The Childhood Liver Disease Research Network strives to provide information and support to individuals and families affected by liver disease through its many research programs.

**IDIOPATHIC NEONATAL HEPATITIS**

**What is Idiopathic Neonatal Hepatitis?**

Idiopathic neonatal hepatitis (INH) or Idiopathic neonatal cholestasis is a general term for inflammation of the liver. It occurs shortly after birth in newborns (less than 3 months of age). The specific cause of the problem cannot be identified. Neonatal cholestasis (poor bile flow in a young baby) can be caused by several things. Sometimes the problems are genetic, meaning passed on to children by their parents at birth, such as in Alagille syndrome (ALGS) and progressive familial intrahepatic cholestasis (PFIC). Other times the problems can be caused by infections like CMV, congenital syphilis, echovirus, and some herpes viruses. The classic hepatitis viruses (A, B, and C) are less common causes. It is also possible for these liver problems to be caused by metabolic diseases like α1-antitrypsin deficiency (Alpha-1), cystic fibrosis (CF), respiratory chain defects, and fatty acid oxidation defects. When doctors check for all these things but still can’t find the reason for the inflammation, they diagnose the problem as INH.

**What are the symptoms?**

The symptoms of idiopathic neonatal hepatitis (INH) can vary greatly from one person to another. Infants with INH may have jaundice as their only symptom. This usually occurs in the first two weeks of life and can last up to the third month of life. Infants may also have dark urine, an enlarged liver, and greyish stools from the lack of bile. Other symptoms may also include poor growth, irritability, and itching.

**How is it diagnosed?**

The greatest challenge in diagnosing INH is in figuring out how it is different from other neonatal liver diseases with known causes. Doctors will run tests to rule out other metabolic, infectious, and genetic causes of the liver problems they see. These include blood tests, scans of the liver and bile ducts, and possibly metabolic or genetic testing and liver biopsy. If biopsy results show that the liver cells are enlarged but no specific cause is found, doctors might diagnose INH.
How is it treated?

Since there is no known cause in INH, treatment is focused on managing symptoms and providing good nutritional support. This includes medications to stimulate bile flow, predigested formulas, and extra vitamins A, D, E, and K.

What is the outlook for infants diagnosed with INH?

Approximately 80% of infants diagnosed with INH recover fully from the condition. As doctors have learned more about INH over the years, and have figured out more specific causes of it, the number of cholestatic infants diagnosed with INH has decreased. In the future we expect that number to continue to decrease as doctors learn more about INH.

Does the ChiLDReN Network have any studies that include patients with INH?

Yes. The ChiLDReN Network currently has one study that includes patients with INH.

The PROBE study is a natural history study that includes patients with INH. A natural history study is aimed at acquiring information and data that will provide a better understanding of rare conditions. Participants will be asked to allow study personnel to obtain information from medical records and an interview, and to collect blood, urine, and tissue samples when clinically indicated, in order to understand the causes of these diseases and to improve the diagnosis and treatment of children with these diseases. All of the information obtained in these studies is confidential and no names or identifying information are used in the study.

**PROBE:** A prospective study of infants and children with cholestasis.

*Eligibility:* Infants up to 6 months of age that have been diagnosed with cholestasis (direct hyperbilirubinemia).

[ClinicalTrials.gov Study NCT00061828](https://clinicaltrials.gov/ct2/show/NCT00061828)

Are there any organizations or foundations that help families dealing with bile acid defects?

Yes. The ChiLDReN Network works with numerous groups that support patients and families who are dealing with rare liver diseases. Please [click here](https://www.childrens.org) to go to that page on our website (Information for Families). You will see the list of groups and information about them.