Northern Care Alliance
NHS Group

Hydrops Fetalis / Erythroblastosis Fetalis

An information guide

Health & care information you can trust

The Information Standard Certified Member

Hydrops Fetalis / Erythroblastosis Fetalis

What is hydrops fetalis?

Hydrops fetalis is not itself a disease, but an ultrasound marker of other fetal complications. It is defined as an abnormal collection of fluid in at least two different fetal organ spaces.

These fluid-filled spaces can occur in different fetal body locations, including:

- The abdominal cavity (ascites)
- Around the heart (pericardial effusion)
- Around the lungs (pleural effusion)
- Generalized oedema or swelling of the skin (anasarca)

Two other signs of hydrops are:

- Polyhydramnios (excess amniotic fluid)
- Placental thickening

Types of hydrops

Hydrops can be divided into two major categories or types: immune hydrops (also called Erythroblastosis Fetalis) and non-immune hydrops.

Immune Hydrops (Erythroblastosis Fetalis)

This occurs when the mother's immune system attacks the blood cells of the baby. For example, a mother who has a Rh-negative blood type who is carrying a baby with a Rh-positive blood type may have an immune response that attacks and destroys the Rh-positive blood cells of the baby.

This leaves the baby anaemic or with low blood count. As the baby tries to make more blood cells to replace those being destroyed, organs that help make blood become enlarged and begin to fail.

These include the liver, kidneys and adrenal gland. The baby's heart is also affected because the low blood count causes it to have to work harder and it can eventually fail.

The blood cells produced in these other organs are usually immature and are referred to as erythroblasts. This gives us the synonym for immune hydrops, erythroblastosis fetalis.

Most cases of Rh incompatibility (Rh negative mother and Rh-positive baby) will result in a mild to moderate haemolytic (blood) problem. However, in 20 to 25 percent of cases, a more severe form develops that leads to hydrops fetalis.

The incidence of immune hydrops has decreased greatly since the introduction of Rhesus Immunoglobulin or Anti-D in the 1960's. Rhesus Immunoglobulin or Anti-D is a shot given to the mother during pregnancy to keep her body from attacking her baby's body.

Non-immune Hydrops

Non-immune hydrops makes up about 75 to 90 percent of all cases of hydrops seen today. Non-immune hydrops can be the symptom of a number of congenital conditions.

Just a few of the many known causes of hydrops include:

- Other conditions causing low blood count in the foetus including fetal bleeding (haemorrhage), problems with fetal blood cell production and genetic disorders that cause the blood cells to be more easily destroyed. These conditions make up about 10 to 27 percent of cases)
- Certain infections the mother catches in pregnancy and passes to baby (congenital Infections) such as syphilis, cytomegalovirus, and parvovirus (approximately 8 percent of cases)

- Genetic syndromes and metabolic disorders (approximately 10 percent of cases)
- Fetal birth defects that affect the heart function. These include defects that cause push on the heart or make the heart work harder. fetal tumours or masses, such as diaphragmatic hernia, cystic adenomatoid malformation, sacrococcygeal teratomas, or polycystic kidneys (about 10 percent of cases)
- Fetal heart conditions or birth defects (approximately 40 percent of cases) such as problems with the placenta, umbilical cord or the mother's body. In approximately 5 to 8 percent of the cases, doctors cannot identify a cause. These are classified as idiopathic cases, meaning the hydrops is the result of an unknown cause.

The incidence of immune hydrops/erythroblastosis fetalis has decreased dramatically since the 1960's due to the introduction of Rh immune globulin or Rhesus Immunoglobulin or Anti-D as well as improvements in treatment options.

The incidence of non-immune hydrops is hard to calculate because many cases will result in fetal death and/or miscarriage of the baby, and some will get better on their own, especially when hydrops is the result of an infectious process. The range of incidence is reported at 1 in 1,500 to 4,000 deliveries.

Prenatal diagnosis of Hydrops Fetalis

Doctors diagnose hydrops prenatally using an ultrasound. If there is abnormal or increased fluid collection in at least two fetal body spaces, the diagnosis can be made.

If fluid accumulation only occurs in one area, doctors cannot make the diagnosis of hydrops. Instead, an increased amount of fluid is simply noted in that area. To differentiate immune from non-immune hydrops involves a blood test of the mother that looks for antibodies associated with blood-group incompatibility.

Other tests after diagnosis

Because heart anomalies or disorders are a common cause of non-immune hydrops, we highly recommend a fetal echocardiogram (a specialized ultrasound of the heart) to look for any structural abnormalities or abnormal heart rhythms.

Other tests may be performed to look for infections, genetic disorders or other congenital issues that have been linked to the development of hydrops. A genetic counsellor may be recommended to assist in this evaluation.

If your doctor believes your baby has hydrops, he or she will refer you to a foetal medicine specialist (a specialist that handles high-risk pregnancies).

How does hydrops fetalis affect my baby?

Hydrops fetalis is a symptom of an underlying problem with your baby.

