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Pulmonary arterial compliance in patients of CHD with increased pulmonary blood flow

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Abstract

Introduction: Pulmonary arterial compliance, the dynamic component of pulmonary vasculature, remains inadequately studied in patients with left to right shunts. We sought to study the pulmonary arterial compliance in patients with left to right shunt lesions and its utility in clinical decision-making. *Materials and methods:* In this single-centre retrospective study, we reviewed cardiac catheterisation data of consecutive patients of left to right shunt lesions catheterised over one year. In addition to the various other parameters, pulmonary arterial compliance was calculated, as indexed pulmonary flow (Qpi) / (Heart rate × pulse pressure in the pulmonary artery). RC time was also calculated, as the product of pulmonary arterial compliance and pulmonary vascular resistance index. Patients were divided into "operable," "borderline," and "inoperable" based on the decision of the treating team, and the pulmonary arterial compliance values were evaluated in these groups to study if it can be utilised to refine the operability decision. *Results:* 298 patients (Median age 16 years, 56% <18 years) with various acyanotic shunt lesions were included. Overall, the pulmonary arterial compliance varied with Qpi, pulmonary artery mean pressure, and pulmonary vascular resistance index, but did not vary with age, type of lesion, or transpulmonary gradients. The median pulmonary arterial compliance in patients with normal pulmonary artery pressure (Mean pulmonary artery pressure less than 20 mmHg) was 4.1 ml/mmHg/m^2 (IQR 3.2). The median pulmonary arterial compliance for operable patients was 2.67 ml/mmHg/m^2 (IQR 2.2). Median pulmonary arterial compliance was significantly lower in both inoperable (0.52 ml/mmHg/m^2, IQR 0.34) and borderline (0.80 ml/mmHg/m^2, IQR 0.36) groups when compared to operable patients ($p < 0.001$). A pulmonary arterial compliance value lower than 1.18 ml/mmHg/m^2 identified inoperable patients with high sensitivity and specificity (95%, AUC 0.99). However, in borderline cases, assessment by this value did not agree with empirical clinical assessment.

The median RC time for the entire study population was 0.47 S (IQR 0.30). RC time in operable patients was significantly lower than that in the inoperable patients (Median 0.40 IQR 0.23 in operable, 0.73 0.25 in inoperable patients ($p < 0.001$). *Conclusions:* Addition of pulmonary arterial compliance to the routine haemodynamic assessment of patients with shunt lesions may improve our understanding of the pulmonary circulation and may have clinical utility.

Pulmonary circulation is a high-volume, low-pressure system with the right ventricle as a flow generator. In patients with congenital acyanotic shunt lesions with increased pulmonary blood flow, this system is overburdened with either increased flow, increased pressure, or both.

The pulmonary circulation has been characterised under three components, viz. pulmonary vascular resistance index, pulmonary arterial compliance, and impedance. While the pulmonary vascular resistance index represents the static right ventricular afterload, impedance and compliance together constitute the dynamic component of the afterload.[1]

Our assessment of pulmonary vasculature in patients with acyanotic heart disease and increased pulmonary blood flow (Qp) is largely based on the calculation of pulmonary vascular resistance index.[2,3] While a few studies have calculated pulmonary arterial compliance values in humans,[1,4–7] pulmonary arterial compliance has not been adequately studied in patients with shunt lesions, nor its utility in clinical decision-making been explored. The product of pulmonary arterial compliance and pulmonary vascular resistance index, the RC time, has also not been well studied in patients with shunt lesions. We studied the pulmonary arterial compliance in large number of patients with acyanotic shunt lesions with increased pulmonary blood flow with a view to evaluate the range of pulmonary arterial compliance at all ages and also studied its relation to other parameters. We evaluated the pulmonary arterial compliance values in operable, borderline, and inoperable patients as decided by the clinicians previously. In addition, the RC time was calculated in all the patients and similarly analysed.

