

LONG-TERM NEURODEVELOPMENTAL OUTCOME IN CHILDREN WITH BILIARY ATRESIA.

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Aim: Little is known about the long-term neurodevelopmental sequelae of biliary atresia (BA). The aim of this pilot study was to get more insight in the long-term neurodevelopmental outcome in BA patients at school-age.

Methods: An observational cohort study of BA patients between 6-12 years who were originally diagnosed in our center (n=10). We assessed neurodevelopmental outcome using motor-, and cognitive tests for the patient and questionnaires for the parents. We compared patient results with the age-specific Dutch norm using one-sample chi-square analyses.

Main results: Nine of ten (90%) children underwent a Kasai porto-enterostomy and eight of ten (80%) had a LTx. In 9 children, motor domains were affected; abnormal fine motor skills, ball skills and balance (resp. 78%, 78% and 44%, $p<0.05$). Children showed lower performance intelligence, attention abilities and perceptual ability (resp. 40%, 33%, 56%, $p<0.05$) compared with the Dutch norm population. The children showed behavioral problems; total behavioral problems, internalizing problems, and problems in attention, hyperactivity and metacognition (resp. 33%, 44%, 33%, 44%, 44%, $p<0.05$), and 56% of the children had learning problems.

Conclusion: This pilot study suggests impaired motor and cognitive outcome in BA patients at school-age, as well as behavioral problems. The data warrant an extension of the study population in combination with investigation of possible factors associated with impaired outcomes. Early identification of subgroups at risk for neurodevelopmental impairments may allow targeting interventions at an early stage, to improve long-term outcome.