Published in final edited form as:

J Am Soc Echocardiogr. 2006 January ; 19(1): 21–27.

Use of Myocardial Performance Index in Pediatric Patients with Idiopathic Pulmonary Arterial Hypertension

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Abstract

Background—The myocardial performance index (MPI) correlates with clinical status in adults with idiopathic pulmonary arterial (PA) hypertension (IPAH). This pediatric study used MPI to assess response to bosentan therapy.

Methods—The study included 12 children with IPAH and 12 healthy control subjects. MPI was correlated with catheterization data at initiation of bosentan and at a median follow-up of 9 months. Therapy responders were defined by a greater than 20% decrease in mean PA pressure.

Results—Right ventricular MPI for patients with IPAH was 0.64 ± 0.30 versus 0.28 ± 0.03 in control subjects ($P < .01$). It had a strong correlation with mean PA pressure ($R = 0.94$; $P < .001$). Right ventricular MPI decreased significantly in responders (range 20%–44%, mean 25%) with a 5 % increase in nonresponders.

Conclusions—Right ventricular MPI in pediatric IPAH correlates with mean PA pressure and response to therapy. This study suggests that this noninvasive Doppler index may be useful to follow up children with IPAH, particularly when tricuspid regurgitation data are insufficient.

Idiopathic pulmonary arterial (PA) hypertension (IPAH), previously known as primary pulmonary hypertension, is a rare but potentially fatal disease that affects adults and children. It is defined by a mean PA pressure (MPAP) greater than 25 mm Hg at rest in the absence of an underlying cause.¹ Historically, IPAH carried a dismal prognosis with a median survival of 10 months in children, mainly as a result of right heart failure.² In recent years, new treatment options have dramatically improved outcome, with some patients surviving more than 5 to 10 years from diagnosis.^{3,4} This improved outcome is directly linked to a better understanding of the pathogenesis and genetics of IPAH, which has led to novel treatment strategies.^{5–9} These advances in treatment further mandate reliable quantitative methods to evaluate right ventricular (RV) function and response to treatment.

The myocardial performance index (MPI) is a noninvasive Doppler measurement of global ventricular function that incorporates both systolic and diastolic function and may be applied to the RV or left ventricle (LV). MPI (or Tei index) is defined as ratio of the sum of the isovolumic contraction time (IVCT) and isovolumic relaxation time (IVRT) divided by the systolic ejection time (ET).¹⁰ The index is easily measured with high reproducibility and is important in the assessment of global RV performance as the active energy cycles of contraction and relaxation occur during IVCT and IVRT.¹¹ RV MPI has been correlated with RV ejection

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fraction measured by cardiac magnetic resonance imaging for patients whose RV functions as the systemic ventricle.¹²

The goals of this study were to: (1) assess reliability and reproducibility of measurements of RV and LV MPI in a pediatric population with IPAH; (2) determine the relationship between RV MPI and MPAP measured at cardiac catheterization; and (3) evaluate the sensitivity of RV and LV MPI to changes in MPAP in response to therapy for IPAH.

METHODS

Study Population

This study includes 12 children diagnosed with IPAH at The Children's Hospital, Denver, Colo, based on clinical and hemodynamic criteria¹³ who were being evaluated for initiation of bosentan, an oral vasodilator. Secondary causes of pulmonary hypertension, such as significant intracardiac shunt, pulmonary disease, thromboembolic disorders, and connective tissue disease, were previously excluded. A group of 12 age-matched healthy children undergoing echocardiograms at our institution served as the control subjects. The reasons for echocardiography in the control group included murmur, chest pain, or palpitations (found later to be isolated premature atrial contractions or sinus arrhythmia). None of the control subjects had structural heart disease or elevated PA pressures estimated by tricuspid regurgitation (TR) jet velocity. Approval for this study was obtained from the institutional review board and all patients or their guardians gave informed consent and assent when indicated, to participate in this study.

