Neurodevelopment in young children with biliary atresia

Infancy and early childhood is a time of critical neurodevelopment. It is also an important time for monitoring that development, particularly in children with chronic illnesses.

The most common cause of chronic progressive liver disease in children is biliary atresia (BA), with symptoms always appearing shortly after birth. BA is the most common reason for the need for liver transplantation in children. Infants with BA are born with obstructed bile ducts, causing bile to be trapped inside the liver. The primary treatment is surgery with a “Kasai procedure” or hepatoportoenterostomy (HPE) performed as early as possible to try to achieve bile drainage.

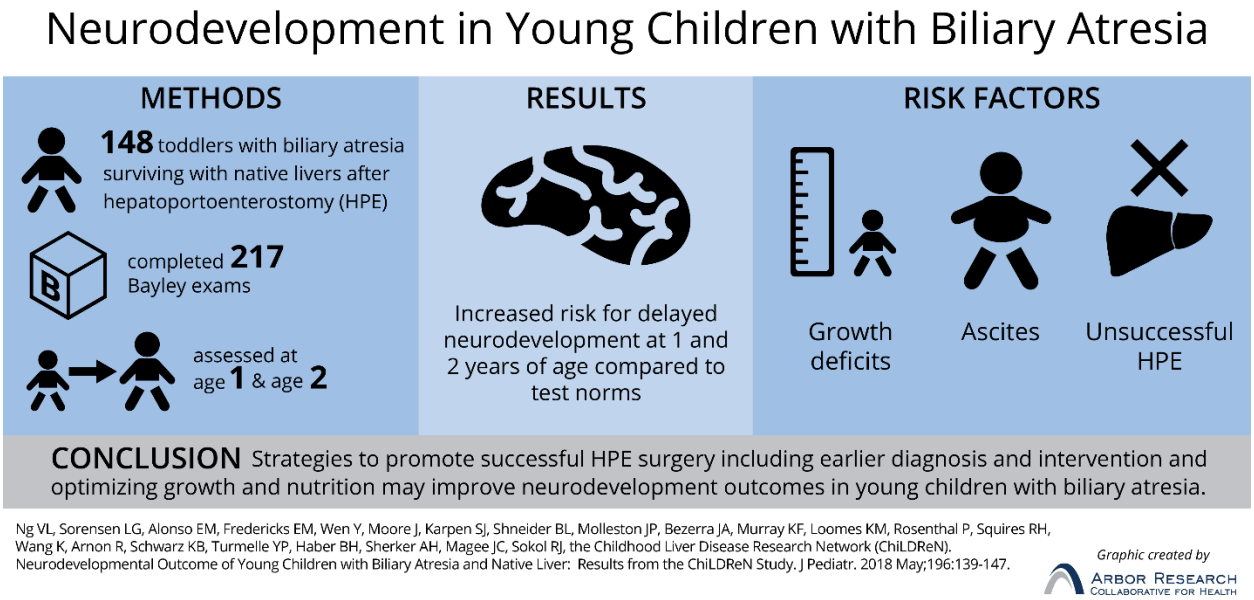
Using data from the Childhood Liver Disease Research Network (ChiLDReN), a National Institutes of Health-funded consortium, researchers assessed neurodevelopment among children with BA and evaluated variables that could predict impaired neurodevelopment.

They found that young children with BA surviving with native livers after HPE are at increased risk for delayed neurodevelopment at both 1 and 2 years of age compared to test norms. Within this group, children with unsuccessful HPE are over 4 times more likely to have impaired neurodevelopment compared to those with successful HPE.

Specific risk factors for impaired neurodevelopment at 1 year of age included growth deficits and ascites (fluid buildup in the abdomen). An unsuccessful HPE was identified as a risk factor at age 2 years.

“It is crucial to identify young children with biliary atresia at risk for delayed neurodevelopment even if they do not meet indications for liver transplantation,” said lead author Dr. Vicky Ng. “If we can mitigate these risks as soon as possible via targeted and tailored interventions, the goal is to enhance functional outcomes for these children.”

These findings suggest that targeting innovations and strategies to promote successful HPE surgery including earlier diagnosis and intervention and optimizing growth and nutrition may improve neurodevelopment outcomes in young children with biliary atresia.



Ng VL, Sorensen LG, Alonso EM, Fredericks EM, Wen Y, Moore J, Karpen SJ, Shneider BL, Molleston JP, Bezerra JA, Murray KF, Loomes KM, Rosenthal P, Squires RH, Wang K, Arnon R, Schwarz KB, Turmelle YP, Haber BH, Sherker AH, Magee JC, Sokol RJ, the Childhood Liver Disease Research Network (ChiLDReN). Neurodevelopmental Outcome of Young Children with Biliary Atresia and Native Liver: Results from the ChiLDReN Study. J Pediatr. 2018 May;196:139-147.

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CITATION:
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