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Performance of the “GUCH morbidity and mortality scores” in cyanotic and non-cyanotic adults with congenital heart disease

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Abstract

Background Cyanotic adults with congenital heart disease (ACHD) rely on life-sustaining adaptations to their chronic cyanosis and are at risk of adverse consequences if such adaptations are out of balance. They may benefit from surgical treatment options that were not available during their childhood. The “GUCH morbidity and mortality scores” were specifically designed to predict risks after surgery in ACHD and included comorbidities and patient age. We aim to assess their performance in cyanotic compared to non-cyanotic ACHD.

Methods Data of all consecutive adults who underwent CHD surgery in 2005–2016, were collected. Mortality was defined as hospital mortality or mortality within 30 days following surgery. Morbidity was defined as occurrence of one or more of the following complications: renal failure requiring dialysis, neurological deficit persisting at discharge, atrioventricular block requiring pacemaker implantation, mechanical circulatory support, phrenic nerve injury and unplanned reoperation. The performance of the GUCH scores was assessed using the area under the receiver operating characteristics curve (C-index, 95% CI).

Results We evaluated 824 operations including 99 performed in cyanotic ACHD. The mean age at operation was 34 ± 13 years (18–72 years). Cyanotic patients had higher hospital mortality and morbidity than non-cyanotic patients (11.1% vs. 2.4%, $P < 0.0001$; and 22.2% vs. 8.4%, $P < 0.0001$ respectively).

C-index for GUCH mortality score in cyanotic and non-cyanotic ACHD were not different: 0.722 (0.536–0.907) and 0.800 (0.712–0.887) ($P = 0.44$), respectively. C-index for GUCH morbidity score was lower in cyanotic than in non-cyanotic ACHD: 0.483 (0.335–0.632) vs. 0.671 (0.601–0.741) ($P = 0.027$).

Conclusion GUCH mortality score is efficient in cyanotic ACHD patients. However the GUCH morbidity score should be refined in these patients.

Keywords Adult with congenital heart disease; Cyanosis; Mortality; Morbidity

Disclosure of interest The authors have not supplied their declaration of competing interest.

<https://doi.org/10.1016/j.acvdsp.2019.06.015>



PC 8

Combined lung transplantation and percutaneous septal defect closure for end-stage atrial septal defect associated pulmonary arterial hypertension

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Abstract

Introduction Pulmonary arterial hypertension (PAH) is a rare but severe complication of unrepaired atrial septal defect (ASD). In patients with ASD, PAH and right ventricle failure, heart-lung transplantation (HLT) is currently viewed as the best ultimate procedure but is limited by the low number of grafts. We aim to report our preliminary experience of double lung transplantation (DLT) followed by percutaneous ostium secundum ASD closure in patients with end-stage ASD associated PAH.

Methods We report 6 patients with end-stage ASD associated PAH treated with DLT followed by percutaneous ASD closure.

Results Median ages at ASD diagnosis and PAH diagnosis were respectively 22 (min:0, max: 37) and 16 (min: 2 max: 40) year-old. Median mean pulmonary artery pressure and pulmonary vascular resistance before DLT were 78 mmHg (min: 49, max: 108) and 14.0 WU.m² (min: 12.5, max: 17.4) respectively.

Median age at DLT was 29 (min 20, max 44) year-old. Severe primary graft dysfunction occurred in 3 (50%) patients and was successfully managed medically. Post operative extracorporeal membrane oxygenation was required in 5 (83%) patients. Median ventilation time was 12 days (min: 2, max 20). Transient tracheotomy was required in 2 patients. Median time of stay in intensive care unit was 15 days (min:7, max: 26).

Median delay between DLT and percutaneous ASD closure occlusion delay was 4.4 months (min: 3.9 max: 18.7). Percutaneous closure was achieved using Amplatzer septal occluder in all patients (size running from n° 16 to 40). Mean pulmonary arterial pressure was within normal range in all patient before ASD closure. All patient

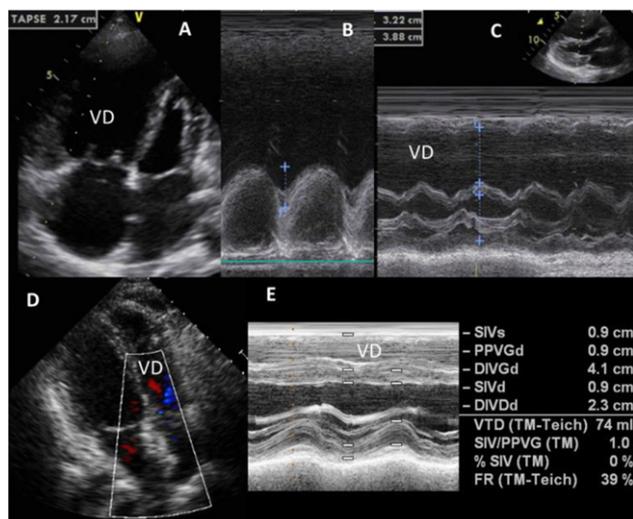


Fig. 1 Echocardiography before (A,B,C) and after (D,E) DLT and ASD closure.