Newborn cholestasis: a liver gone haywire

Neonatal cholestasis, commonly known as newborn “jaundice”, afflicts one in every 2,500 babies born. In reality, the term “cholestasis” includes all disorders linked to abnormal bile flow or formation. Newborns are especially vulnerable to this disease because their process of bile production is still undergoing maturation. If left untreated, a large percentage of newborns face serious complications, or even death.

In their article, Muriel Girard and Florence Lacaille¹ cover the remarkable process of bile production, showing how deregulation of these steps can occur in newborns. They review the various origins of cholestasis, including genetic and individual factors.

The surprise comes when the authors reveal the diverse conditions that can result in cholestasis, such as mitochondrial disorders, infections, or even intravenous feeding. The damaging feature of cholestasis - decreased bile flow - causes an increase in serum bile salts, which can damage the liver. Another consequence for the baby is poor absorption of fats and fat-soluble vitamins, in turn leading to rickets, neuropathy, and other disorders of vitamin deficiency.

What makes cholestasis a potentially deadly condition in young babies? The hidden danger is when the bile duct connecting the liver and small intestine is blocked or absent, a condition known as biliary atresia. Girard and Lacaille reveal that “Biliary atresia is the most frequent cause of neonatal cholestasis and accounts for almost 50% of cases.” Because of the high likelihood of a blocked or missing bile duct, the authors advise clinicians to err on the side of caution when making diagnoses.

As with many diseases, early diagnosis is key to improving the odds for survival. Liver transplantation is the main treatment for a severely blocked or missing bile duct. Only one drug, ursodeoxycholic acid, is available to treat cholestasis. However, Girard and Lacaille remain hopeful that a better understanding of this disease will pave the way for less invasive treatment options, such as gene therapy.

References