Hydrops usually will rapidly become fatal for your baby if left untreated. An infant/foetus with hydrops is severely compromised. The earlier in the pregnancy the diagnosis is made, the worse the prognosis is. Some babies with hydrops may even die before they are born.

If your child is diagnosed with immune hydrops, urgent evaluation and possible treatment is necessary to prevent stillbirth or fetal death. The treatment will partly depend upon the gestational age at diagnosis and the determination of underlying cause.

There is a 60 to 90 percent fetal mortality associated with non-immune hydrops. This mortality rate is dependent on the underlying cause. Many of the underlying congenital anomalies (birth defects) have an extremely high mortality rate in and of themselves.

Sometimes, no cause is ever discovered, but the foetal medicine specialist will be aggressively working to try to find out why your baby has developed hydrops. Some causes can be treated, so the doctor will especially be looking to see if there is a treatable cause. Generally, the earlier in gestation that fetal hydrops is seen, the poorer the prognosis is.

Complications with Hydrops

- Babies born with hydrops are very swollen and have a large, round abdomen due to the fluid collection in the abdominal cavity.
- Often, these infants will have severe respiratory distress or breathing problems due to a variety of causes.
- Many have lungs that did not develop completely. This is thought to be a result of limited space in the chest due to a swollen liver and a diaphragm that is pushed upward from the fluid that collected in the abdomen.
- The polyhydramnios (larger than normal volume of amniotic fluid) may cause increased risks of preterm labour and birth.
- Many of these babies have fluid collection around the lungs and/or heart, which complicates already compromised breathing and heart function.
- Another common complication for these infants is severe low blood sugar (hypoglycaemia), which is thought to be the result of altered function of cells in the pancreas. The cause of this altered function is not well understood. Hypoglycaemia (low blood sugars) put these babies at risk for seizures, apnoea (difficulty breathing) and possible brain injury.
- If the underlying cause of the hydrops can be linked to anaemia (low blood count), the baby is at an increased risk of developing hyperbilirubinemia and heart failure. This problem can become severe very quickly and lead to neurologic injury or fetal death.

How does hydrops fetalis affect my pregnancy?

If your obstetrician suspects a problem with your baby, you will likely be referred to a foetal medicine specialist.

This doctor will perform a targeted ultrasound to look at all of your baby's anatomy (the body and its systems, such as heart, lungs, abdomen, head, brain, arms, legs) and assess the location and amounts of fluid accumulation.

The foetal medicine specialist perinatologist will assess you and your baby for:

- The causes hydrops
- The progression of hydrops
- Possible treatment options
- Overall well-being including maternal well-being and baby's growth and activities.

Other specialists you may see are:

- A neonatologist, a specialized paediatric doctor who will care for your baby after delivery
- A genetic physician or genetic counsellor who will be evaluating your family health history for possible inheritable conditions that would cause hydrops
- Any other specialists as necessary to address underlying issues or abnormalities uncovered by the overall workup.

Mirror Syndrome

In up to 50 percent of pregnancies complicated by hydrops, a condition call mirror syndrome (or Ballentine syndrome) will develop.

This condition is thought to be a variant of a pregnancy-related blood pressure disease called preeclampsia and may result in life-threatening maternal hypertension (elevated blood pressures) or seizures. The only treatment for mirror syndrome is immediate delivery of the baby

How do you treat hydrops fetalis?

Treatment for hydrops depends on the determined cause, if any. Otherwise, the care of a baby born with complications of hydrops is primarily focused on comfort measures, if no therapy or treatment is available.

When the underlying cause is determined to be complications of low blood counts, one potential therapy is transfusion of blood products to the foetus, just as would be done for an adult with critically low blood. This procedure is called an In Utero Transfusion (IUT).

An IUT involves an amniocentesis where a needle is guided by ultrasound into the umbilical cord of the baby so that the blood can be sampled for testing and new blood may be transfused to restore the blood levels. Blood levels are expressed as haematocrit and the initial procedure goal is a haematocrit of 20-25 percent.

A repeat procedure is likely necessary to achieve a final haematocrit of 45-50 percent within 48-72 hours. (Then, transfusions are done at two- to three-week intervals, with the last one done at 34 to 35 weeks gestation. These babies should improve before birth.

The treatment protocol for other cases of non-immune hydrops is aimed at the underlying cause. If the underlying cause is known, the benefits and risks of the treatment will be weighed against likelihood of survival.

Babies who have not shown improvement before birth should be delivered at a tertiary care centre with a level III Neonatal Intensive Care Unit (NICU) capable of extensive evaluation and complex care of the compromised newborn immediately after delivery.

Because hydrops of uncertain causes are associated with such poor outcomes, you should expect your team of doctors to have very open and frank conversations with you and your family about options of palliative care versus aggressive resuscitation, prior to

delivering your baby. A neonatologist will work with you to develop a plan of care for your baby once he or she is born.

For infants without any specific diagnosis, we offer supportive treatment as we give the baby time to reabsorb all the oedema (fluid from swelling). Often, there are complications like infection, and sometimes the baby is unresponsive to the support provided.

Delivery method

The best method for delivery of babies with hydrops is uncertain and a Caesarean section may be advised.

