



Title	Comprehensive analyses of neurodevelopmental outcomes and quality of life of children with biliary atresia
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論文内容の要旨  
Synopsis of Thesis

氏名 Name	里村 宜紀
論文題名 Title	Comprehensive analyses of neurodevelopmental outcomes and quality of life of children with biliary atresia (小児胆道閉鎖症患者における神経発達およびQOLの包括的な解析)
論文内容の要旨	
〔目的(Purpose)〕	
To holistically evaluate neurodevelopmental outcomes and quality of life (QOL) of Japanese patients with biliary atresia (BA) and to investigate the factors associated with the outcomes.	
〔方法ならびに成績(Methods/Results)〕	
Methods	
This study enrolled patients with BA aged 5-18 years who visited Osaka University Hospital in 2021. Neurodevelopmental assessments were performed to evaluate intellectual ability, cognitive functions and adaptive skill levels. Furthermore, emotional and behavioral issues, characteristics of attention deficit hyperactivity disorder, and QOL were concomitantly assessed in the same cohort. Biochemical and social factors associated with the results were examined.	
Results	
Fifty-three patients, with a median age of 11.2 years were included in the analyses. Patients with BA had a significantly lower Full-Scale Intelligence Quotient or developmental quotient (FSIQ/DQ) score and Vineland Adaptive Behavior Scale (VABS) composite score than the general Japanese population. Household education level and short stature were associated with low and borderline FSIQ/DQ and VABS composite scores, respectively. Among patients with low and borderline FSIQ/DQ scores, those with average or high VABS composite scores received significantly less neuroeducational care than those with low and borderline VABS composite scores. Despite the low FSIQ/DQ and VABS composite scores, the total QOL scores were higher than those of the general population.	
〔総括(Conclusion)〕	
Patients with BA had intellectual and behavioral impairments. Notably, patients with intellectual impairments are overlooked and not followed up, especially if adaptive skills are maintained.	

## 論文審査の結果の要旨及び担当者

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## 論文審査の結果の要旨

この論文は、日本人胆道閉鎖症患者の神経発達の予後とQOLを包括的に評価すること目的として、認知機能、適応行動能力、注意欠陥・多動性障害(ADHD)およびQOLを同時に評価した。対象は5歳から18歳の53人であった。解析の結果、胆道閉鎖症患者では認知機能と適応行動能力は日本人の標準集団と比較し低値であった。また、情緒・行動の問題、ADHD特性は標準集団よりも高かったが、患者の認知機能と相関を認めず、適応行動能力と相関を認めた。さらに、認知機能は境界域だが行動障害を伴わない胆道閉鎖症患者は、見逃されやすく、学校や医療機関での十分なサポートが提供できていない可能性があり、注意深く診療する必要が示された。以上から、学位の授与に値すると考えられる。