



The Childhood Liver Disease Research and Education 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) is a general term for inflammation of the liver that occurs shortly after birth in newborns (less than 3 months of age) for which a specific cause cannot be identified. Neonatal hepatitis can have one of a number of causes including metabolic, infectious, and genetic causes. Metabolic diseases include α 1-antitrypsin deficiency, cystic fibrosis, neonatal iron storage disease, respiratory chain defects, and fatty acid oxidation defects. Infectious causes include congenital syphilis, echovirus, and some herpes viruses. The classic hepatitis viruses (A, B, and C) are less common causes. There are also a number of less common genetic defects, such as Alagille syndrome and progressive familial intrahepatic cholestasis. In Idiopathic Neonatal Hepatitis, however, the cause of inflammation remains unknown.

What are the symptoms?

The symptoms of idiopathic neonatal hepatitis can vary greatly from one individual to another. Infants with INH may have jaundice as their only symptom; usually in the first two weeks of life developing up to the third month of life. There may also be the presence of dark urine, hepatomegaly, and acholic stools. Other symptoms may also include poor growth, irritability, pruritis.

How is it diagnosed?

The greatest challenge in diagnosing idiopathic neonatal hepatitis is in differentiating it from other neonatal liver diseases with known causes. INH is diagnosed when tests run to rule out other metabolic, infectious, and genetic causes are complete. This is done through blood tests, hepatobiliary scans, possible metabolic or genetic testing, and possibly liver biopsy. On biopsy, the hepatocytes are enlarged and no cause is found.

How is it treated?

Since there is no known cause in idiopathic neonatal hepatitis, treatment is focused on symptom management and 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 idiopathic neonatal hepatitis recover fully



from the condition. Over the years, as more specific causes of this syndrome are diagnosed, the number of cholestatic infants given the diagnosis decreases. In the future it is expected that this number will continue to decrease as more specific causes are described.