

**ACC.14**

TCT@ACC-12 | Innovation in Intervention

A588

JACC April 1, 2014

Volume 63, Issue 12



Congenital Heart Disease

MAJOR ADVERSE CARDIAC EVENTS IN CHILDREN WITH WILLIAMS BEUREN SYNDROME UNDERGOING SURGERY: AN ANALYSIS OF THE SOCIETY OF THORACIC SURGEONS CONGENITAL HEART SURGERY DATABASE

Poster Contributions

Hall C

Sunday, March 30, 2014, 3:45 p.m.-4:30 p.m.

Session Title: Advances in Congenital Heart Surgery

Abstract Category: 10. Congenital Heart Disease: Pediatric

Presentation Number: 1227-272

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Background: Children with Williams Beuren Syndrome (WBS) undergoing cardiac surgery are at risk for major adverse cardiac events (MACE) but the frequency of and risk factors for such events have not been well described. We sought to better define frequency and risk of MACE in WBS using the STS-CHS Database.

Methods: We examined patient characteristics and in-hospital outcomes for cardiac operations in WBS patients (2000-2012) using the STS-CHS Database. The cohort was divided into 4 procedural groups: isolated supravalvar aortic stenosis, complex left ventricular outflow tract (LVOT), isolated right ventricular outflow tract (RVOT) and combined LVOT + RVOT surgeries. We used univariate logistic regression to evaluate the association between preoperative factors and a composite outcome of in-hospital death, cardiac arrest or postoperative mechanical circulatory support.

Results: Of 447 index operations (87 centers), median (IQR) age and weight at surgery were 2.4 years (0.6, 7.4) and 10.6 kg (6.5, 21.5). Overall mortality was 4.5%. The composite outcome occurred in 9.2% but with significant procedural variability ($p < 0.001$) [Table1]. Odds of the composite outcome were lower with increasing weight (OR=0.97; 95% CI 0.93, 0.99) and age (OR=0.99; 95% CI 0.98, 0.99) at surgery, but higher in the presence of any preoperative risk factor (OR=2.1; 95% CI 1.1, 4.0).

Conclusion: Major adverse cardiac events are common in WBS following left sided cardiac surgery but less frequent after isolated RVOT surgery.

	Overall N=447	Isolated SVAS1 N=129	Complex LVOT N=131	Isolated RVOT N=44	LVOT + RVOT N=87
Preoperative factors					
Any preoperative risk factor	112 (25%)	22 (17%)	35 (27%)	20 (46%)	19 (22%)
Mechanical circulatory support	6 (1%)	1 (1%)	2 (2%)	0 (0.0%)	2 (2%)
Mechanical ventilatory support	26 (6%)	3 (2%)	8 (6%)	3 (7%)	6 (7%)
Renal failure	2 (<1%)	0 (0%)	0 (0%)	1 (2%)	0 (0%)
Neurological deficit	14 (3%)	2 (2%)	5 (4%)	3 (7%)	3 (4%)
Outcomes					
Any postoperative complication	190 (42%)	33 (25%)	59 (45%)	18 (39%)	51 (58%)
In-hospital mortality	20 (5%)	2 (2%)	7 (5%)	0 (0%)	6 (7%)
Cardiac arrest	14 (3%)	1 (1%)	2 (2%)	(0%)	8 (9%)
Mechanical circulatory support	25 (6%)	4 (3%)	7 (5%)	0 (0%)	11 (13%)
Composite outcome ²	42 (9%)	5 (4%)	12 (9%)	0 (0%)	18 (21%)

1. SVAS represents supravalvar aortic stenosis

2. Composite outcome represents in-hospital death, cardiac arrest or postoperative mechanical circulatory support