RV Function is A key Parameter in Assessment of Patients with Pulmonary Arterial Hypertension

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PULMONARY HYPERTENSION CLASSIFICATION (PHTN)

1. Pulmonary arterial hypertension (PAH)
    a. idiopathic (iPAH)
    b. familial
    c. secondary: congenital heart disease (cPAH)
2. Pulmonary venous hypertension
3. PHTN associated with hypoxia and other resp. disease
4. PHTN associated with chronic thrombotic disease
5. PHTN associated with miscellaneous things

PULMONARY ARTERIAL HYPERTENSION (PAH)

PAH defined:

mean pulmonary artery pressure (PAP) of > 25 mmHg.

pulmonary vascular resistant (PVR) > 3 Wood Unite.

PULMONARY ARTERIAL HYPERTENSION (PAH)

PAH progressive disease that carries high morbidity and mortality in children

In children, idiopathic PAH carries worse prognosis than PAH in congenital heart disease

RV FUNCTION IS AKEY PARAMETER IN ASSESSMENT OF ADVERSE OUTCOM

- RV ejection fraction: an indicator of increased mortality in patients with CHF associated with CAD

(Polak et al. J Am Coll Cardiol 1983)

- RV function predicts exercise capacity and survival in advanced heart failure

(Di Salvo et al. J Am Coll Cardiol 1983)

- RV function is a crucial determinant of short-term prognosis in severe chronic heart failure

(Gavazzi et al. J Heart Lung Transplant 1997)

RV FUNCTION IS AKEY PARAMETER IN ASSESSMENT OF ADVERSE OUTCOM

- RV ejection fraction: independent predictor of survival in patients with moderate heart failure

(De Groote et al. J Am Coll Cardiol 1998)

- RV function predicts prognosis in patients with chronic pulmonary disease

(Burgess et al. J Am Soc Echocardiogr 2002)

- RV contractile reserve is associated with one year mortality in patients with DCMP

(Otasevic et al. Eur J Echocardiography 2005)

RV FUNCTION IS AKEY PARAMETER IN ASSESSMENT OF PATIENTS WITH PAH

Echocardiography is the most common modality used for assessment and serial follow up of RV function in children with PAH

Nat (nature) Rev Cardiol 2010;7:551-63.

RV FUNCTION IS AKEY PARAMETER IN ASSESSMENT OF PATIENTS WITH PAH

Two- dimensional, M-mode, and Doppler echo measures have been associated with adverse outcomes in adults with iPAH

Am J Cardiol 2011;107:628-32

RV FUNCTION IS AKEY PARAMETER IN ASSESSMENT OF PATIENTS WITH PAH

The objective of this study was to investigate conventional (2D, M-mode, and Doppler) echocardiographic indices of RV function in relation to death or lung transplant in children with iPAH and cPAH