Materials and methods

We retrospectively evaluated records from consecutive patients with acyanotic shunt lesions with increased Qp who underwent right heart catheterisation during the study period, that is from 01 January to 31 December, 2019. Patients with a concomitant lung disease, surgical interventions in the pulmonary vasculature, or anatomic abnormality of the pulmonary artery, for example branch pulmonary artery stenosis, hypoplastic pulmonary arteries, patients not in sinus rhythm at the time of study, patients with severe right ventricular dysfunction, and patients with incomplete records, were excluded.

Ethical approval was taken from the institutional ethics committee, and all the patient-related data were collected anonymously.

Cardiac catheterisation was done under conscious sedation as per routine; the baseline haemodynamic data were collected in all patients. Oxygen consumption was assumed using the published charts;[8] vasoreactivity testing was done with 100% oxygen given by the face mask for 10 mins, when indicated. We have not included haemodynamic data with oxygen in this study.

Detailed clinical information was available for all the patients, and the haemodynamic data were analysed to calculate the following additional parameters retrospectively.

$$\mathrm{PCa} = \frac{\mathrm{QPi}}{\text{Heart Rate} \times \text{Pulmonary artery pulse pressure}}$$

$$\text{RC Time (S)} = \mathrm{PCa}(\mathrm{mlmmHg}^{-1}) \times \mathrm{PVRi}(\mathrm{mmHgL}^{-1}\,\mathrm{min}) \times 0.06$$

Equation 1: Formulae for calculation of various parameters

(Qpi = indexed pulmonary blood flow, HR = heartrate, PCa = Pulmonary arterial compliance)

Qpi measurement in our laboratory is based on the assumed values of oxygen consumption routinely. For patients where pulmonary artery wedge pressure was not measured, left ventricular end-diastolic pressure was taken as equal to mean wedge pressure for calculation of pulmonary vascular resistance index. Patients were classified into clearly operable, clearly inoperable, and with borderline operability, based on the final decision of the treating team. The decision regarding operability was based on a comprehensive clinical evaluation which included age of the patient, clinical details, and haemodynamic parameters. Operability decisions were guided by the conventionally used criteria.[9] Patients with raised pulmonary vascular resistance index > 6 unit but persistent significant left to right shunts were considered borderline operability. Although vasodilatory testing with oxygen was done whenever the resting pulmonary vascular resistance index was high, but the operability decisions relied heavily on the basal haemodynamic data and a comprehensive clinical evaluation. All the contentious cases were discussed in the haemodynamic conference, and patients were labelled as operable, inoperable, or having borderline operability by the consensus of experienced cardiologists.

There are various methods of measurement of pulmonary arterial compliance.[10–12] We calculated pulmonary arterial compliance using the pulse pressure method[13,14] using formulae as above (equation 1), as this is a simple and accurate method of measurement of pulmonary arterial compliance.[13,15,16]

Statistical analysis

Data were analysed by Stata 11.2 (Stata Corp. 4905 College Station, Texas 47845, USA) and presented as mean ± SD, median (range), median (interquartile range), or frequency (percentage). Parameters of the individuals between operable and non-operable groups were compared using chi-square test or Fisher's exact test for categorical variables, independent *t*-test for variables following normal distribution, and Wilcoxon rank sum test for parameters following non-normal distribution. Spearman's correlation was used to correlate the continuous variables. Receiver operating characteristic curves were generated to identify the cut-offs for various parameters for association with operability. Univariate and stepwise multivariate logistic regression was used to calculate unadjusted and adjusted OR after assessing multicollinearity and mediators among the variables.

Results

Baseline characteristics

A total of 324 right heart catheterisation records were evaluated for inclusion. Of these, 26 were excluded (11 pulmonary artery stenosis, surgery or hypoplasia, 08 incomplete data, 04 non-sinus rhythm, 03 right ventricular dysfunction). Remaining 298 patients (148 males, M:F 0.99:1) were included. The median age of study patients was 16 years (range 2 months to 70 years, 167 patients (56%) less than 18 years). Atrial septal defect was the most common defect (n = 124, 41.6%), followed by patent ductus arteriosus (n = 95, 31.9%) and ventricular septal defect (n = 64, 21.5%). The patients were catheterised for diagnostic or therapeutic purposes (n = 168 for device closure, 130 diagnostic catheterisation). The baseline characteristics of study population are shown in Table 1. Overall, the median Qpi was 5.5 L/min/m^2 (IQR 4.18), the mean (±SD) of pulmonary artery mean pressure was 38.52(±24.35) mmHg, and median pulmonary vascular resistance index was 3.3 (IQR 5.84) Wood units m^2 (Table 1). Lesion-wise haemodynamic details of the patients are given in Supplementary Table 1.

