary care center
 3. Consider risks of:
 - birth trauma
 - PPH
 - non reassuring fetal heart
 - dystocia
 - caesarean

Low recurrence if no inheritable disorder identified



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