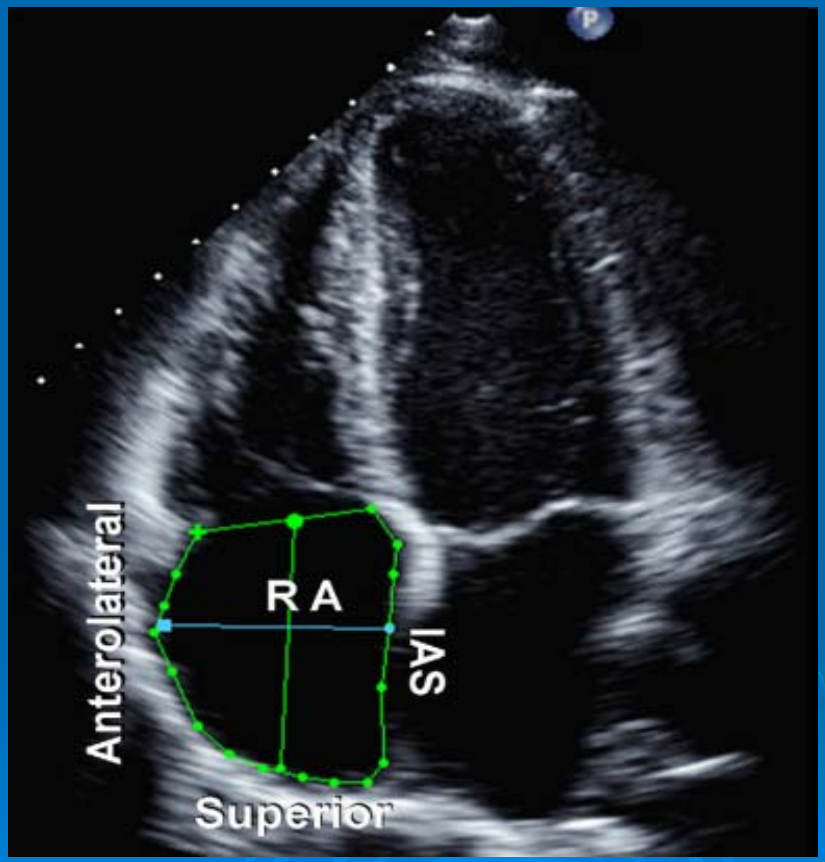
Right Atrial VOLUME

RA enlargement correlate to RV dysfunction

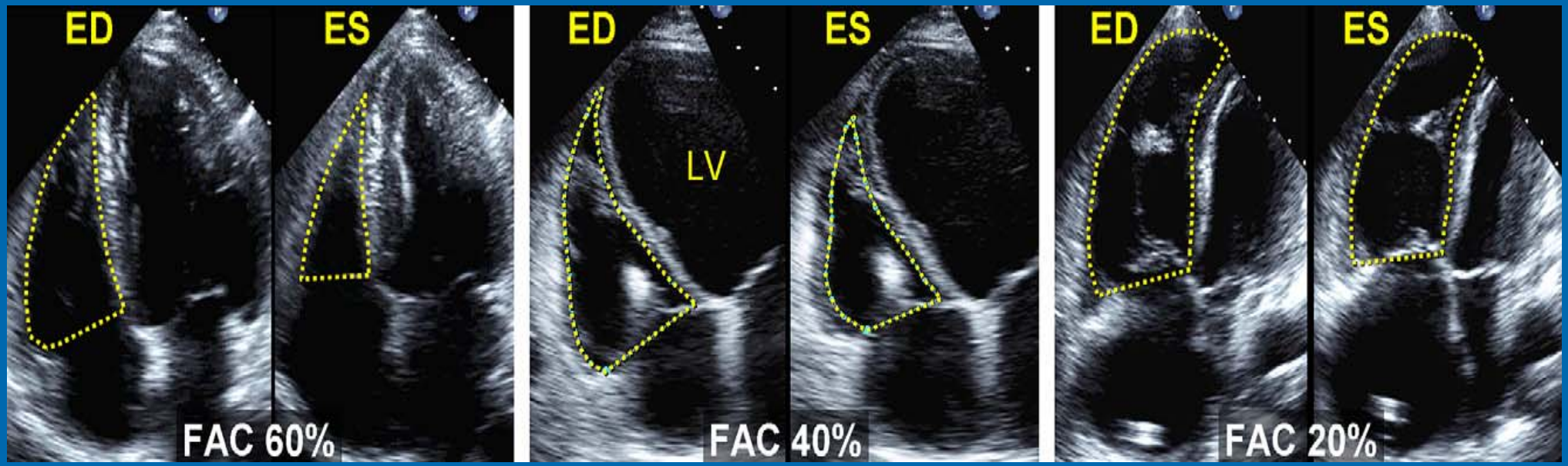
(Bustamante-Labarta, J Am Soc Echocardiogr 2002;15:1160-4)
(Lopez,J Am Soc Echocardiogr 2010;23:465-95).

Have been found to be predictive of survival or need for transplantation in adult patients with PAH.

(Bossone E, J Am Soc Echocardiogr. 1999;12:655-62

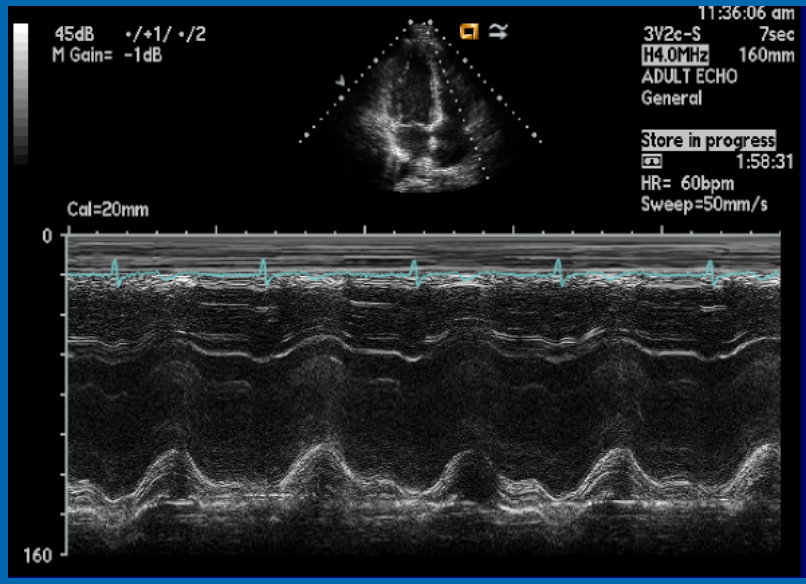


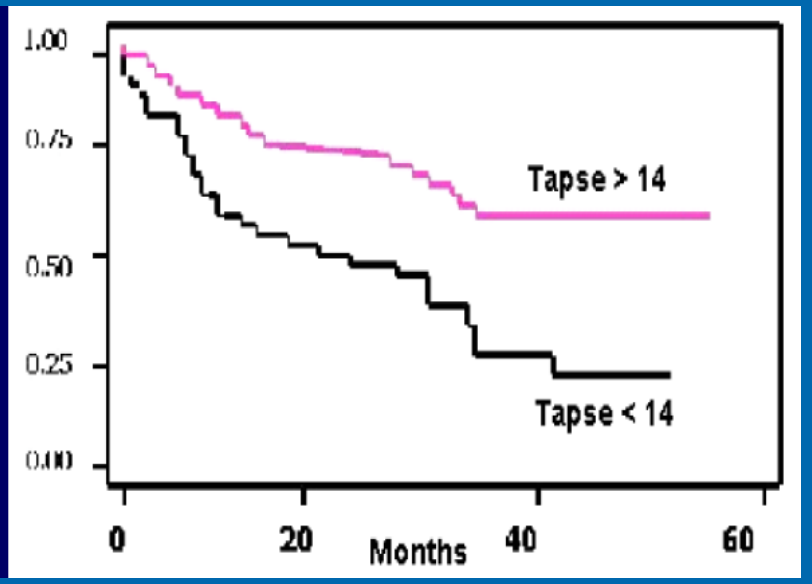
RV Area and Fractional Area Change (%FAC)



- Well correlated with RV function measured by radionuclide ventriculography or MRI
- Good predictor of prognosis
- Limitations: fail to measure FAC due to inadequate RV tracing

Tricuspid Annular Plane Systolic Excursion (TAPSE)





In adults TAPSE correlates well with RV EF, FAC, RV stroke volume

(López-Candales, Am J Cardiol 2006;98:973)
(Saxena, Echocardiography 2006 :23, 750–755)
(Forfia, Am J Respir Crit Care Med 2006 174:1034–1041)

Event free survival according to TAPSE in patients with CHF

(Forfia, Am J Respir Crit Care Med, 2006 174:1034)