Subgroups of congenital heart lesions (Supplementary Table 1)

The clinical characteristics including age, pulmonary artery pressures, and the magnitude of the shunts are shown in Supplementary Table 1. Amongst the subgroups, mean pulmonary artery pressure for the atrial septal defect patient subgroup was 30.03 mmHg and median pulmonary vascular resistance index was 2.57 Wood units m^2. Patients with patent ductus arteriosus had a mean pulmonary artery pressure of 31.68 mmHg and median pulmonary vascular resistance index of 3.0 Wood units m^2. The mean pulmonary artery pressure for ventricular septal defect subgroup was 61.6 mmHg and median pulmonary vascular resistance index 10.18 wood units m^2.

Pulmonary arterial compliance

Mean pulmonary arterial compliance for the overall study group was 2.88 ml/mmHg (±3.2) per m^2 (Median 2.13, IQR 2.1) (Table 2). The mean pulmonary arterial compliance for patients with a normal pulmonary artery pressures (Mean pulmonary artery pressure less than 20 mmHg) was 5.37 mm Hg (±5.05), median 4.1, IQR 3.2. Pulmonary arterial compliance was significantly lower in the inoperable group (mean 0.61 ± 0.36, Median

Table 1. Baseline characteristics of the study population.

Age (Years)	
Median (IQR)	16 (26)
Range	2 months – 70 years
Age 18 or less n (%)	167 (56)
Age >18 n (%)	131 (43.9)
M:F	148:150 (0.99:1)
BSA (Mean ± SD)	1.13 (±0.48)
Predominant defect n (%)	
ASD	124 (41.6)
VSD*	64 (21.5)
PDA	95 (31.9)
AVSD	12 (4)
Ap Window	1
Persistent truncus arteriosus	2
Mean Pa Pressure; Mean (±SD)	38.52 (±24.35) mmHg
PVRi, Median (IQR)	Wood Units m^2
Overall study group	3.3 (5.84)
Operable patients	2.73 (2.2)
Borderline patients	11.2 (4.1)
Inoperable patients	23.82 (13.19)
Operability n (%)	
Clearly inoperable	223 (74.83)
Borderline	35 (11.75)
Clearly operable	40 (13.43)
Indication for catheterisation n (%)	
ASD Device closure	82 (27.5)
VSD Device closure	1 (0.34)
PDA Device closure	86 (28.76)
Operability baseline data	128 (42.81)
Operability post-O_2	85
Qpi Median (IQR)	5.5 (4.18)
Qsi Median (IQR)	2.4 (1.2)
Qp:Qs Median (IQR)	2.1 (1.6)

*Including 05 patients with Down Syndrome.

0.52 ml/mmHg, IQR 0.34), when compared to both, borderline (mean PCa 0.87 ± 0.3 ml/mmHg, median 0.80 IQR 0.36) ($p < 0.001$) as well as clearly operable patients (mean pulmonary arterial compliance 3.61 ± 3.42 ml/mmHg, median 2.67 ml/mmHg IQR 2.2) ($p < 0.001$).

Relationship of pulmonary arterial compliance to pulmonary blood flow and pulmonary artery pressure

Pulmonary arterial compliance showed a linear relation with Qpi, with a loose fitting curve (R^2 0.37) (Fig 1). Pulmonary arterial compliance showed an inverse parabolic relationship with pulmonary artery systolic, diastolic, and mean pressures. The best correlation was seen between pulmonary arterial compliance and pulmonary artery systolic (power, r^2 0.44) (Fig 1). The correlation with mean and diastolic pulmonary artery pressures was a loose fit (r^2 0.29 and 0.15, respectively).

