

HYDROPS FETALIS

DEFINITION

- Abnormal accumulation of fluid in ≥ 2 compartments of the fetus (pleural and pericardial effusions, ascites and/or subcutaneous oedema)
- Often associated with polycythaemia and placental thickening
- High but variable mortality rates dependent on underlying cause

TYPES

- Traditionally classified into 2 types:
- non-immune hydrops fetalis occurs in the absence of maternal antibodies; accounts for 90% of fetal hydrops in Western countries
- immune hydrops fetalis occurs when maternal allo-immune antibodies are produced against fetal red cells causing haemolysis; rare since introduction of anti-D immunoglobulins

AETIOLOGY

- Imbalance of fluid movement between fetal intravascular and interstitial spaces
- Multiple causes including cardiac abnormalities (structural or arrhythmias), chromosomal/genetic, infection, haematological, metabolic and non-cardiac structural anomalies
- No identifiable cause found in 15–31% of babies

ANTENATAL MANAGEMENT

- Hydrops fetalis is diagnosed antenatally via ultrasound
- Refer to fetal medicine team [important as confirmed antenatal diagnosis aids appropriate counselling of families, and further intensive monitoring required throughout pregnancy (discussion of this is beyond the scope of this guideline)]
- Possible antenatal interventions include intra-uterine blood transfusion and in-utero procedures e.g. paracentesis/thoracentesis
- High risk of premature delivery

Refer all antenatally diagnosed hydrops fetalis to a regional fetal medicine centre for further assessment and management

NEONATAL MANAGEMENT

Resuscitation

- Resuscitation and stabilisation can be difficult
- An expert team including a neonatal consultant should be present at delivery
- Manage according to Neonatal Life Support (NLS)

Consider concurrent pleural/ascitic drains to facilitate resuscitation

- In cases of severe anaemia, give urgent Group O RhD negative blood transfusions
- Baby may need further grouped and crossmatched blood transfusions in NNU

Give only CMV negative and irradiated blood

Ventilation

- Ensure adequate oxygenation and ventilation
- May require high frequency oscillatory ventilation [see **Ventilation: high frequency oscillatory ventilation (HFOV)** guideline] and muscle relaxation
- If pulmonary hypertension present may require nitric oxide (see **Nitric oxide** guideline)

Cardiovascular system

- Use inotropes to support heart and blood pressure
- If intravascular fluid depletion give colloid

- Strict fluid balance
- If severe compromise may require further pleural and ascitic taps
- Immune hydrops may require exchange transfusion. See **Jaundice** and **Exchange transfusion** guidelines

NEONATAL INVESTIGATIONS

- Due to the extensive list of potential causes, direct investigations according to clinical history and presentation
- Initial investigations to consider include:

	Initial investigations	Further investigations to be considered if underlying cause is not ascertained
Haematology	<ul style="list-style-type: none"> • FBC (including blood film) • Group and direct Coombs test • Maternal Kleihauer test 	<ul style="list-style-type: none"> • Red cell enzyme deficiency (e.g. G6PD deficiency) • Red cell membrane defects (e.g. hereditary spherocytosis) • Haemoglobinopathies (e.g. thalassaemia)
Biochemistry	<ul style="list-style-type: none"> • Liver function tests including albumin • Urea, creatinine and electrolytes 	<ul style="list-style-type: none"> • If pleural/ascitic tap done – send for fluid MC+S and biochemistry
Cardiac	<ul style="list-style-type: none"> • ECG to exclude cardiac dysrhythmias • Echocardiography to exclude structural heart defects 	
Placenta	<ul style="list-style-type: none"> • Send to pathologist 	
Genetic testing	<ul style="list-style-type: none"> • Chromosomes • Microarray 	<ul style="list-style-type: none"> • Investigate for congenital metabolic conditions • Non-immune hydrops – discuss whole exome sequencing with genetics team
Infection	<ul style="list-style-type: none"> • Toxoplasma, rubella, CMV, parvovirus, herpes simplex virus 	
Radiology	<ul style="list-style-type: none"> • Chest X-ray • Abdominal X-ray • Cranial ultrasound scan 	<ul style="list-style-type: none"> • Further investigations to be guided by clinical picture

Even with optimal management, the mortality rate is high. Suggest a post-mortem in the event of a death