Tricuspid Annular Plane Systolic Excursion (TAPSE) – In Children

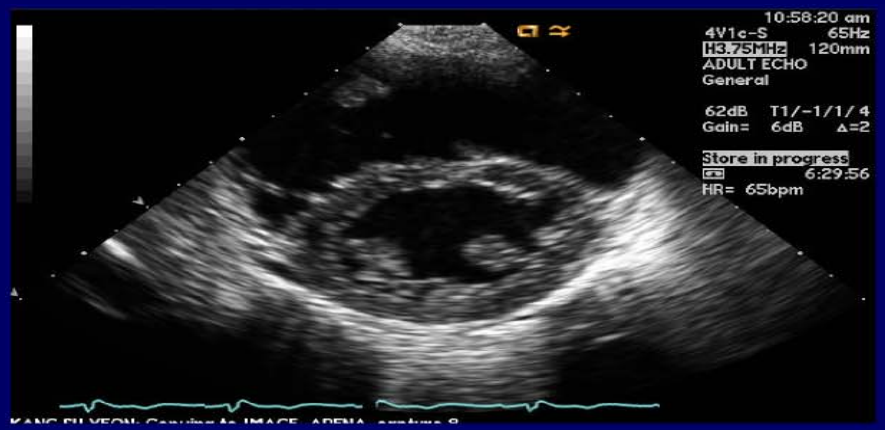
- Normal values have been established for children and neonates

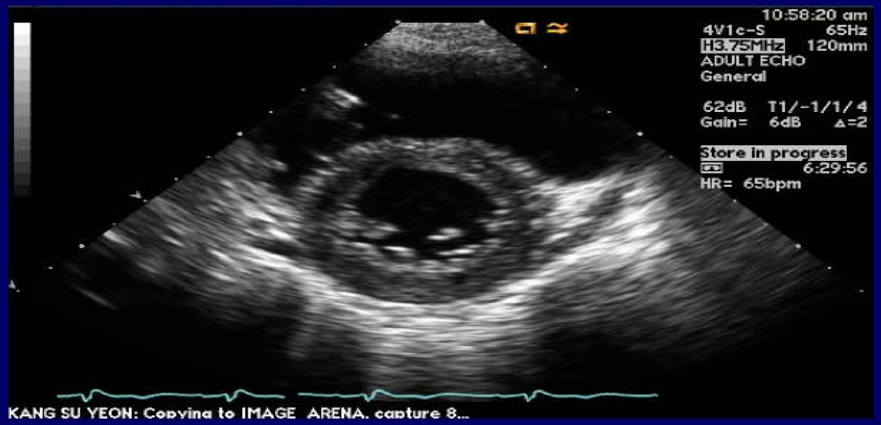
(Koestenberger J Am Soc Echocardiogr. 2009;22:715)

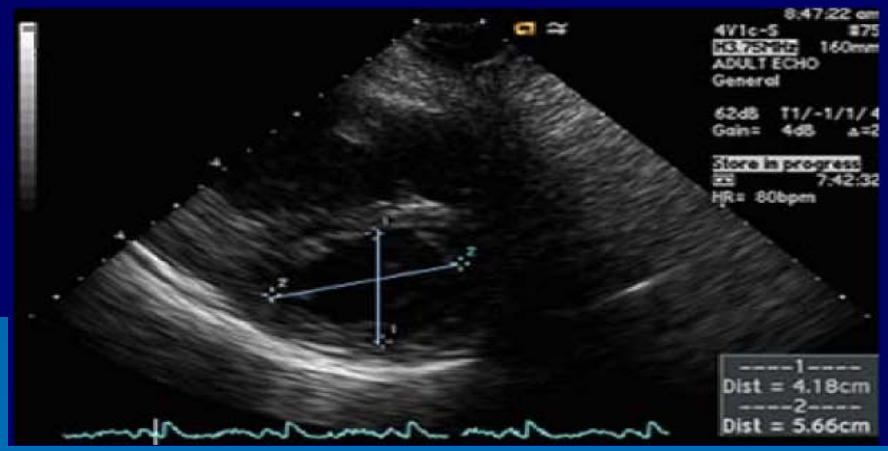
(Koestenberger M, Neonatology. 2011;100:85-92)

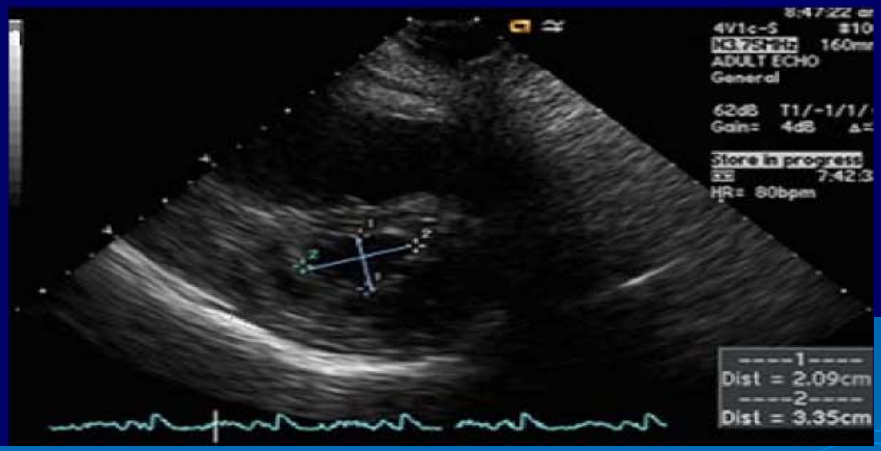
- Little pediatric data investigating its use for evaluation of RV function or its prognostic value in children with PAH.