On multivariate analysis, nature of the defect and higher pulmonary artery mean significantly contributed to the lower pulmonary arterial compliance.

Relationship of pulmonary arterial compliance with other factors

Pulmonary arterial compliance did not correlate with the age, neither for the overall study group, nor for individual lesions atrial septal defect, ventricular septal defect, patent ductus arteriosus, or AVSD.

Pulmonary arterial compliance did not correlate with the transpulmonary pressure gradient (calculated as the difference between pulmonary artery diastolic and mean pulmonary artery wedge pressure) in the overall study group, or in any of the subgroups of operable, inoperable, and borderline patients.

Pulmonary arterial compliance - pulmonary vascular resistance index relationship

Pulmonary arterial compliance values in the entire study population showed inverse parabolic relation with the pulmonary vascular resistance index values (Power, $r^2 = 0.66$) (Fig 2). For the entire study group, the pulmonary arterial compliance and pulmonary vascular resistance index relationship was maintained across the operability spectrum (Fig 3) (power r^2 0.7, 0.8,0.9 for operable, borderline, and inoperable cases).

The relationship was also maintained across lesion subtypes, across the operability spectrum (Supplementary Figures 1–3). When plotted against age, the pulmonary arterial compliance pulmonary vascular resistance index relationship curve showed a shift to left at younger age (Supplementary Fig 4).

The pulmonary arterial compliance value started to show a decline in the overall group as well as in individual lesions, with minimal increase in pulmonary vascular resistance index (Fig 2).

Pulmonary arterial compliance and operability

As the median pulmonary arterial compliance value for inoperable patients was significantly lower than the operable patients, both in the overall study population as well as among individual lesions, an receiver operating characteristic analysis was done. A value of pulmonary arterial compliance less than 1.18 ml/mmHg correctly identified inoperable patients with a sensitivity and specificity of 95% (area under the receiver operating characteristic curve 0.99).

Review of borderline patients

Records from the patients classified as patients with borderline operability were reviewed by a team of experienced cardiologists, who classified these patients into “Likely operable” and “Likely inoperable” groups, based on the overall clinical picture and right heart catheterisation. These patients were then classified based on pulmonary arterial compliance alone as operable and inoperable. The calculated coefficient of agreement (kappa) was near zero, signifying no agreement.

RC time

The median RC time for the entire study population was 0.47 S (IQR 0.30). RC time in operable patients was significantly lower

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Table 2. Statistical analysis of PCa values.

Lesion	Category/PCa Median (IQR)	Category/PCa Median (IQR)	p
Overall Study group	Operable 2.67 (1.88,4.08)	Inoperable 0.52 (0.39,0.73)	<0.001
	Operable 2.67 (1.88,4.08)	Borderline 0.80 (0.66,1.02)	<0.001
	Borderline 0.80 (0.66,1.02)	Inoperable 0.52 (0.39,0.73)	0.13
Atrial septal defect			
	Operable 3.05 (2.07,4.76)	Inoperable 0.45 (0.28, 0.53)	<0.001
	Operable 3.05 (2.07,4.76)	Borderline 0.78 (0.55,0.94)	<0.001
	Borderline 0.78 (0.55,0.94)	Inoperable 0.45 (0.28, 0.53)	0.98
Ventricular septal defect			
	Operable 2.3 (1.8,2.64)	Inoperable 0.59 (0.43,0.75)	<0.001
	Operable 2.3 (1.8,2.64)	Borderline 0.85 (0.73,1.04)	<0.001
	Borderline 0.85 (0.73,1.04)	Inoperable 0.59 (0.43,0.75)	0.12
Patent ductus arteriosus			
	Operable 2.54 (1.8,3.36)	Inoperable 0.68 (0.42,0.76)	<0.001
	Operable 2.54 (1.8,3.36)	Borderline 1.1 (0.82,1.52)	0.03
	Borderline 1.1 (0.82,1.52)	Inoperable 0.68 (0.42,0.76)	0.89
AVSD			
	Operable 1.77 (1.21,3.57)	Inoperable 0.6 (0.56,0.66)	0.02
	Operable 1.77 (1.21,3.57)	Borderline 0.73 (0.70,0.76)	0.32
	Borderline 0.73 (0.70,0.76)	Inoperable 0.6 (0.56,0.66)	0.71

