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Congenital Heart Disease

CARDIAC SURGERY OUTCOMES IN HETEROTAXY SYNDROME: 25 YEARS EXPERIENCE FROM A MULTICENTER CONSORTIUM

Poster Contributions
Hall C
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Session Title: Advances in Congenital Heart Surgery
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Background: Heterotaxy syndromes of asplenia and polysplenia encompass a wide spectrum of complex cardiovascular abnormalities associated with left-right asymmetry. The combination of structural and electrical cardiac abnormalities along with extracardiac co-morbidity makes cardiac surgery in heterotaxy syndrome challenging and associated with high morbidity and mortality. We report the mortality after cardiac surgery in patients with heterotaxy from the Pediatric Cardiac Care Consortium (PCCC).

Methods: We identified 1,169 patients with a diagnosis of heterotaxy that underwent cardiac operation in a PCCC center between 1982 and 2005.

Results: 753 (64.4%) had asplenia and 416 (35.6%) polysplenia. Detailed results are presented in the following table.

A. Initial Procedure				
	Asplenia		Polysplenia	
	n	Mortality	n	Mortality
Aorto-Pulmonary shunt	375	17%	114	13.20%
Pulmonary band	25	28%	21	38.10%
Glenn shunt	44	9.10%	18	0.00%
Fontan	8	37.50%	11	27.30%
Total anomalous pulmonary venous return repair	157	47%	17	23.50%
Pacemaker	8	25%	30	46.7%
Miscellaneous*	136	13.4%	205	24.50%
Total	753	27.8%	416	30.5%
B. Subsequent Procedures				
	Asplenia		Polysplenia	
	n	Mortality	n	Mortality
Aorto-Pulmonary shunt	110	10.90%	34	0%
Glenn shunt	257	5.80%	92	5.40%
Fontan	274	15.30%	97	9.30%
Total anomalous pulmonary venous return repair	45	24.40%		
Miscellaneous*	233	6.9%	116	6.9%
Total	919	11.6%	339	6.3%

C. Accumulative outcomes after 1st and subsequent procedures			
	Asplenia	Polysplenia	All Heterotaxy
n (total patients)	753	416	1,169
n1 (survivors 1st procedure)	543 (72.1%)	289 (69.5%)	832 (71.2%)
n2 (survivors subsequent procedure)	480 (63.7%)	272 (70.4%)	752 (66.3%)
Univentricular palliation (% of n1)	232	89	321
Cardiac transplantation	20 (2.7%)	13 (3.1%)	33 (2.8%)
Pacemaker/AICD	65 (8.7%)	89 (21.4%)	154 (13.2%)
Overall mortality	267 (35.8%)	114 (27.4%)	381 (32.6%)

*Miscellaneous procedures include: atrioventricular valve repair, closure of atrial or ventricular septal defects, augmentation of the aorta and others.

Conclusion: Our results illustrate a high overall surgical mortality (32.6%) in patients with heterotaxy syndrome (35.9% in asplenia and 27.4% in polysplenia). Univentricular palliation occurs frequently in asplenia (48.3%) than polysplenia (32.7%) survivors ($p<0.01$). Pacemaker implantation occur in 1% of the asplenia and 7% in polysplenia ($p<0.0001$) with significant mortality (25% and 46.7% respectively).