LV - Eccentricity Index





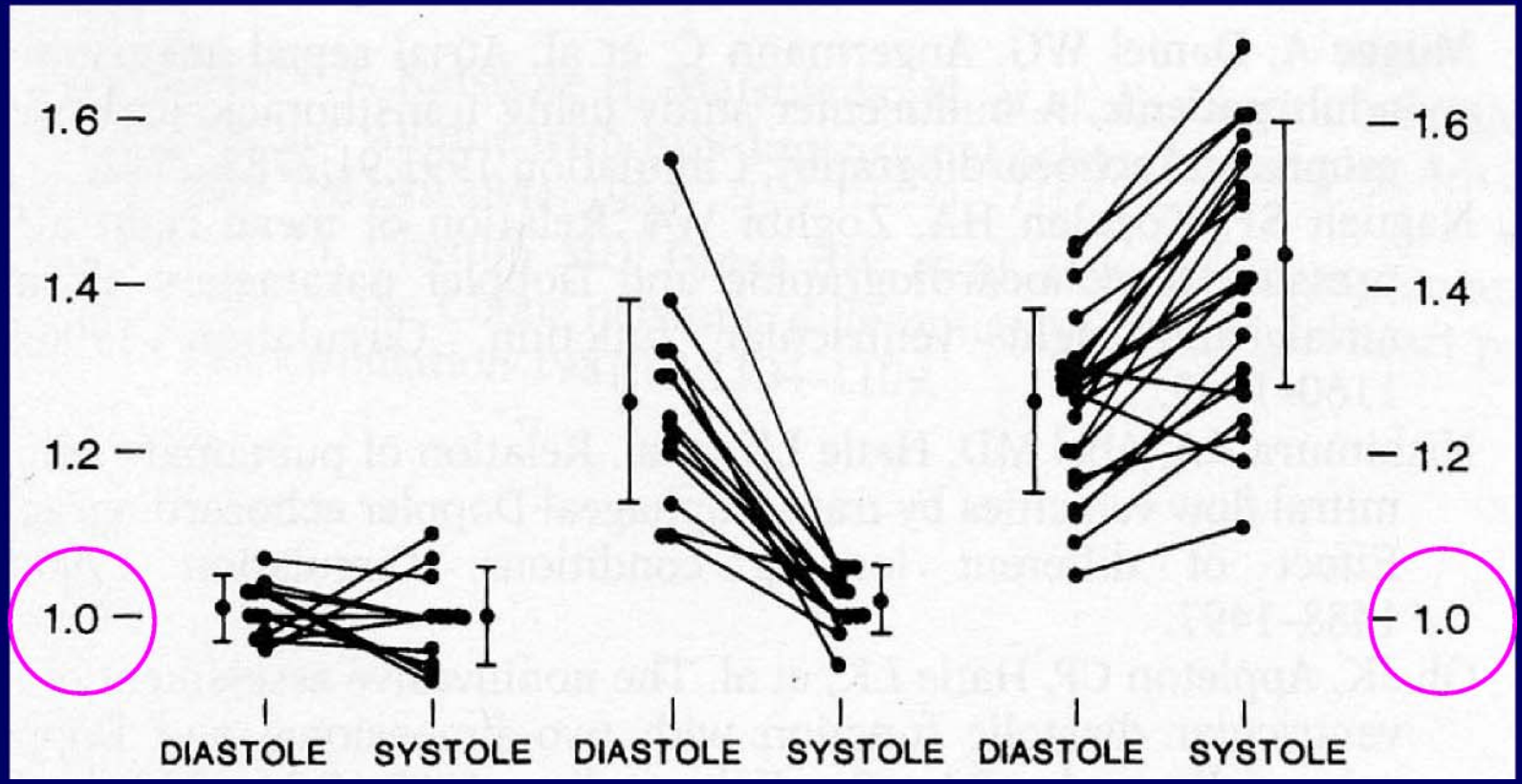




- Reflects the degree of septal flattening resulting in abnormal LV shape

- Normal: approximately 1.0 in both diastole and systole

LV - Eccentricity Index

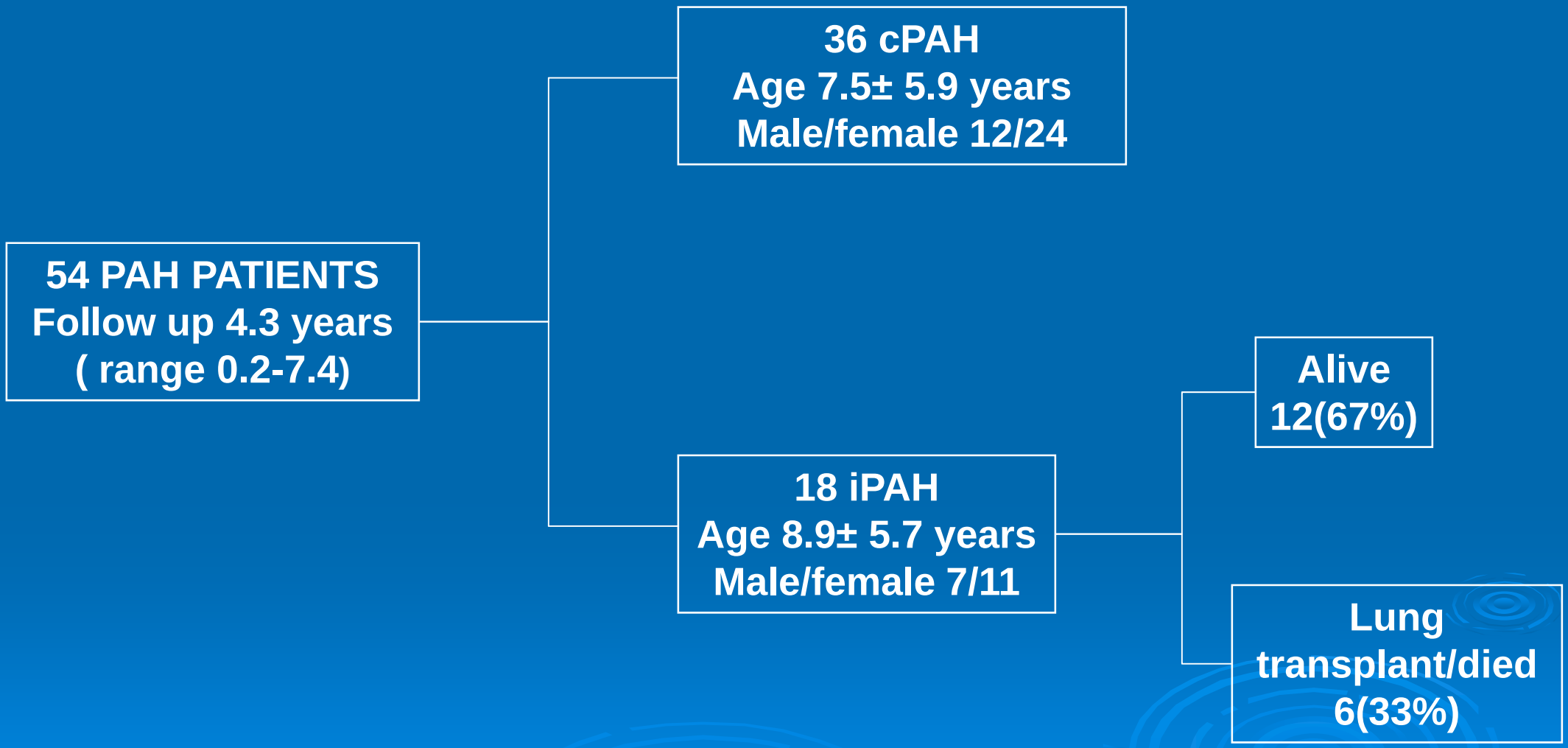


RV volume Overload

RV pressure Overload (PAH)

