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Occurrence of pulmonary hypertension in Heterotaxy syndrome: A retrospective observational study

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Introduction: Heterotaxy Syndrome (HS) is defined as an abnormality of thoraco-abdominal visceral arrangement across the left-right axis of the body leading to either polysplenia or asplenia. Mortality in these patients depends on associated cardiac, pulmonary and abdominal lesions. We evaluate the hypothesis that Pulmonary Hypertension (PH) increases the mortality of HS in pediatric population.

Methods: After thorough review of electronic databases, patients with HS were classified as polysplenia or asplenia. PH prevalence was determined in each group based on echocardiogram (pulmonary vascular systolic pressure as definitively ≥ 50% systemic) and/or cardiac catheterization (pulmonary vascular resistance > 3WU/m^2) results. Associated cardiac lesions were categorized as either 1) Simple lesions (e.g., ASD, common AV canal defect); 2) Complex two ventricle palliation (e.g., TGA, DORV); or 3) Single ventricle palliation (e.g., HLHS). Chi square test was used to test differences among groups. Mortality and median survival in each was determined.

Results: Of 236 patients initially coded for HS, 56 patients (born between 1954 and 2006) met unequivocal criteria for HS: 21(37.5%) polysplenia and 35 (62.5%) asplenia. Simple cardiac lesions were more common in polysplenia (50% vs 17.9%, $p = 0.021$). PH was present in 33.9% of patients, predominantly in the polysplenia group (52.4% vs 22.9%, $p = 0.023$). PH occurred with simple cardiac lesions more frequently than with complex cardiac lesions (64.2% vs 23.8%, $p = 0.005$). The overall mortality rate for HS was 37.5% without a significant difference between polysplenia vs asplenia ($p > 0.05$) or between simple vs complex lesions ($p > 0.05$). However, the association of PH increased mortality to 68.4% ($p < 0.001$).

Conclusions: PH is present in about one third of cases of HS and in more than half of cases with polysplenia. The simple cardiac lesions seen in polysplenia tend to be at a higher risk of developing PH. Mortality is significantly increased in HS when PH is present, suggesting patients with polysplenia die from PH complications whereas patients with asplenia die from complications of their complex cardiac lesions. Evaluation of this precisely phenotyped historical HS cohort confirms our hypothesis that PH is a potential cause of death in HS. Early diagnosis and treatment of PH may prevent premature mortality, especially in patients with polysplenia.

Biography

Keyur Mehta graduated from Grant Medical College in Mumbai at the age of 23 years. He completed his pediatrics training from State University of New York, Downstate Medical Center. Before starting residency, he worked at the National Cancer Center in Singapore. Currently, he is working as a Clinical Assistant Instructor and Chief Resident at SUNY Downstate. He is a member of the Medical Council of India as well as the American Academy of Pediatrics. He is going to begin his Pediatric Cardiology fellowship at Nicklaus Children's Hospital (formerly Miami Children's Hospital) in Florida next year.

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