

Tricuspid annular peak systolic velocity (TAPSV) in children and young adults with pulmonary artery hypertension secondary to congenital heart disease and tetralogy of Fallot: comparison with MRI data

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Aims:

The tricuspid annular peak systolic velocity (TAPSV), as a new echocardiographic index to assess right ventricular (RV) systolic function, has not been investigated thoroughly in children and young adults with tetralogy of Fallot (TOF) and pulmonary artery hypertension secondary to congenital heart disease (PAH-CHD). Ghio et al* have shown that the TAPSV is a reproducible index of systolic RV function in patients with CHD. One aim of this study was to determine TAPSV values in pediatric patients with RV volume overload (TOF), and severe pressure overload (PAH-CHD), and to compare with age-matched normal values in a cross-sectional study design. Second aim was to compare the TAPSV values with the RV ejection fraction (EF) and RV indexed end-diastolic volume (EDVi) values measured by MRI.

Methods:

TAPSV values of patients with TOF (n=185) and PAH-CHD (n=55) (Table 1) were compared to age-matched normal subjects. TAPSV values were compared to RVEF and RVEDVi determined by MRI. Guided by the 4-chamber view, a 5 mm sample volume was placed at the lateral corner of tricuspid annulus (Figure 1). Peak annular velocities during systole were recorded and analyzed off-line.

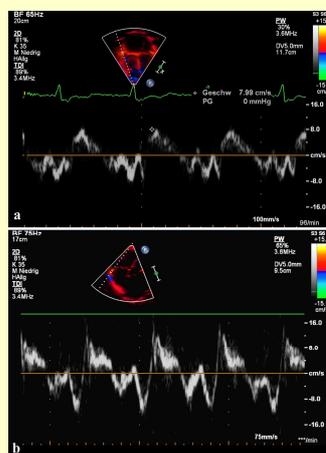


Figure 1: Apical 4-chamber view. The white broken line indicate M-Mode cursor placement at the tricuspid lateral annulus. Representative image of the tissue Doppler measured TAPSV in a 18 year old PAH-CHD patient (Figure 1a), and in a 16 year old healthy adolescent (Figure 1b).

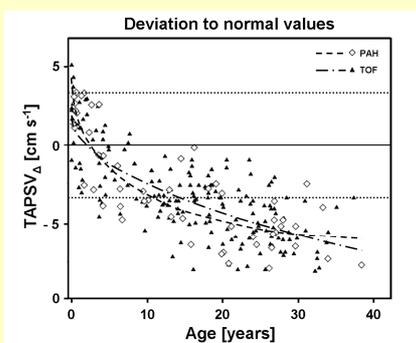


Figure 2: Deviation of TAPSV values (TAPSV_Δ) in patients with PAH-CHD, and TOF from mean reference values versus increasing time. The TAPSV_Δ value data points for PAH-CHD, and TOF patients are given as diamonds, and triangles, respectively. The interpolated mean values of control group are given as the black thin line. The -2 standard deviation line (-2SD) of the control group measurements is given as a black smooth-dashed line. The absolute deviation of the measured mean TAPSV values of our patients compared to age-related TAPSV reference values (interpolated black thin line) is demonstrated.

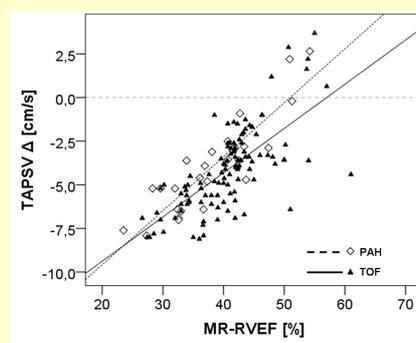


Figure 3: Relationship between TAPSV values and RVEF in PAH-CHD and TOF patients. TAPSV_Δ value data are given as diamonds for PAH-CHD patients and triangles for TOF patients, respectively. Deviation of TAPSV_Δ values in patients with PAH-CHD and TOF from mean age-related reference values is shown. The horizontal broken line at 0 indicates normal TAPSV values. The difference of TAPSV_Δ values to reference values is expressed in centimetre (cm/s).

