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The Prevalence of Congenital Heart Defects in Infants with Cholestatic Disorders of infancy: A single center study

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Introduction: There is deficiency of data about congenital heart defects (CHD) in cholestatic disorders of infancy other than Alagille syndrome. There are many conditions that cause neonatal cholestasis, the most commonly identifiable are biliary atresia (BA), genetic disorders, metabolic diseases, and α1-antitrypsin deficiency. BA could be associated with other congenital anomalies. Aim of work: To define the prevalence and types of CHD in infants with various causes of cholestatic disorders of infancy. Methods: This cross sectional study was conducted on 139 infants presenting with cholestasis whether surgical or non-surgical. The study was carried out at the Pediatric Hepatology Unit, Cairo University Children's Hospital, Egypt. Full examination and investigations were done in an attempt to reach an etiologic diagnosis for cholestasis; in addition to a comprehensive echocardiographic study. Results: The age at the onset of cholestasis ranged from 1 day to 7 months. Males constituted 61.2%. BA was diagnosed in 39 patients (28%), AGS in 16 patients (11.5%), 27 patients had miscellaneous diagnoses and 57 cases had indeterminate etiology. CHD were detected in 55 patients (39.5%). Shunt lesions were detected in 24 patients; 43.6%, pulmonary stenosis in 18 patients (32.7%) and combined lesions in 9 cases (16.4%). Three patients (5.5%) had abnormal cardiac situs. Only seven patients had clinical presentation suggestive of CHD. CHD were detected in 14 patients with BA (35.9%), 15 patients with AGS (93.7%) and 26 patients in the remaining group (30.9%). Among the 39 patients with BA, 14 (35.7%) had CHD: 9 patients (23%) had shunt lesions, the most common cardiac anomaly was PDA in 5 patients (12.8%) and the second most common was ASD secondum in 3 patients (7.7%). Only 3 patients had major congenital cardiac defects (7.7%) with two of whom having situs inversus associated with cardiac defects (5.1%).
Conclusion: CHD are not uncommon among cholestatic infants other than AGS. Echocardiography is recommended as part of the preoperative assessment infants with BA before undergoing hepatic portoenterostomy to exclude presence of CHD which may impact the anesthetic planning, timing of surgery and the outcome of hepatobiliary surgery.