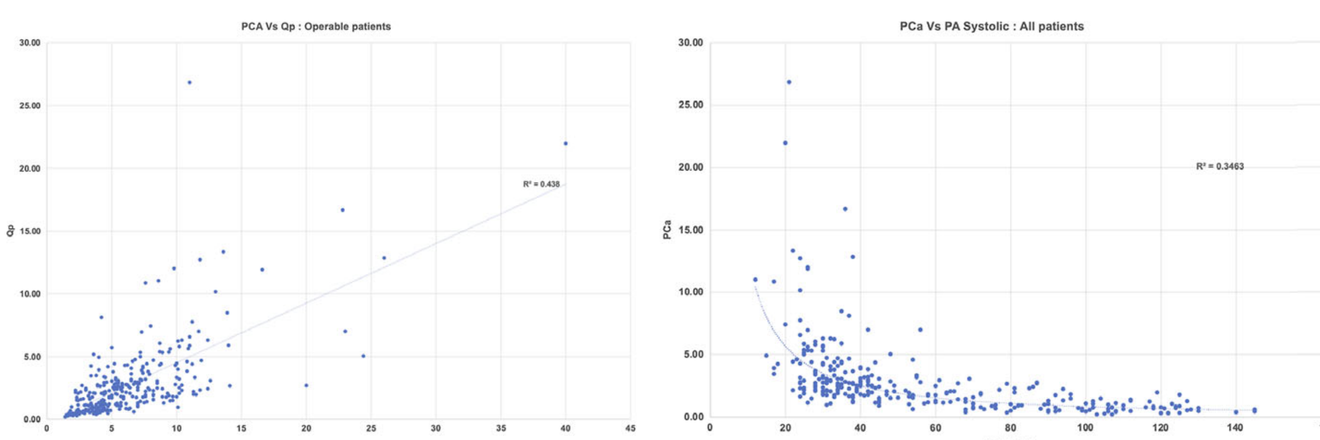

Figure 1. Relationship of PCa to Qpi (Operable patients) and PA systolic (all patients), all lesions.

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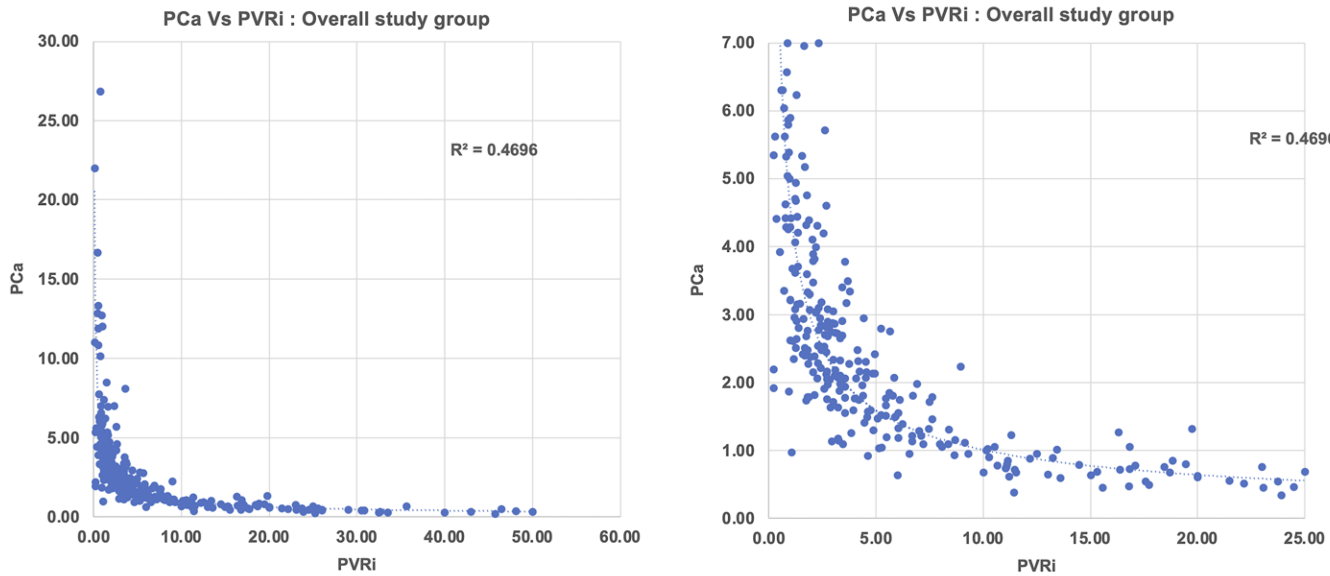