Prognostic significance of 2 dimensional, M-mode and Doppler echo indices of right ventricular function in children with pulmonary arterial hypertension

Eias Kassem and Mark Friedberg


54 PAH PATIENTS
Follow up 4.3 years
(range 0.2-7.4)
36 cPAH
Age 7.5± 5.9 years
Male/female 12/24
18 iPAH
Age 8.9± 5.7 years
Male/female 7/11
Alive
12(67%)
Lung
transplant/died
6(33%)

Demographic data from all iPAH and cPAH patients at time of the first and last echocardiogram

Variable	First Echo			Last Echo		
	iPAH (n=18)	**cPAH (n=36)**	***p-value**	**iPAH (n=17)**	**cPAH (n=31)**	***p-value**
Age (years)	8.9±5.7	7.48±5.9	0.37	11.7±6	11.1±6	0.70
Female n, (%)	11 (55%)	24 (66%)		10 (59%)	18 (58%)	
Systolic BP (mmHg)	90±11	98±16	0.13	104±19	99±11	0.45
Diastolic BP(mmHg)	54±8	57±10	0.24	56±9	63±19	0.43
6-minute walk distance (meter)	372±77	392±82	0.40			
PVRi (WU x m^2)	23.4±10.6	16±9.6	0.006			
Number of medications						
1	9 (50%)	24 (67%)				
2	4 (22%)	4 (11%)				
3	3 (16%)	3 (8%)				
4	1(6%)	0 (0%)				

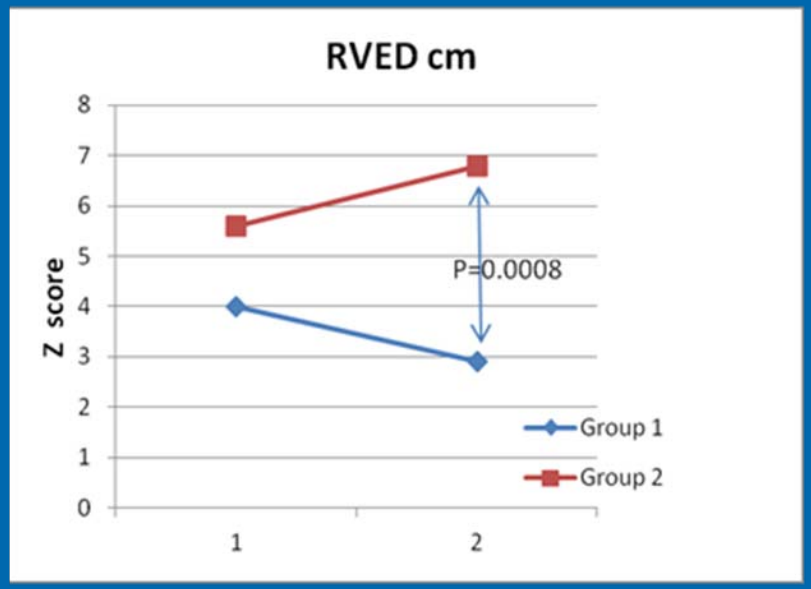
Demographic data from iPAH patients comparing patients from group 1 (survivors) vs. group 2 (death/transplant) at first and last echocardiogram

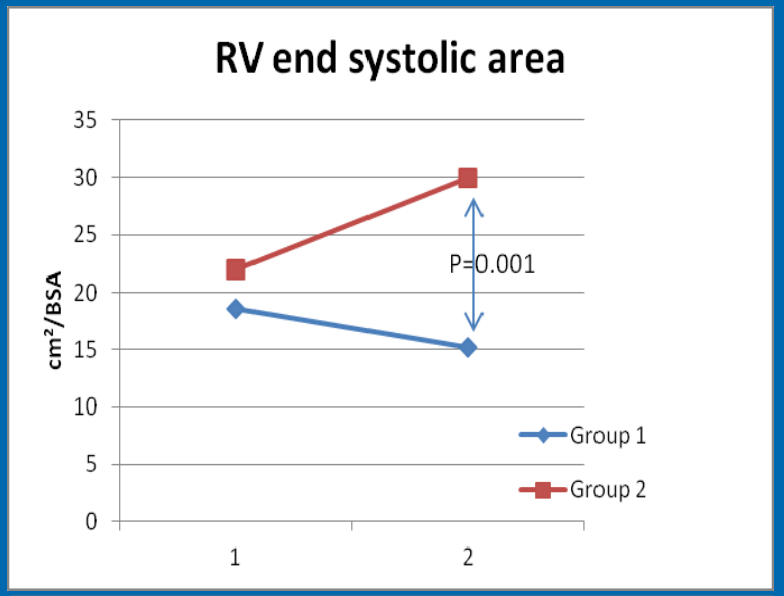
Variable	First Echo			Last Echo		
	Group 1 n=12	**Group 2 n=6**	**p-value**	**Group 1 n=12**	**Group 2 n=6**	**p-value**
Age (years)	9.3±5	9.3±6	0.99	12.7±5.8	11.7±6.5	0.80
Female n, (%)	7 (63%)	4 (66%)		6 (55%)	4 (66%)	
Heart rate (bpm)	92±21	103±29	0.57	79±20.5	103±20.4	0.06
Systolic BP (mmHg)	92±12	92±10	0.93	99±15	106±22	0.61
Diastolic BP (mmHg)	52±8	58±7	0.14	58±15	64±20	0.52
6-MW (meters)	397±58	371±149	0.60			
PVRi (WU)	24±12	19±6	0.66			
Number of medications:						
0	1 (8%)	0 (0%)				
1	6 (50%)	3 (50%)				
2	4 (34%)	0 (0%)				
3	1 (8%)	2 (33%)				
4	0 (0%)	1 (17%)				

