Pretriscuspid shunt and Eisenmenger syndrome: a deadly combination

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Introduction: Pulmonary arterial hypertension (PAH) is a common comorbidity in congenital heart disease (CHD) and significantly affects prognosis. Our aim was to investigate how the location of the shunt influences the outcome of these patients.

Methods: The REHAP registry is a Spanish multicentre voluntary registry of patients over 14 years of age, which includes patients with congenital heart disease and PAH, starting in 2007. Baseline characteristics, functional class, right catheterization data, treatment and survival were compared. We performed a cox regression model for assessing the impact of the type of shunt on prognosis, after looking for confusing and effect modifying variables.

Results: 664 patients were analyzed, their characteristics are detailed in the table. Mean follow up time was 10 years. We performed a survival analysis adjusted for the confounding variables: age (Hazard ratio (HR) 1.02 p<0.001), functional class (HR 2.45 p 0.001) and clinical group. The clinical group was also an effect-modifying variable, which means there is no unique effect (HR) of the type of shunt, instead it depends on the clinical group. We did a multiple regression analysis, adjusting for the confounding variables, to calculate the HR for each clinical group: in patients with Eisenmenger syndrome, the HR is 0.27 (p<0.001) for simple posttricuspid shunt and HR 0.38 (p<0.001) for complex posttricuspid shunt (reference group: pretricuspid shunt). In the others clinical groups, the type of shunt did not reach statistical significance.

Conclusion: The type of shunt significantly affects prognosis in Eisenmenger patients, with a much worse outcome for those with pretricuspid shunts. Functional class and clinical group are also closely related to outcome.

	All	Simple pretricuspid shunt	Simple posttricuspid shunt	Complex posttricuspid shunt	p
N	664	209 (31)	288 (43)	167 (25)	
Age (years)	38±17	48±17	36±16	28±12	<0.001
Sex (female)	447 (67)	157 (75)	185 (64)	105 (63)	0.014
Clinical group					<0.001
Eisenmenger	371 (56)	53 (26)	184 (64)	134 (80)	
Large left to right shunt	68 (10)	46 (22)	17 (6)	5 (3)	
Small shunt	62 (9)	40 (19)	19 (7)	3 (2)	
Postoperative	159 (24)	68 (33)	66 (23)	25 (15)	
NYHA Functional class III–IV	331 (50)	109 (52)	136 (47)	86 (52)	0.492
Pulmonary vascular resistance (mmHg)	10±7	9±5	13±7	12±8	<0.001