Figure 2. PCa versus PVRi correlation: all patients.

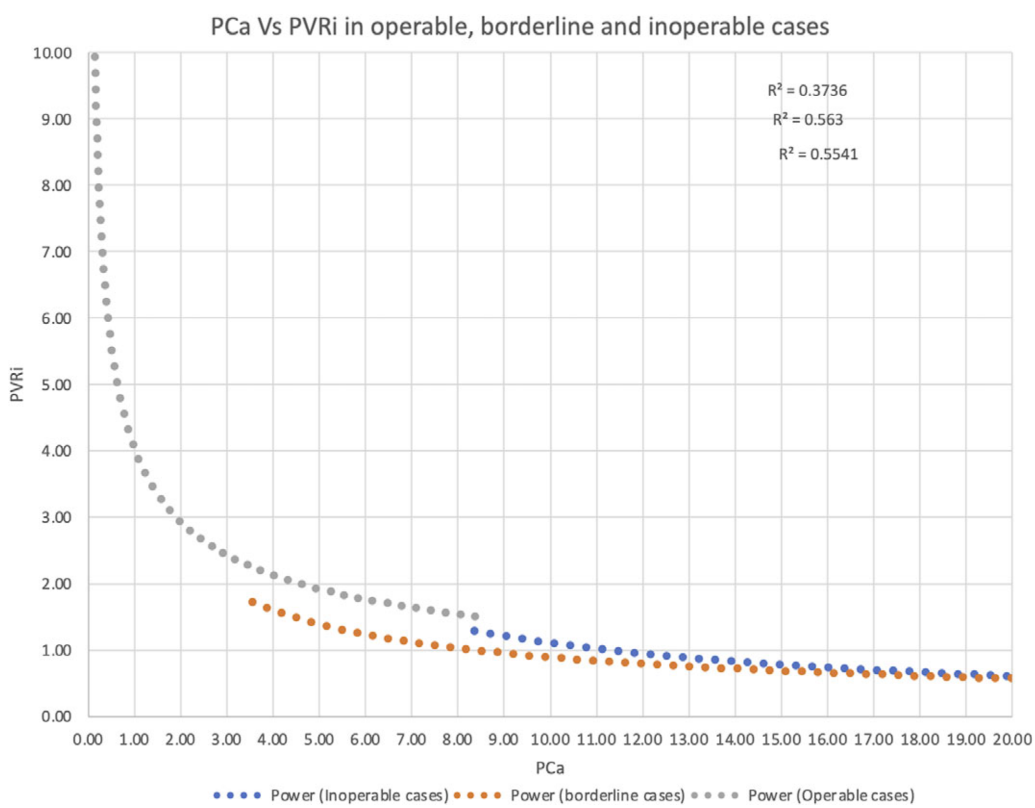

Figure 3. Relation between PCa and PVRi in operable, borderline, and inoperable cases.

than the inoperable patients (Median 0.40 IQR 0.23 in operable, 0.73 0.25 in inoperable patients, $p < 0.01$).