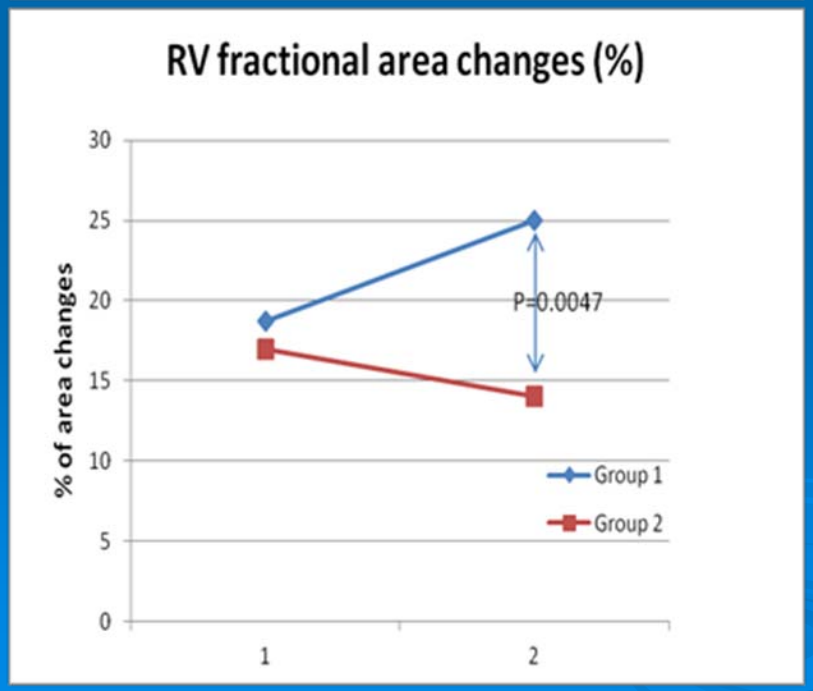
RV indices in iPAH and cPAH patients at presentation and at last echo.

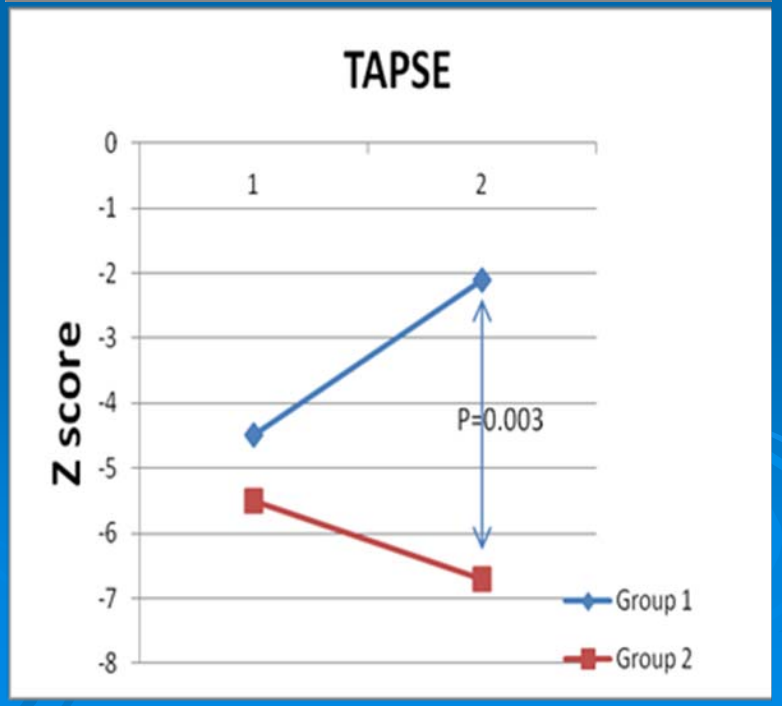
Variable	First Echo			Last Echo		
	iPAH (n=18)	cPAH (n=36)	p*	iPAH (n=17)	cPAH (n=31)	p*
RVSp (mmHg)	85±17	71±3	0.02	83±16	60±24	0.02
indexed RV end-diastolic area (cm^2)	26±10.6	21±5	0.07	27±13	17.6±8	0.01
index RV end-systolic area (cm^2)	21±7.4	14.8±4.4	0.001	24±15	13.3±5.3	0.002
RVED Z-score	4.2±1.9	2.7±1.6	0.02	5±2.4	2±2.1	0.001
RV FAC%	19±6	30±7	0.0001	21.1±9	30±10	0.003
indexed RA volume	39±15	35±20	0.184	60±42	35±20	0.02
TAPSE Z score	-4.7±2.4	-3.4±2.5	0.044	-3.6±3.4	-4.2±2.1	0.44
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	0.01

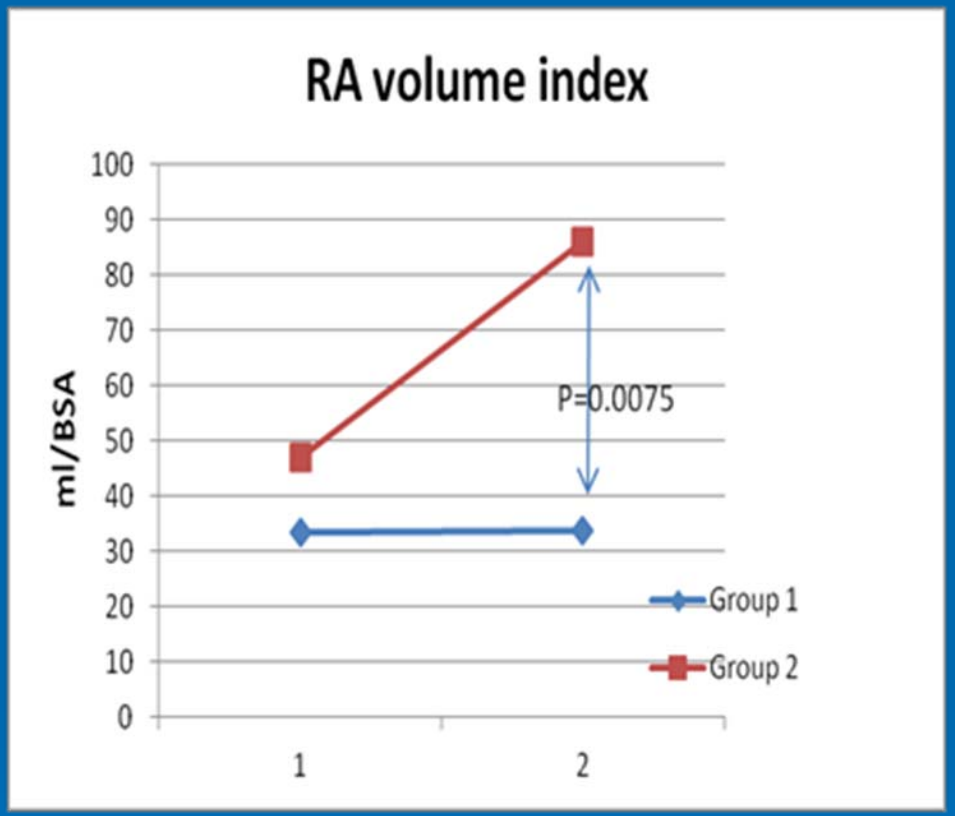
Changes in indices of RV function from presentation to last echo in iPAH patients in group 1 compared to group 2.

