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, as well as evidence of infection and metabolic diseases. An ultrasound examination of the liver and hepatobiliary scan may then 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 baby is diagnosed with liver disease, can it 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, surgery on the liver is required.

Is a liver transplant necessary?
Some severe liver disorders will require a liver transplant in order to be cured. This is rarely necessary in the first months of life.

What are 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 recovers 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?
Neonatal hepatitis from viral infection 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 subsequent babies should be followed carefully after birth. Prenatal diagnosis is available for some of these disorders, and genetic counselling may be useful.

Canadian Liver Foundation
Since its inception in 1969, the Canadian Liver Foundation (CLF) has been committed to reducing the incidence and impact of liver disease by providing support for research and education into the causes, diagnosis, prevention and treatment of the more than 100 diseases of the liver. This is a rapidly changing field of medicine. Information in this brochure is current for January 2004.

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Canadian Charitable Registration No. 10686 2949 RR0001
Ce dépliant est également disponible en français.
When you see a yellow traffic light, you know that you must proceed with caution. It’s the same thing when you see jaundice in your baby.

Jaundice can be an early warning sign of liver disease.

As a parent, you need to proceed with caution. Help protect your baby by learning to recognize the early warning signs of liver disease.

The most common sign of liver disease in new-borns 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. This type of jaundice usually clears up by itself and is not an indication of liver disease.

Breastfeeding may prolong jaundice in some new-borns. “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. This jaundice will usually disappear on its own but it should be checked by your doctor.

What are the causes of liver disease in infants?

Liver disease in infants is usually the result of:
1. viral infection of the liver;
2. blockage of the flow of bile from the liver (usually a disease called “biliary atresia”);
3. inherited or metabolic liver diseases.

What is viral infection of the liver?

Viruses which can cause liver disease in infants include cytomegalovirus (CMV), 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 block bile flow from the liver?

- **Biliary atresia** is a blockage of the duct system that carries bile from the liver to the intestine. Surgery to restore bile flow from the liver to the intestine (Kasai procedure) is more likely to be effective the earlier it is performed.

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

- **In Allagile syndrome and certain other conditions**, the smallest bile ducts inside the liver are abnormal and reduced in number.

- **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.

What are metabolic liver disorders?

These are numerous, but most are rare. Most 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 an enzyme made in the liver.

- **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.