Discussion

The pulmonary circulation, despite being the focus of investigations for CHD-associated pulmonary hypertension for a long time, remains an enigma. Pulmonary arterial compliance, a marker of elastic properties of pulmonary circulation and an indicator of the pulsatile load of the pulmonary circulation,[17] represents an important part of the right ventricular afterload.[18] There is a surprising scarcity of data on pulmonary arterial compliance in patients with congenital left to right shunt lesions despite it is obvious physiological importance. While ideal measurement of pulmonary arterial compliance may be difficult,[19] the method used in this study is easily available to the clinicians. Earlier studies have documented pulmonary arterial compliance to be a function of pulmonary flow. The values of pulmonary arterial compliance for children with shunt lesions but no pulmonary hypertension, as obtained in our study are higher than that reported in normal

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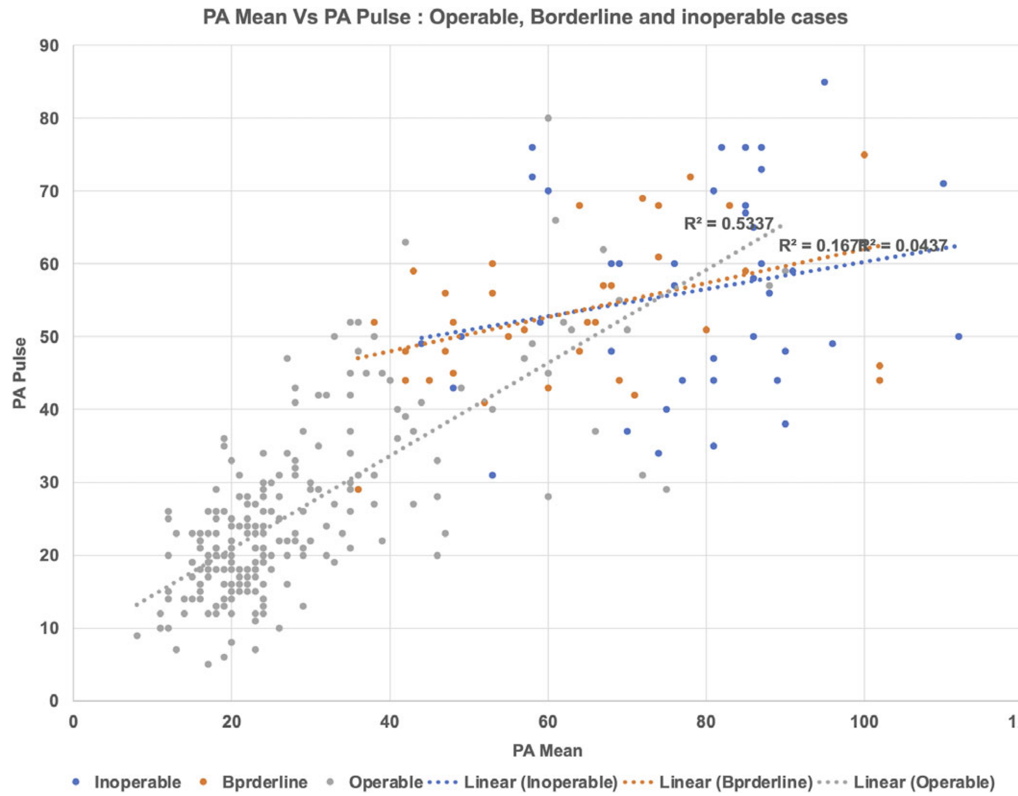

Figure 4. Relation between PA mean and PA pulse pressure in study patients.

children,[5] probably owing to the higher pulmonary to systemic blood flow ratio in our study patients. Very recently, a study of pulmonary artery compliance in children with shunt lesions and normal pulmonary vascular resistance index reported pulmonary arterial compliance values ranging from 2.44–3.88 mmHg/ml/m^2, similar to the values found in children with normal pulmonary artery pressures in our study.[20]

Our study found significantly lower pulmonary arterial compliance in inoperable patients across lesion subtypes. With a more advanced pulmonary vascular disease, right ventricular afterload increases. While a part of it is obviously due to an increase in pulmonary vascular resistance index, stiffer pulmonary arteries also increase the right ventricular afterload, just as stiffening of aorta with ageing increase the left ventricular afterload.[21] Assessment of pulmonary arterial compliance along with pulmonary vascular resistance index thus may yield a better overall picture of the right ventricular afterload.