At delivery, most babies who are being aggressively resuscitated will require endotracheal intubation (the placement of a special tube in the windpipe) to help with breathing.

Placing the breathing tube can be difficult because of the swelling from the excess fluid in the baby's body. High-frequency ventilation and high airway pressure settings may also be required to provide the baby with enough oxygen.

To help your baby breathe, we may need to place tubes in the chest to help remove fluid from the abdomen and around the lungs.

We may also place special IV lines. Normally, in the umbilical cord, there are two arteries and one vein. We also put an umbilical artery catheter in one of the arteries of the umbilical cord. With this special line, we can:

- Provide fluids
- Give medications
- Monitor blood pressure
- Remove blood for blood tests

What about surgery?

Under some circumstances, surgical treatment of the suspected cause of hydrops may be discussed.

For example, congenital cystic adenomatoid malformation and bronchopulmonary sequestration may be responsive to surgical treatment, but will only be considered if hydrops is diagnosed at certain gestational ages.

You can discuss this option with your foetal medicine specialist.

Will I be able to help care for my baby after birth?

Yes. Please ask your baby's nurse about ways to interact with and care for your baby.

These babies are typically very ill at birth and will require aggressive treatments. To help in your understanding about what is being done and why, ask questions about the treatments and procedures and visit often with your baby.

If you had planned to breastfeed your baby, you can begin to pump and freeze your breast milk while you are still in the hospital. A lactation consultant can assist in answering your questions.

Your milk will be frozen and stored in the Neonatal Intensive Care Unit until your baby is ready for it. The NICU has breast pumps and private rooms available to you when you are visiting.

You can bring in pictures, small toys, booties and blankets for your baby while he or she is in the NICU.

When can my baby go home?

If the treatment leads to a reversal and the hydrops resolves, your infant may eventually go home. However, for an infant with non-immune hydrops, the prognosis is very poor.

Your baby must be able to eat enough to maintain and gain weight and breathe effectively by himself or herself before going home. It is important to remember the complication of hydrops has a poor prognosis of survival except for those cases that have a definite cause with established fetal treatment.

What is my baby's long-term prognosis?

Long-term prognosis is guarded.

These babies are critically ill even if they do survive to birth. Of the foetuses diagnosed prenatally, only about 20 percent survive to delivery.

Of this number, approximately half will survive the neonatal period. Long-term survival for those that make it through the newborn period is based on the underlying cause of the hydrops. The data currently shows an optimistic outlook for those babies who do survive.

If English is not your first language and you need help, please contact the Interpretation and Translation Service

Jeśli angielski nie jest twoim pierwszym językiem i potrzebujesz pomocy, skontaktuj się z działem tłumaczeń ustnych i pisemnych

اگر انگریزی آپ کی پہلی زبان نہیں ہے اور آپ کو مدد کی ضرورت ہے تو ، براہ کرم ترجمانی اور ترجمہ خدمت سے رابطہ کریں

Dacă engleza nu este prima ta limbă și ai nevoie de ajutor, te rugăm să contactezi Serviciul de interpretare și traducere

ইংরাজী যদি আপনার প্রথম ভাষা না হয় এবং আপনার সাহায্যের প্রয়োজন হয় তবে অনুগ্রহ করে দোভাষী এবং অনুবাদ পরিষেবাটিতে যোগাযোগ করুন

إذا لم تكن الإنجليزية هي لغتك الأولى وتحتاج إلى مساعدة ، فيرجى الاتصال بخدمة الترجمة الشفوية والتحريرية

: 0161 627 8770

: interpretation@pat.nhs.uk

To improve our care environment for Patients, Visitors and Staff, **Northern Care Alliance NHS Group** is Smoke Free including buildings, grounds & car parks.
For advice on stopping smoking contact the Specialist Stop Smoking Service on 01706 517 522

For general enquiries please contact the Patient Advice and Liaison Service (PALS) on 0161 604 5897

For enquiries regarding clinic appointments, clinical care and treatment please contact 0161 624 0420 and the Switchboard Operator will put you through to the correct department / service

The Northern Care Alliance NHS Group (NCA) is one of the largest NHS organisations in the country, employing 17,000 staff and providing a range of hospital and community healthcare services to around 1 million people across Salford, Oldham, Bury, Rochdale and surrounding areas. Our Care Organisations are responsible for providing our services, delivering safe, high quality and reliable care to the local communities they serve.

The NCA brings together Salford Royal NHS Foundation Trust and the hospitals and community services of The Royal Oldham Hospital, Fairfield General Hospital in Bury, and Rochdale Infirmary (currently part of The Pennine Acute Hospitals NHS Trust).

www.facebook.com/NorthernCareAllianceNHSGroup

www.linkedin.com/company/northern-care-alliance-nhs-group

Northern Care Alliance NHS Group (NCA) @NCAlliance_NHS

Date of publication: April 2020
Date of review: April 2020
Date of next review: April 2022
Ref: PI(WC)448

www.pat.nhs.uk
www.northerncarealliance.nhs.uk