New Home Screening Program for Liver Disease in Newborns

By Lubna Ekramoddoullah

Parents in British Columbia (BC) are being asked to check their newborn's stool colour in an effort to detect early signs of a rare but fatal liver disease, as part of the Biliary Atresia Home Screening Program launched by Perinatal Services BC, an agency of the Provincial Health Services Authority.

Biliary atresia is a liver disease resulting from blockage of the bile duct, which prevents bile from leaving the liver, resulting in damage and scarring that can lead to death by the age of two, if not treated. The disease begins to affect newborns in the first month of life. While it is normal to see jaundice in the first few days after birth, some babies may have jaundice that lasts longer than two weeks, as well as pale yellow, chalk-white, or clay-coloured stools, an indication that very little or no bile is reaching the intestine.

After the birth of the baby and before mom and baby leave the hospital, parents are given a stool colour card that contains photos of normal and abnormal infant stool colours. Mothers who have a home birth are given the stool colour card by their midwives. Parents are asked to check their newborn's stool colour against the colour card every day for the first month after birth. If they see an abnormal stool colour, they are to contact the Biliary Atresia Home Screening Program.

The preferred treatment is the Kasai procedure, a surgical method that re-establishes bile flow from the liver to the intestine by joining the two. The diseased bile duct is removed and a small segment of the small intestine is attached to the liver at the spot where bile is expected to drain.

The effectiveness of this surgery depends on timing. If the Kasai procedure is performed in the first two months of life, it has an 80 per cent chance of success. But after three months, it drops to 20 per cent. If the procedure is unsuccessful, a liver transplant is required.

That is why detecting biliary atresia early is so important. There is no single blood test for biliary atresia, so stool colour is the main tool for early detection.

"We need to build awareness among parents and healthcare providers about the need to look at stool colour as a disease indicator," says Dr. Richard Schreiber, Director, BC Pediatric Liver Transplant Program, BC Children's Hospital and Professor of Pediatrics, University of British Columbia. "Poor outcomes due to late diagnosis and surgery of infants three months of age or older remain a problem throughout Canada and elsewhere in the world."

The Biliary Atresia Home Screening Program is the first of its kind in Canada. The program is based on best practices in Taiwan, as well as research conducted in BC and Quebec involving over 9,500 families. The stool colour card used in BC also has a Quick Response code, so parents can use their smartphones to sign up for weekly text or email reminders to check their baby's stool. The reminders are available in 12 languages, including Punjabi, Arabic and Persian.

"BC is the first province in Canada to implement this unique type of home screening program," says Kim Williams, Provincial Executive Director, Perinatal Services BC. "There are no blood tests or samples to collect, and it is family-centered — done at home by parents or other family members. Parents can feel empowered because they are taking a proactive role in identifying a life-threatening disease and improving the health of their newborns."

"It took many visits to our family doctor over the course of the first three months of our newborn son's life to ultimately receive the biliary atresia diagnosis," says Mitzi Mogden-Dupuis. "Prolonged jaundice and stunted growth were clear indicators of illness, but due to a partially functioning bile duct, his stool was still light brown and yellow in colour. We explored several possibilities, but the most important piece of the puzzle was absent simply because we did not have a reference tool to indicate and report where he registered on the scale of healthy versus unhealthy stool colour. It was not until we presented a physical stool sample that our doctors were able to establish the cause and severity of his illness. Within hours of this evidence, and just inside the window of probability to have a successful outcome through the Kasai procedure, our son was admitted into the hospital. We now have a thriving five-year-old son and consider our family incredibly fortunate to have caught this in time. However, a resource such as the stool colour card could have dramatically improved the timeline of achieving the operation necessary to save our son's life."

Facts about Biliary Atresia

- The cause of biliary atresia is not known; it affects only newborns and it is not preventable.
- Biliary atresia occurs in one out of every 19,000 births per year in Canada and 1/10,000 births per year in BC (about three to five cases).
- While rare, it is the most common cause for cirrhosis- and liver disease-related death among children, accounting for more than 60 per cent of all pediatric liver transplants.
- In Canada, only eight per cent of infants with biliary atresia have the Kasai procedure done at 30 days of age or younger.
- The Kasai procedure is named after Dr. Morio Kasai, the Japanese surgeon who developed the procedure in 1951. The diseased bile duct is removed and a small segment of the small intestine is attached to the liver at the spot where bile is expected to drain.

For more information about the Biliary Atresia Home Screening Program, visit Perinatal Services BC at www.perinatalservices.bc, click Screen Programs, and then Biliary Atresia.