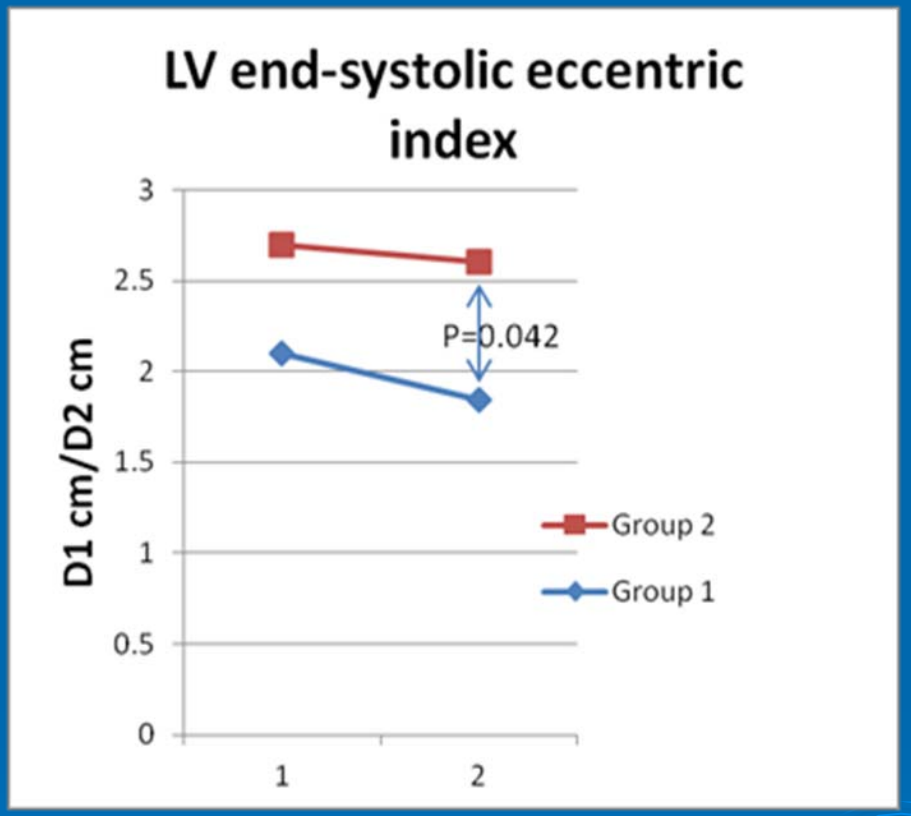




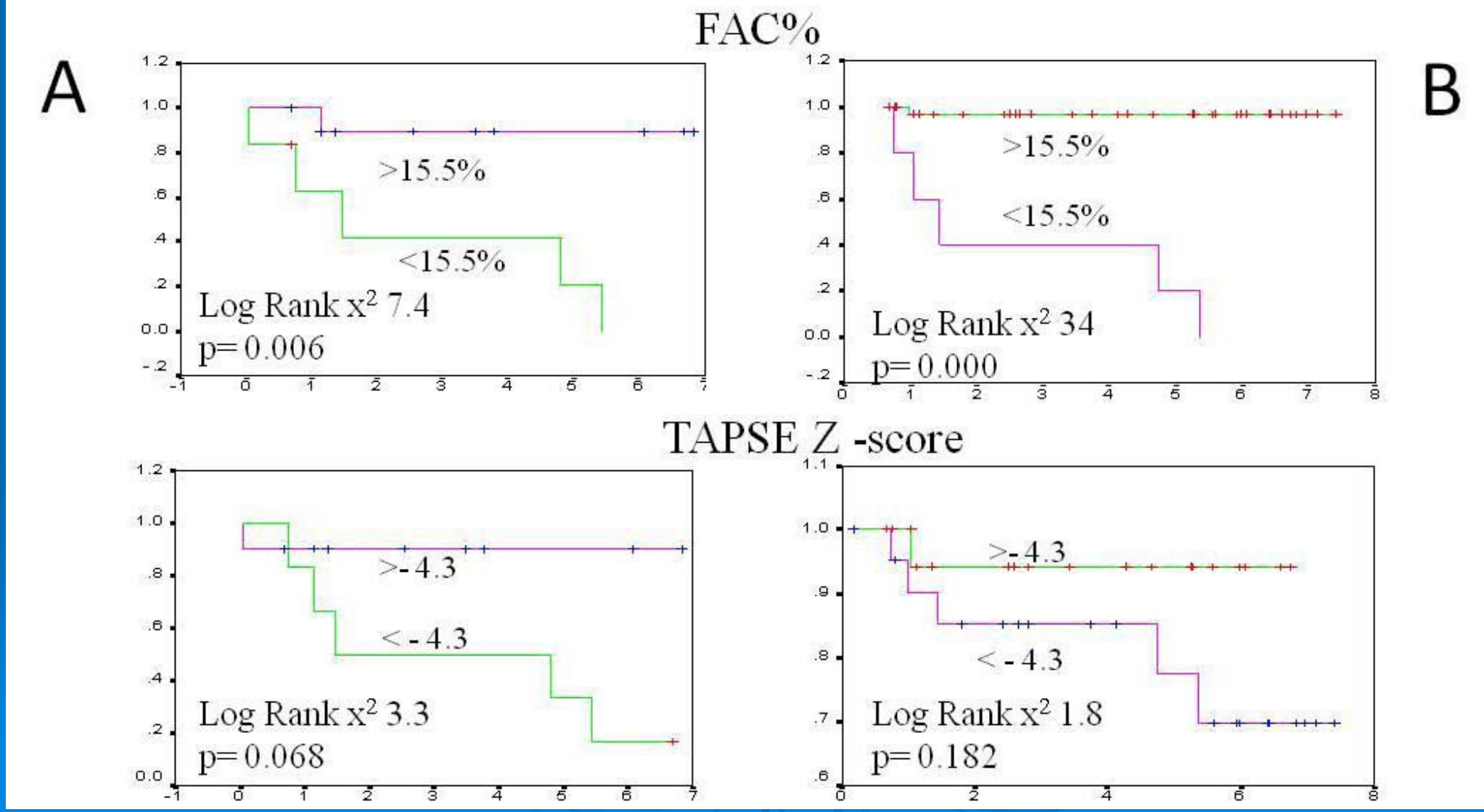




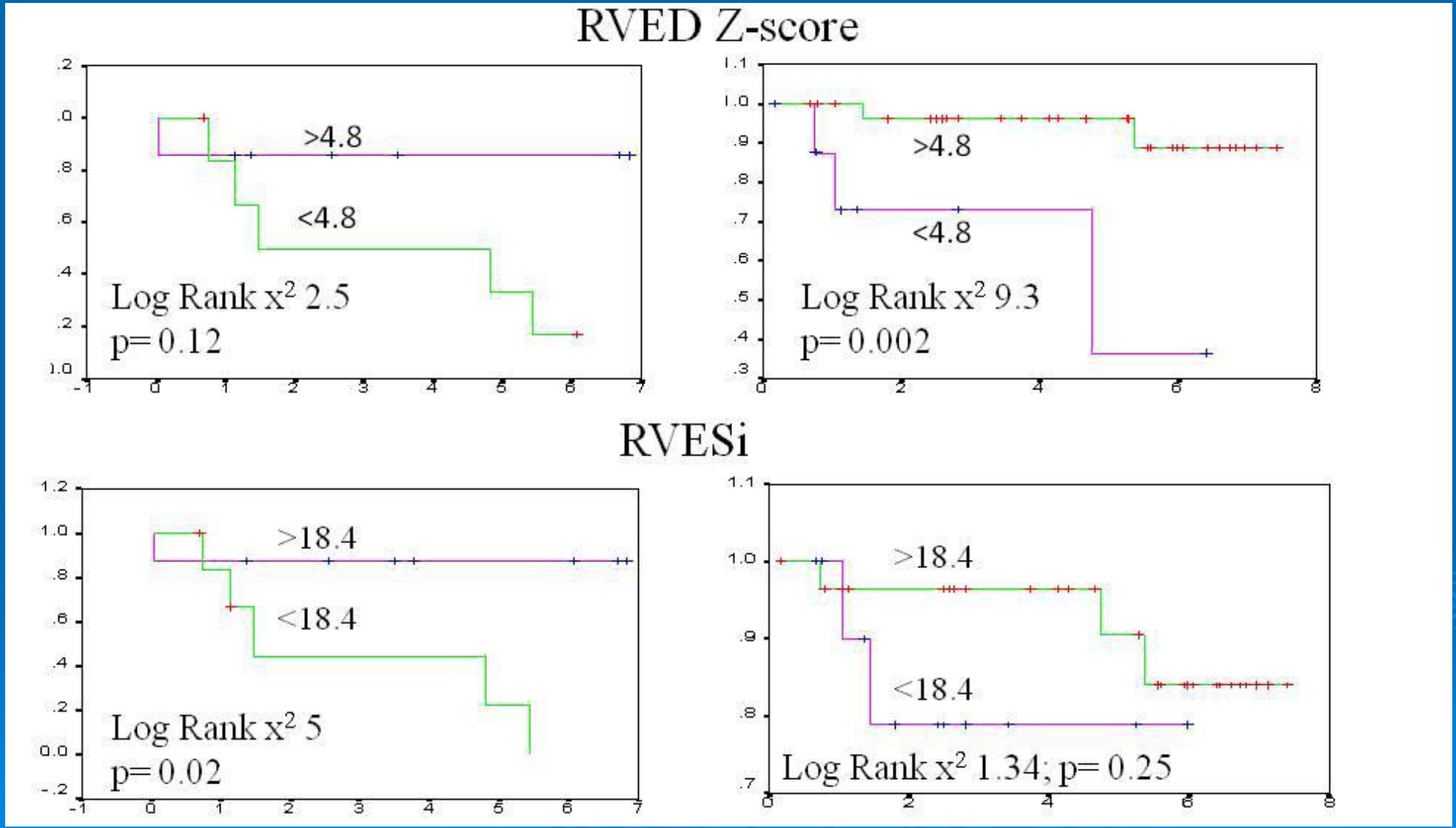

RA volume index
ml/BSA
100
90
80
70
60
50
40
30
20
10
0
1
2
P=0.0075
Group 1
Group 2


LV end-systolic eccentric index
D1 cm/D2 cm
3
2.5
2
1.5
1
0.5
0
1
2
P=0.042
Group 2
Group 1

Kaplan-Meier probability of survival stratified by RV function. Column A depicts survival for iPAH patients. Column B depicts survival in all patients



Kaplan-Meier probability of survival stratified by RV function. Column A depicts survival for iPAH patients. Column B depicts survival in all patients



Conclusion

Echo parameters of RV function were worse in iPAH patients compared to cPAH patients.

RV function was similar between the two groups of iPAH at presentation but worsened in patients who died or were transplanted.

תודה

איאס קאסם