Echocardiographic Examination

Complete 2-dimensional, spectral Doppler, and color flow Doppler examinations were performed on all patients and healthy control subjects (Vivid 5, GE Vingmed, Milwaukee, Wis). Doppler measurements were performed using 3.5-, 5-, or 8-MHz transducers, as appropriate for patient size. Echocardiographic studies were obtained within 24 hours before the cardiac catheterizations performed at the patient's baseline and at follow-up after initiation of bosentan. Oral bosentan was started after the baseline cardiac catheterization was completed. Only patients in sinus rhythm were included in this study. A 12-lead electrocardiogram was also recorded and reviewed for the presence of ventricular conduction delays (QRS duration > 100 milliseconds). Velocities of TR and pulmonary valve regurgitation were quantified by spectral Doppler flow imaging. Assessment for low velocity and bidirectional atrial level shunts was performed with narrow imaging sectors and careful adjustments of the color Doppler aliasing velocities from subcostal views.

Doppler tracings were recorded using a sweep speed of 50 to 100 mm/s. The tricuspid inflow velocity was recorded from the apical 4-chamber view with the pulsed wave Doppler sample volume positioned at the tips of the tricuspid leaflets. Continuous wave Doppler recording of the TR signal was obtained from the apical 4-chamber and parasternal RV inflow views. Doppler-derived peak systolic RV pressure was then estimated from the peak TR velocity using the simplified Bernoulli equation without correcting for right atrial pressure.¹⁴ The RV outflow tract velocity was recorded from the parasternal short-axis view with the Doppler sample volume positioned just below the pulmonary valve. Measurement of LV MPI was made from the apical 4-chamber view with the cursor placed between the mitral valve inflow and LV outflow. A simultaneous electrocardiogram was obtained in all patients. All studies were recorded digitally for later offline analysis.

Doppler Measurement of MPI

Doppler time intervals were measured offline using echocardiographic raw data stored in a digital format (Echopac, GE Vingmed). Figure 1 shows the calculation of the RV MPI. Interval “a” is measured from cessation to onset of tricuspid inflow and is equal to the sum of the RV IVCT, ET, and IVRT. RV ET (interval b) is measured from the onset to the end of the RV outflow velocity trace. RV MPI was defined as the sum of IVCT and IVRT divided by the ET and was calculated as: $(a - b/b)$ as described by Tei et al.^{10,15} Because interrogation of the tricuspid valve inflow and the RV outflow is obtained from different echocardiographic views, care was taken to select traces with R-R intervals within 5% in duration from each other. LV MPI was calculated by measuring IVCT and IVRT from an apical 4-chamber view that displayed the mitral valve inflow and LV outflow during the same cardiac cycle. To avoid observer bias, these measurements were made at a different time from the echocardiographic assessment described above, blinded to patients’ names and diagnoses. Five consecutive beats were measured and averaged for each measurement.

Cardiac Catheterization

All patients underwent cardiac catheterization before and at a mean follow-up of 9 months after starting bosentan therapy. One patient was under general anesthesia (patient 9) for both catheterization studies; the remaining 11 were under conscious sedation. PA pressures were measured using standard fluid-filled catheters. For patients not on supplemental oxygen therapy at the time of the study, MPAP obtained on room air was used for comparison with the echocardiographic RV MPI. For those patients already receiving supplemental oxygen, the MPAP measured on their baseline amount of oxygen was used for comparison to RV MPI. All 12 patients underwent acute vasoreactivity testing per protocol during both initial and follow-up catheterizations.¹⁶ Cardiac output was measured by the thermodilution method in those without intracardiac shunts and by Fick’s method in those with a patent foramen ovale.

Statistical Analysis

All data are expressed as mean value and SD. Student *t* test was used for 2-way comparison of clinical characteristics between patients and control subjects. Correlation between RV MPI and MPAP was determined by linear regression analysis. The RV MPI was also correlated to age, heart rate, and body surface area for patients and control subjects. Statistical significance was defined as a *P* value less than .05. Intraobserver and interobserver variability was assessed by blinded repeated measurements. Intraobserver variability was based on measurements made by the same observer at different times assessing the same 5 beats and Bland-Altman analysis was performed.¹⁷ Interobserver variability was based on measurements made by different observers (K. L. D. and B. D.), blinded to each other’s results, assessing the same 5 beats.

