Hydrops Fetalis

Synonyms: hydrops, fetal hydrops, foetal hydrops, universal oedema of the newborn

First described by Ballantyne in 1892, this is a serious condition defined as abnormal fluid accumulation in two or more fetal compartments. These may include ascites, pleural effusion, pericardial effusion and skin oedema. It may also be associated with polyhydramnios and placental oedema.

Hydrops is traditionally classified into either immune (particularly rhesus (Rh) blood group isoimmunisation) or non-immune hydrops fetalis (NIHF). However, over 90% of hydrops in the Western world is of non-immune origin.[1]

Epidemiology

The incidence of fetal hydrops is reported to be 3 to 24 per 10,000 live births.[2] The incidence varies according to the population risk of the conditions listed under 'Aetiology', below. For example, in Thailand the expected frequency of hydrops, from homozygous alpha thalassaemia or Bart's hydrops is much higher.

Aetiology

One review found the causes of NIHF as follows: cardiovascular (20.1%), haematological (9.3%), chromosomal (9.0%), syndromic (5.5%), lymphatic dysplasia (15.0%), inborn errors of metabolism (1.3%), infections (7.0%), thoracic (2.3%), urinary tract malformations (0.9%), extra thoracic tumors (0.7%), twin-to-twin transfusion syndrome (4.1%), gastrointestinal (1.3%), miscellaneous (3.6%) and idiopathic (19.8%).[3]

Haematological causes

- Isoimmunisation (haemolytic disease of the newborn, erythroblastosis fetalis), antibodies to red cell antigens including Rh, ABO, Duffy and Kell.
- Other haemolytic disorders - eg, glucose-6-phosphatase dehydrogenase (G6PD) deficiency, glucose phosphate isomerase (GPI) deficiency, pyruvate kinase (PK) deficiency.
- Disorders of red cell production - eg, congenital dyserythropoietic anaemia, Diamond-Blackfan syndrome, lethal hereditary spherocytosis, congenital erythropoietic porphyria ( Günther's disease), alpha thalassemia (Bart's haemoglobinopathy).
- Fetal haemorrhage - eg, intracranial or intraventricular, hepatic laceration or subcapsular, fetomaternal haemorrhage or twin-to-twin transfusion.

Cardiac causes

- Abnormalities of left ventricular outflow - eg, aortic valvar stenosis or atresia, coarctation of the aorta, truncus arteriosus, hypoplastic left heart, endocardial fibroelastosis.
- Abnormalities of right ventricular outflow - eg, pulmonary valvar atresia or insufficiency, Ebstein's anomaly, arteriovenous malformations, haemangiomas.
- No structural anomalies - eg, superior vena cava or Inferior vena cava occlusion, intrathoracic or abdominal masses, disorders of lymphatic drainage, arrhythmias; supraventricular tachycardia or congenital heart block (66-75% occur in pregnancies complicated by maternal collagen disease; prenatal closure of the foramen ovale or ductus arteriosus, myocarditis or idiopathic arterial calcification or hypercalcaemia).
Infective causes

- Parvovirus B19 (slapped cheek syndrome) - this is increasingly recognised as being important. Use of polymerase chain reaction diagnostic testing has demonstrated that perhaps 20% of hydrops fetalis is associated with this maternal/fetal infection.
- Cytomegalovirus (CMV).
- Syphilis.
- Herpes simplex virus.
- Toxoplasmosis.
- Hepatitis B.
- Adenovirus.
- Coxsackievirus type B.
- Listeria monocytogenes.
- Ureaplasma urealyticum.

Metabolic and other causes

- Inborn errors of metabolism - eg, glycogen-storage disease type IV; lysosomal storage diseases.
- Hypothyroidism and hyperthyroidism.
- Chromosomal syndromes - eg, trisomies 10,13,15,18 or trisomy 21 (Down’s syndrome); Turner syndrome (45, X).
- Numerous other autosomal-recessive genetic disorders.

Tumours

Especially sacrococcygeal teratoma.

Investigations

Evaluation of hydrops begins with an antibody screen (indirect Coombs’ test) to determine if it is non-immune, detailed sonography of the fetus(es) and placenta, including echocardiography and assessment for fetal arrhythmia, and middle cerebral artery Doppler evaluation for anaemia, as well as fetal karyotype and/or chromosomal micro-array analysis, regardless of whether a structural fetal anomaly is identified. [4]

One review recommended the following for NIHF: [5]

- Fetal chromosome analysis and genetic testing.
- Imaging studies, including comprehensive obstetric ultrasound (including arterial and venous fetal Doppler) and fetal echocardiography.
- Investigation for maternal-fetal infections.
- Alpha-thalassaemia testing in the mother if at risk because of ethnicity.
- To evaluate the risk of fetal anaemia, Doppler measurement of the middle cerebral artery peak systolic velocity should be performed in all hydropic fetuses after 16 weeks of gestation.
- For suspected fetal anaemia, fetal blood sampling and intrauterine transfusion should be offered rapidly.
- All cases of unexplained fetal hydrops should be referred to a medical genetics service where available. Detailed postnatal evaluation by a medical geneticist should be performed on all cases of newborns with unexplained NIHF.
- Amniotic fluid and/or fetal cells should be stored for future genetic testing.

Management

This depends on an accurate diagnosis of the cause of the hydrops. Interventions are sometimes possible and helping the parents decide whether to continue the pregnancy or opt for a termination depends upon whether the abnormalities are compatible with the continued survival of the fetus and problems likely after birth.

Recommended treatment depends on the underlying cause and gestational age. Preterm delivery is recommended only for obstetric indications. Candidates for corticosteroids and antepartum surveillance include those with an idiopathic aetiology, an aetiology amenable to prenatal or postnatal treatment, and those in whom intervention is planned if fetal deterioration occurs. Such pregnancies should be delivered at a facility with the capability to stabilise and treat critically ill newborns. [4]

Parental involvement and guidance are fundamental requirements and require full knowledge by the parents of all possible potential consequences. If the decision is made to continue the pregnancy, the next step is to decide whether to intervene with invasive fetal treatment. Consideration should also be given to the point at which preterm delivery represents less risk for the fetus than continuing the pregnancy.

Intrauterine intraperitoneal fetal transfusion with packed RBCs has been replaced by intravascular (umbilical vein) transfusion as the treatment of choice for fetal anaemia.

Treatment for fetal arrhythmias has included doing nothing, administering drugs and immediate delivery if maturity permits. Fetal pacing has been reported. [6] Various drugs have also been used. Adenosine is effective with supraventricular arrhythmias and maternal oral dexamethasone has been used in cases of hydrops-related fetal heart block. [7] A combination of amiodarone and digoxin has been used for treatment of refractory atrial flutter. [7]
Surgery is occasionally used to correct malformations associated with hydrops — eg, cystic adenomatoid malformations and bronchial sequestration.

**Prognosis**

Survival in hydrops depends on the underlying disease, on available fetal therapies to resolve hydrops and on the gestational age of delivery.\[8\]

Spontaneous remission has been reported in hundreds of cases. Underlying causes in these cases include cardiac arrhythmias, twin-to-twin transfusion syndrome, cystic hygroma with or without Noonan’s syndrome,\[9\] both parvovirus B19 and CMV infections and idiopathic ascites or pleural effusions.\[10\]

**Further reading & references**

- Friedman DM; Congenital Heart Block. Clinical neonatology and perinatology, 2009

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