### Liver Disease in Infants and Children





Canadian Liver Foundation Fondation canadienne du foie Bringing liver research to life Donner vie à la recherche sur le foie

### Jaundice can be an early warning sign of liver disease.

The most common sign of liver disease in newborns is jaundice (a yellowing of the skin and whites of the eyes).

Other symptoms and signs include:

- dark urine and/or pale stools;
- an enlarged abdomen;
- vomiting;
- failure to grow or develop normally.

### If my baby is jaundiced, does it mean he/she has liver disease?

Jaundice develops when blood contains an excess of bilirubin — a pigment produced during the normal breakdown of red blood cells.

Many babies have "newborn jaundice" lasting three to five days after birth because the liver is not fully developed. The newborn liver cannot remove bilirubin from the blood effectively enough. With newborn jaundice, the colour of the urine and stools is normal.

Breastfeeding may also prolong jaundice in some newborns. "Breast milk jaundice" develops gradually about one week after birth and can last as long as four to six weeks if the infant is breastfeeding.

Both "newborn" and "breastmilk" jaundice usually clear up on their own and are not an indication of liver disease. However any jaundice that does not clear up after 14 days of life should be checked by your doctor.

### What can cause liver disease in infants and children?

There are many causes of liver disease in infants and children. Some still remain unrecognized. Liver disease in children can be grouped into the following categories:

- 1. Viral infection of the liver
- **2.** Decreased or impaired bile flow from the liver
- 3. Inherited or metabolic liver diseases

### What is viral infection of the liver?

Viruses which can cause liver disease in infants include cytomegalovirus (CMV, a common virus that can infect almost anyone), rubella (German measles) and hepatitis B and C viruses.

There is a high risk that pregnant women who are infected with the hepatitis B virus will pass the virus on to their babies around the time of birth. That is why screening of pregnant women and immunization of newborns against the hepatitis B virus is extremely important. The risk of passing the hepatitis C virus from mother to child is much lower (4–7%).



# What conditions can lead to decreased bile flow from the liver?

**Biliary atresia** begins in early infancy and causes damage to the liver due to the destruction of ducts which carry bile from the liver to the small intestine. Surgery can restore bile flow from the liver to the intestine but this surgery (known as Kasai procedure) has the highest likelihood of success if it is performed within the first 8–12 weeks of life.

**Choledochal cyst** is dilatation or enlargement of the bile duct outside the liver due to abnormal liver development. This condition can be diagnosed by ultrasound before birth but is usually found in early infancy. Corrective surgery is required.

Children with **Allagile syndrome** usually have a liver disease due to a progressive loss of the bile ducts within the liver over the first year of life. This leads to a buildup of bile in the liver, causing damage to liver cells. Treatment of Alagille syndrome is based on trying to increase the flow of bile from the liver, maintain normal growth and development, and prevent or correct any of the specific nutritional deficiencies that often develop.

### What are metabolic liver disorders?

There are many metabolic liver diseases that affect infants and children. Most are rare and have a genetic basis. The common ones include:

Alpha 1-antitrypsin deficiency causes liver disease, and later on lung disease, because of a deficiency of a protein made in the liver. One of its functions is to protect the lungs from enzymes produced by inflammatory cells. These enzymes are aimed at killing bacteria but can also cause damage to lung tissue if not properly controlled.

Galactosemia occurs in babies who cannot metabolize simple milk sugar (galactose) found in breast milk and most baby formulas. They may develop vomiting and dehydration. Treatment is dietary.

Hereditary tyrosinemia is due to a defect in an enzyme metabolizing a protein building block (tyrosine). Since tyrosinemia is particularly common in northern Quebec, doctors test the blood of every newborn baby in the province thus allowing early detection and appropriate treatment.

#### What tests are performed to diagnose liver disease in infants?

For an accurate diagnosis, doctors will order blood tests for bilirubin levels, and other measurements of liver function, and will look for any evidence of infection and metabolic diseases. An ultrasound examination may be used to examine the liver and bile ducts. Finally, doctors may do a liver biopsy (take a sample of tissue) to inspect the liver tissue more closely.



#### If my infant or child is diagnosed with liver disease, can they be treated?

The treatment varies with the cause. In general, doctors may prescribe a special baby formula as well as vitamin supplements. Some metabolic diseases however, require specific diets or drug treatments. For some diseases, such as biliary atresia, surgery on the liver is required. Please consult your healthcare provider regarding specific treatments and diets your child may require.



#### Is a liver transplant necessary?

In many cases, childhood liver diseases can be managed or controlled through diets, medications or other interventions. If the liver is too damaged to function effectively, a liver transplant may be necessary. This is sometimes necessary in the first year of life.

### What are the warning signs of continuing liver disease?

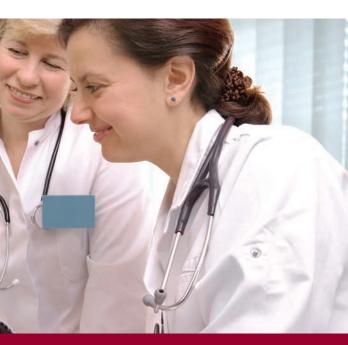
Jaundice which does not disappear is a sign of ongoing liver disease and generally indicates severe liver problems. In an infant who appears to recover from a liver disorder, the return of jaundice is a sign of ongoing disease. Your doctor should check your child at least yearly.



### **Does liver disease recur in the same family?**

Viral infections and biliary atresia are extremely unlikely to affect more than one child in a family. Metabolic disorders of the liver are usually inherited and therefore may occur in more than one child. Older children in the family should be checked for these diseases, and infants should be followed carefully after birth.

Prenatal diagnosis is available for some of these disorders, and genetic counselling may be useful.



## CANADIAN LIVER

1 in 4 Canadians may be affected by liver disease, including everyone from newborn babies to older adults.

Founded in 1969 the Canadian Liver Foundation (CLF) is the only national charity in Canada focused on liver health, and the main source of non-profit funding for liver health research.

Today, we are *bringing liver research to life* by raising funds to promote liver health, improve public awareness, fund research and provide support to individuals affected by liver disease.

To support liver research visit **liver.ca/donate** 

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