RESULTS

Clinical Patient Characteristics

The clinical characteristics of the 12 children with IPAH and 12 age-matched control subjects are summarized in Table 1. The clinical profile of each of the 12 patients is illustrated in Table 2. The mean age at diagnosis was 3.5 ± 3.4 years (range: 1 month-10 years). The mean QRS duration for all patients was 90 ± 18 milliseconds. Ventricular conduction delays (QRS duration > 100 milliseconds) were present in 2 of 12 patients (Table 2). None of the patients or control subjects had a complete right bundle branch block. Selected echocardiographic findings on the 12 patients with IPAH are summarized in Table 3. The hemodynamic measurements and calculated data are shown in Table 4.

MPI

Baseline RV and LV MPI values were elevated in the IPAH group compared with the control group (Figure 2). The RV MPI for the IPAH group was elevated at 0.64 ± 0.30 compared with 0.28 ± 0.03 for healthy age-matched control subjects ($P < .01$). The LV MPI was also elevated at 0.44 ± 0.15 for patients compared with 0.34 ± 0.03 in healthy control subjects ($P < .05$). Figure 3, A, shows the relationship of MPAP obtained at catheterization and RV MPI within 24 hours of the catheterization before treatment with bosentan. There was a strong correlation between the two variables ($R = 0.94$; $P < .01$).

Response to Treatment

The RV MPI reflected response to treatment. Follow-up data are summarized in Table 4 and Figure 3, B. Based on the change in MPAP at follow-up catheterization, patients were divided into two groups, responders and nonresponders. Responders were defined as those patients who had a 20% decrease in MPAP between the initial and follow-up catheterization. All 4 patients who responded to bosentan with a greater than 20% decrease in MPAP also had an improvement in RV MPI, which decreased by a mean of 25% (range: 20%–44%). The 8 nonresponders had a mean increase in RV MPI of 5% (range: 0%–43%). It is interesting to note that one patient with severe IPAH and a borderline decrease in MPAP of 18% at follow-up catheterization did not have a significant change in RV MPI (patient 9). The other patient with ventricular conduction delay on the electrocardiogram (patient 12 in Tables 2 and 4) improved with treatment.

The LV MPI did not change significantly with therapy. The LV MPI values for responders were 0.45 ± 0.25 before versus 0.44 ± 0.02 after (not significant). The LV MPI values for nonresponders were 0.42 ± 0.12 before compared with 0.41 ± 0.10 after (not significant).

Correlations with Physiologic Parameters

The relationship of RV and LV MPI to age, heart rate, or body surface area was assessed for patients and healthy control subjects. There was no statistically significant correlation.

Reproducibility

Interobserver and intraobserver variability were assessed using Bland-Altman analysis (Figure 4). The mean percentage variation for intraobserver measurements was less than 10%. The mean percentage variation in interobserver measurements was less than 15%. The LV MPI reproducibility results were similar with less than 10% variation for intraobserver variability and less than 15% for interobserver variability.

DISCUSSION

This study demonstrates that the MPI for the RV and LV can be measured reliably in children with IPAH with acceptable reproducibility. We found that RV MPI is elevated in pediatric patients with IPAH compared with age-matched control subjects. Importantly, among patients with IPAH, RV MPI correlated with invasive measurements of MPAP and trends in RV MPI correlated with trends in MPAP. These findings support the value of MPI in the clinical monitoring of these patients. The advantages of MPI over TR velocity are 2-fold: (1) not all patients with pulmonary hypertension have sufficient TR for accurate measurement of the peak jet; and (2) MPI appears to correlate with MPAP, which was chosen to define this disease because it is more reliable than the peak instantaneous PA pressure derived from the TR jet velocity.¹

In adults with IPAH, RV MPI has been shown to predict outcome.^{18,19} It is useful in assessing the effects of prostacyclin therapy.²⁰ Our findings correspond to the experimental study by

Sugiura et al²¹ who demonstrated a significant positive linear correlation between the RV MPI and changes in MPAP in neonatal piglets with hypoxic pulmonary hypertension. Other studies have shown that the MPI correlates with invasive measures of both RV and LV function^{22, 23} and is useful in the follow-up of patients with cardiomyopathy.^{24,25} Studies in children with RV volume and pressure overload have demonstrated the feasibility of measuring this index in the pediatric age group.^{26,27}

