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## Aspects of biliary atresia and other pediatric cholestatic diseases

Yang, Huiqi

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## 中文摘要

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在本论文中，我们研究了与胆道闭锁相关的多个方面和其它儿童黄疸性疾病。

我们主要研究胆道闭锁的病理机制。胆道闭锁的动物模型是通过采用轮状病毒注射新生小鼠建立的。首先，对这一动物模型相关的研究进行了系统回顾。然后，在这个动物模型基础上，证实了肝纤维化开始于胆道闭锁发展的早期。肝脏纤维化与上皮细胞向肝实质细胞的转化和 Hedgehog 信号通路的激活同时发生。在胆道梗阻形成之前，肝脏纤维化的过程已经发生。随后，对炎症对肝胆运输基的影响作了研究。炎症导致肝胆运输基相关基因下调，从而导致了肝内黄疸。这是导致肝脏纤维化早期发生的主要原因。

在论文的第二部分，对胆道闭锁和其它儿童黄疸性疾病的临床方面做了研究。在这部分的第一章中，回顾了部分胆道外引流治疗进展性家族性肝内黄疸性疾病和 Alagille 综合征的长期结果。研究结果证明这种手术对术前肝脏活检没有纤维化的儿童取得很好的效果。

Meta 统计分析证明胆道闭锁经手术治疗后的患者大剂量服用类固醇类药物可能会取得较好的术后效果。回顾性研究胆道闭锁经手术治疗的临床资料，证实术中可见胆道引流是影响术后预后的因素。