

Intensive Care Nursery House Staff Manual

Hydrops Fetalis

INTRODUCTION: Hydrops fetalis is an excess accumulation of fluid in the fetus. Depending on the severity and cause of hydrops, there may be edema of fetus and placenta, ascites, pleural effusions and/or pericardial effusions. In previous years, most cases of hydrops were caused by severe erythroblastosis fetalis secondary to Rh iso-immunization (see Hemolytic Disease of the Newborn, P. 121). With the marked decrease in this condition (due to prophylaxis with immune globulin), most cases of hydrops fetalis are now caused by other conditions and are known as non-immune hydrops. **The rest of this section deals only with non-immune hydrops fetalis.**

ETIOLOGY: Non-immune hydrops fetalis can be caused by a wide variety of factors. A list of the more common causes is shown in Table 1. In approximately 1/4 of all cases, the cause is not determined.

MANAGEMENT:

1. Antenatal management should be directed towards making a diagnosis, with the aim of identifying those in whom either prenatal or immediate post-natal intervention may be effective. Diagnostic techniques include fetal ultrasonography, fetal echocardiography, examination of maternal blood for fetal erythrocytes (Kleihauer-Betke test), amniocentesis and sampling of fetal blood. In some cases, fetal intervention is effective (*e.g.*, fetal transfusion for anemia due to Parvovirus B19 infection, treatment of fetal tachycardia). In others, delivery corrects the underlying problem (*e.g.*, chorioangioma of placenta). Very few hydropic infants survive if delivered before 30 weeks of gestation.

2. Postnatal treatment includes

- A. Vigorous resuscitation with interventions (*e.g.*, thoracentesis, paracentesis) to remove excess fluid (see Resuscitation of hydropic infants, P. 6).
- B. Treatment of asphyxia, which is common in these infants.
- C. Diagnosis of cause of hydrops, both for management of the patient and counseling of the parents regarding risk of recurrence in future pregnancies. This may require extensive diagnostic interventions, including careful post-mortem examination, skin sample for karyotyping and full body radiographs in those who do not survive.
- D. Treatment of underlying cause

OUTCOME: Approximately 50% of fetuses with non-immune hydrops fetalis die *in utero*, and about half of the liveborn infants survive. As can be seen from the table, survival and long term outcome are dependent upon the underlying condition.

Table 1. Conditions associated with non-immune hydrops fetalis.

(Adapted from Phibbs RH: Non-immune hydrops fetalis. In *Rudolph's Pediatrics*, 20th Edition, AM Rudolph, JIE Hoffman, and CD Rudolph, Eds., Appleton & Lange, Stamford, CT, 1996, p. 256)

Hemolytic anemia

- α -thalassemia
- RBC enzyme deficiencies

Other anemias

- Feto-maternal hemorrhage
- Twin-twin transfusion (donor)

Cardiac

- Fetal arrhythmias
- Premature closure of foramen ovale
- Hypoplastic left heart
- Hypoplastic right heart
- Ebstein's anomaly of tricuspid valve
- Cardiomyopathy
- Cardiac tumors
- Premature closure of ductus arteriosus
- Other structural anomalies

Chromosomal abnormalities

- Trisomy 21, 18
- Turner syndrome

Infections

- Viral (Parvovirus B19, Herpes, CMV)
- Toxoplasmosis
- Syphilis
- Chagas Disease

Vascular malformations

- Chorioangioma (placenta, umbilical vessels)
- Liver hemangioma
- Cerebral A-V malformation
- Sacrococcygeal teratoma
- Klippel-Trenaunay syndrome

Vascular accidents

- Intracranial hemorrhage
- Thrombosis of renal veins, IVC
- Twin-twin transfusion (recipient)

Lymphatic malformations

- Pulmonary lymphangiectasis
- Cystic hygroma
- Multiple pterygium syndrome
- Noonan syndrome

Chest masses

- Cystic adenomatoid malformation
- Diaphragmatic hernia
- Pulmonary sequestration
- Intrathoracic mass

Skeletal conditions

- Asphyxiating thoracic dystrophy
- Osteogenesis imperfecta
- Chondrodysplasia

Genetic metabolic disease

- Gaucher Disease
- Mucopolysaccharidosis
- Nieman-Pick Disease
- Neonatal hemochromatosis

Fetal Hypomobility

- Arthrogyposis
- Neu-Laxova syndrome
- Pena-Shokier syndrome
- Myotonic dystrophy

CNS anomalies

- Absent corpus callosum
- Encephalocele
- Holoprosencephaly

Other

- Bowel obstruction with perforation (meconium peritonitis, volvulus)
- Infant of diabetic mother
- Prune belly syndrome
- Congenital nephrosis
- Maternal indomethacin therapy