The use of time intervals to detect and monitor pulmonary hypertension is well established²⁸ and particularly useful because they are not limited by the geometric shape of the ventricle, a fact that is important in these patients who usually have distorted ventricular geometry. Several studies have documented the use of the pre-ejection period, ET, and acceleration time for patients with pulmonary hypertension.^{29–31} The MPI also includes changes of the IVRT, a diastolic time interval. The systolic and diastolic time intervals are easily obtained by routine Doppler techniques during standard echocardiographic examination and are simple to measure, with excellent reproducibility. In addition, MPI is independent of heart rate.³²

All patients in this study who had an improvement in MPAP of 20% or more in response to vasodilator therapy also had an improvement in RV MPI. One patient with severe elevation of PA pressure had a decrease in MPAP of 18% between initial and follow-up catheterization, without an improvement of the RV MPI. A possible explanation could be permanent loss of RV function secondary to the severe pulmonary hypertension and chronic high afterload. In this case, the reduction in MPAP could be the result of remodeling in the pulmonary vasculature whereas the MPI could reflect persistent RV dysfunction. Our data cannot answer the question of how right bundle branch block would affect this marker because none of the patients had significant RV conduction delay.

The relationship of RV MPI to ventricular loading conditions remains unclear. Eidem et al²⁶ demonstrated that RV MPI was increased for patients with combined pressure and volume overload, and did not change after intervention to relieve the volume overload. In addition, data from a porcine model also supported relative load independence of this index.³³ However, more recently, Cheung et al³⁴ presented experimental data to suggest significant alterations of the MPI in response to acute changes of ventricular loading conditions. In light of these data, it is possible that the abnormal RV MPI in our children with pulmonary hypertension at least in part reflected altered RV afterload. It has been difficult to study the response of the RV to changes in loading conditions although it is clear that it is significantly different from that of the LV.³⁵ In children, the noninvasive quantitation of RV function has been particularly difficult because magnetic resonance imaging often requires general anesthesia and radionuclide studies are hindered by the need for sedation, limited resolution, and radiation exposure.^{36–38}

Our data support that the RV MPI is a sensitive tool to estimate MPAP in children with IPAH and to monitor response to therapy. No conclusions can be made regarding the role of RV dysfunction in our observations. Further studies with longer follow-up periods will be beneficial to further delineate the use of MPI in predicting prognosis in pediatric patients with IPAH.

Limitations

The primary limitation of this study is the small size of the study group. However, the population is highly selected to patients with IPAH, without other disease. Another limitation is the fact that the follow-up catheterizations were performed at different times after initiation of vasodilator therapy. The RV MPI appears to agree well with changes in MPAP for both responders and nonresponders but, because the relatively short follow-up time and the small

number of patients, no comment can be made regarding the possible role of ventricular remodeling on MPI measurements.

Conclusion

The MPI provides an easily obtainable and noninvasive tool for monitoring MPAP in pediatric IPAH. RV MPI is independent of quantity of TR and can be especially useful in those children without measurable TR. We propose that RV MPI may be a useful adjunct in the clinical assessment of pediatric patients with IPAH.

Acknowledgements

We would like to thank Mr Scott Kirby, RCDS, and the staff of the Cardiac Imaging Laboratory at The Children's Hospital for their help in collecting the data.

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Figure 1.

Calculation of right ventricular (RV) myocardial performance index (MPI). Interval “a” (from onset of isovolumic contraction to onset of diastolic filling) was determined from tricuspid inflow trace (*left*). Interval “b” (systolic ejection time) was measured from RV outflow trace (*right*). RV MPI was calculated as: $MPI = (a - b)/b$.

Figure 2. Difference between right and left ventricular myocardial performance index (*MPI*) in idiopathic pulmonary arterial hypertension (*IPAH*) group compared with normal (*NL*) group (* $P < .05$).

Figure 3.

Right ventricular (RV) myocardial performance index (MPI) correlated with mean pulmonary artery pressure (MPAP) obtained at initial (*pre*) (A) and follow-up (*post*) (B) cardiac catheterization.

Figure 4.
Bland-Altman plots of intraobserver and interobserver variability indicate good reproducibility over broad range of values.