Demographic Data: PAH-CHD Patients	
PAH-CHD patients	(n)
Female	51
Age, mean	26.24
Age range	15-54
BSA, mean	2.01
BSA range	1.51-2.68
NYHA	(n)
Class I	26
Class II	12
Time of surgical repair	(months)
Mean	5.4
Range	0-15.8
Diagnosis	
Atrial	(n)
AVSD	16
PA with VSD	11
TAPSV	(n)
TOF	5
Treatment	
Diuretic	(n)
Diuretic + Sildenafil	11
Sildenafil	17
Calcium Antagonist	6
Eurosteroid	(n)
Diuretic	21
Echocardiography	
TAPSV Data available	(n)
Male	46
Female	39
MRI Data	
PAH patients	(n)
Female	37
PAH + TOF patients	147
Female	87
RVEF > 50% (mean)	(%)
Range	47-78
RVEF < 50% (mean)	(%)
Range	18-63
Range	9-91
< 30% (n)	5
< 30% (n)	11
< 30% (n)	4
ECG	
PAH patients	(n)
Mean ± SD (ms)	126 (20)
QRS duration	Mean ± SD (ms)
Range (ms)	80-205
QRS duration > 180 ms	(n)
(% of measured)	4
Right Heart Catheterization	
mPAP	(Mean ± Range, mm)
Mean ± Range, WJL	38.2 (20-55)
Mean ± Range, WJL	33 (21-48)
Mean ± Range, WJL	35.4 (9-49)
Demographic Data: TOF Patients	
TOF Patients	(n)
Female	104
Age, mean	12.24
Age range	5-34
BSA, mean	1.18
BSA range	0.71-1.91
NYHA	(n)
Class I	52
Class II	20
Time of surgical repair	(months)
Mean	33.1
Range	0-104
Echocardiography	
TAPSV Data available	(n)
Female	103
Mean ± SD (cm/s)	33.4
MRI Data	
TOF patients	(n)
Female	114
PAH + TOF patients	116
Female	63
RVEF > 50% (mean)	(%)
Range	47-78
RVEF < 50% (mean)	(%)
Range	18-63
Range	9-91
< 30% (n)	5
< 30% (n)	11
< 30% (n)	4
ECG	
TOF patients	(n)
Mean ± SD (ms)	113 (20)
QRS duration	Mean ± SD (ms)
Range (ms)	75-210
QRS duration > 180 ms	(n)
(% of measured)	3

Table 1: Demographic data of our TOF and PAH-CHD patients.

Results:

TAPSV values become significantly reduced after an age of 10.4 years in PAH-CHD patients, and after 13.6 years in TOF patients when compared to the lower bound of the ± 2 SD interval of normal subjects (Figure 2). A significant positive correlation between TAPSV with RVEF was seen in both, TOF ($r = 0.66$, $p < 0.001$) and PAH-CHD ($r = 0.82$, $p < 0.001$) patients (Figure 3). A significant negative correlation between TAPSV with RVEDVi was seen in TOF ($r = -0.29$, $p = 0.002$) as well as in PAH-CHD patients ($r = -0.59$, $p < 0.001$).

Discussion:

Little is known of systolic RV function in PAH-CHD in the pediatric age group. Because of the preserved RV systolic function in our infants and young children with PAH-CHD we assume that they were in the stage of "adaptive hypertrophy" as described by Bogaard et al**. TAPSV values of patients with PAH-CHD become significantly reduced compared to age-matched controls after an age of 10.4 years, i.e. the longer the RV suffers from severe pressure overload the more depressed systolic RV function becomes. This might be explained by the fact that the RV is not capable to sustain high long-term pressure overload**. The increased RVEDVi can be explained by the fact that with increasing afterload the ventricular septum becomes flat and eventually inverts into the LV cavity with time.

Conclusion:

Although initially preserved, we found impaired TAPSV values with increasing age in patients with TOF and PAH-CHD. This indicates that persistent pressure overload in PAH-CHD patients as well as volume overload in TOF patients can lead to an impairment of systolic RV function and increased RVEDVi. The validity of TAPSV data could be confirmed by MRI data (RVEDVi and RVEF).

References:

- * Ghio S, et al. Prognostic relevance of the echocardiographic assessment of right ventricular function in patients with idiopathic pulmonary arterial hypertension. Int J Cardiol 2010; 140: 272-8.
- ** Bogaard et al. The right ventricle under pressure: cellular and molecular mechanisms of right-heart failure in pulmonary hypertension. Chest 2009; 135: 794-804.