

## Prognostic Significance of 2-Dimensional, M-Mode and Doppler Echo Indices of Right Ventricular Function in Children with Pulmonary Arterial Hypertension

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### Background:

Right ventricular (RV) function may be a key determinant of mortality in pediatric idiopathic pulmonary artery hypertension, (iPAH) and that associated with congenital heart disease (cPAH) but echo indices of RV function have not been adequately studied.

### Methods:

Children (0-18 yr) with iPAH and cPAH were retrospectively studied. RV function indices (indexed RV end diastolic area, fractional area change, tricuspid annular excursion, right atrial volume) were analyzed at diagnosis and at last follow up. Indices were compared between iPAH and aPAH pts at baseline and follow up. iPAH pts alive (group 1) were compared with those dead/ transplanted (group 2) at time of study. Cut points predictive of survival were generated (ROC) and Kaplan Meier survival analyzed.

### Results:

54 pts (36 cPAH (7.5±5.9 yrs; M:F 12:24); 18 iPAH (8.9±5.7 yrs; M:F 7:11; group 1 n=12, group 2 n=6) were studied. Median follow up was 4.3 (0.2-7.4) yrs. Despite similar PVRi, RV function was significantly decreased in iPAH vs cPAH pts during follow up (table). Group 1 iPAH maintained RV function on follow up, while it worsened in group 2 (fig 1). In iPAH survival was significantly different based on RV function cutpoints (fig 2).

### Conclusion:

Conventional echo RV function indices including RV end diastolic area, fractional area of change and tricuspid annular excursion appear to be useful for prognosis in children with PAH.

	Variable First Echo			Last Echo		
	iPAH (n=18)	cPAH (n=36)	P*	iPAH (n=17)	cPAH (n=31)	P*
RV systolic pressure mmHg	85±17	71±3	0.024	83±16	60±24	0.025
RV end-diastolic area cm <sup>2</sup> /m <sup>2</sup>	26±10.6	21±5	0.074	27±13	17.6±8	0.01
RV end-systolic area cm <sup>2</sup> /m <sup>2</sup>	21±7.4	14.8±4.4	0.001	24±15	13.3±5.3	0.002
RV ED Z score	4.2±1.9	2.7±1.6	0.027	5±2.4	2±2.1	0.001
RV fractional area change (%)	19±6	30±7	0.0001	21.1±9	30±10	0.003
RA volume index	39±15	35±20	0.184	60±42	35±20	0.016
TAPSE Z score	-4.7±2.4	-3.4±2.5	0.044	-3.6±3.4	-4.2±2.1	0.442
LV end-systolic eccentric index	2.3±0.7	1.9±1.8	0.002	2.4±1.1	1.4±0.33	0.003
LV end-Diastolic eccentric index	1.7±0.37	1.3±0.26	0.01	1.6±0.5	1.3±0.19	0.012