Table 1

Clinical characteristics of 12 children with idiopathic pulmonary arterial hypertension and 12 control subjects

	Patients (n = 12)	Control subjects (n = 12)	<i>P</i>
Age, y	9.6 ± 4.8	9.2 ± 4.3	NS
Weight, kg	35.9 ± 18	36.9 ± 15.6	NS
Height, cm	132 ± 25	138 ± 24	NS
Body surface area, m ²	1.13 ± 0.39	1.18 ± 0.36	NS
Heart rate, beats/min	94 ± 16	88 ± 15	NS
Systolic blood pressure, mm Hg	99 ± 13	104 ± 12	NS
Fractional shortening, %	39 ± 5	38 ± 6	NS

NS, Not significant.Means and SD, Student *t* test.

Table 2

Clinical profile for 12 children with idiopathic pulmonary arterial hypertension

Case	Age, y	Age at diagnosis, y	QRS duration, ms	Medications at baseline	Time interval between start of bosentan and follow-up catheterization, mo
1	4	4	62	Oxygen	4
2	7	0.8	83	Prostacyclin, dipyridamole, warfarin	3
3	15	0.25	93	Oxygen, treprostinil, warfarin, dipyridamole, furosemide	17
4	8	0.10	90	Prostacyclin, warfarin	19
5	15	0.3	96	Warfarin	12
6	14	9	94	Prostacyclin, oxygen, clopidogrel, furosemide, bumetadine	3
7	6	5	83	Prostacyclin, warfarin	3
8	5	2	98	Furosemide, digoxin, L-arginine	3
9	11	10	124	Prostacyclin, oxygen, digoxin, furosemide, warfarin	12.5
10	13	5	70	Prostacyclin, warfarin, dipyridamole, nifedipine	18
11	2	2	72	Oxygen, warfarin	12
12	15	1	111	Dipyridamole, warfarin	3

Table 3

Echocardiographic data of 12 patients with idiopathic pulmonary arterial hypertension

Echocardiographic characteristic	Responders (n = 4)		Nonresponders (n = 8)	
	Pre	Post	Pre	Post
Peak TR velocity, m/s	4.1	3.2	3.8	3.7
Atrial level shunt				
None	4	4	4	4
Left to right	0	0	1	1
Bidirectional	0	0	2	2
Right to left	0	0	1	1

TR, Tricuspid regurgitation.

Table 4

Myocardial performance index and cardiac catheterization data

Case	Baseline						Follow-up					
	MPAP, mm Hg	SPAP (% SBP)	PVRI U × m ²	PVR/ SVR	CI, L/ min/ m ²	RV MPI	MPAP, mm Hg	SPAP (% SBP)	PVRI U ×m ²	PVR/ SVR	CI, L/ min/ m ²	RV MPI
1	63	88 (80)	15.58	0.68	3.53	0.83	44	70 (75)	11.78	0.56	2.88	0.72
2	39	53 (53)	6.63	0.39	4.39	0.47	35	54 (49)	6.52	0.46	4.06	0.47
3	60	90 (90)	11.1	0.71	4.95	0.70	62	68 (82)	13.85	0.81	2.44	0.71
4	26	36 (39)	5.60	0.27	4.86	0.37	24	38 (51)	3.09	0.31	3.19	0.34
5	30	38 (35)	3.48	0.23	3.41	0.34	39	52 (49)	6.03	0.34	3.82	0.33
6	66	100 (83)	12.96	0.71	4.92	0.81	58	80 (73)	7.95	0.66	5.32	0.69
7	56	79 (77)	11.3	0.69	4.34	0.72	27	40 (42)	4.27	0.29	4.45	0.4
8	73	92 (86)	32.03	0.76	2.22	1.16	57	79 (83)	11.74	0.72	4.26	0.87
9	90	120 (133)	67.06	2.3	2.27	1.13	74	96 (123)	17.86	1.45	3.02	1.15
10	27	38 (37)	6.73	.31	4.46	0.35	30	42 (41)	7.15	0.28	3.15	0.36
11	38	51 (89)	8.78	0.82	3.46	0.44	55	71 (85)	11.33	0.80	5.39	0.63
12	35	48 (45)	4.76	0.25	3.99	0.38	20	35 (32)	3.66	0.17	3.72	0.32

CI, Cardiac index; MPAP, mean pulmonary artery pressure; MPI, myocardial performance index; N, nonresponder; PVR, pulmonary vascular resistance; PVRI, PVR indexed; R, responder; RV, right ventricular; SBP, systolic blood pressure; SPAP, systolic pulmonary artery pressure; SVR, systemic vascular resistance; SVRI, SVR indexed.