Pulmonary arterial compliance has been linked to a more advanced disease state and prognosis[22] earlier, but the relation to operability has not been studied. Among haemodynamic parameters, operability assessment is generally based on the pulmonary vascular resistance index measurements only, and since the formulae used to calculate pulmonary arterial compliance and pulmonary vascular resistance index share many parameters, it might seem that the pulmonary arterial compliance and pulmonary vascular resistance index are mathematically measuring the same thing. However, two variables that are different between the formulae are the heart rate (instead of Qpi) and the pulse pressure instead of mean pulmonary artery pressure. As can be seen from Figure 4, the relationship between pulse pressure and mean pressure does not always remain constant, and therefore, pulmonary arterial compliance and pulmonary vascular resistance index might potentially yield different dimensions. That such is indeed the case has been documented in patients with thromboembolic pulmonary hypertension,[23] and also in patients with raised Left atrial pressures.[24,25] Further, heart rate variation may alter the relationship between pulmonary vascular resistance index and pulmonary arterial compliance. Heart rate has been shown to be of prognostic importance in patients with idiopathic pulmonary arterial hypertension.[26] Obviously, heart rate might vary due to multiple reasons, but the importance of heart rate measurement in clinical approach to pulmonary arterial hypertension in shunt lesions has not been considered. Thus, pulmonary arterial compliance and pulmonary vascular resistance index are complementary and not necessarily identical measures.

Whether the change in compliance can occur in the early stages of rising pulmonary vascular resistance index is not clear, but the idea is provocative. On the pulmonary arterial compliance-pulmonary vascular resistance index correlation curve (Fig 2), Pulmonary arterial compliance shows an earlier decline. Pulmonary arterial compliance values fall significantly when the pulmonary vascular resistance index is higher than normal, but still in "operable range." Though we did not study patients temporally, but if an initial rise of pulmonary vascular resistance index is taken as an early event, we can infer from the correlation curve that significant pulmonary arterial compliance fall in patients with a left to right shunt may allow early diagnosis of pulmonary vascular disease. Other studies on patients with pulmonary arterial hypertension have documented a similar inference.[17] Systematically conducted prospective studies may provide a better insight into this hypothesis.

The product of pulmonary vascular resistance index and pulmonary arterial compliance (RC time) has been argued both as constant,[6] and not constant,[18,27] in the literature based on theoretic assumptions. We found varying RC time in operable and inoperable patients. This also supports our contention that the study of pulmonary arterial compliance in patients with shunt lesion may

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provide additional information, in addition to the conventional measures of pulmonary vascular resistance index.

Unfortunately, the study of pulmonary arterial compliance did not improve our decision-making in borderline cases. There is too much empiricism involved in decision-making in shunt lesions in borderline cases at older ages: and in the absence of long-term data, we can only draw limited inference from this. However, it seems that study of pulmonary arterial compliance in shunt lesions may add to our understanding of pulmonary circulation. Further prospective studies seem warranted.

Limitations

The major limitations of our study are that it is a retrospective analysis of records of a number of patients who have undergone catheterisation with a shunt lesion for diagnostic and/or therapeutic purpose. The quality of data may not be as good as that of a prospective study. Oxygen consumption was assumed in all these patients, and in patients where pulmonary artery wedge pressure was not measured, left ventricular end-diastolic pressure was presumed to be equal to mean pulmonary artery wedge pressure. However, the significant number of patients of varying ages, and different levels of pulmonary vascular resistance index make the analysis relevant to patients with shunt lesions. Our analysis of operability in borderline cases and its relation to pulmonary arterial compliance is empirical. To the best of our knowledge, our study is the first one addressing the role of pulmonary arterial compliance in operability assessment of patients with shunt lesions and increased pulmonary blood flow.

Conclusions

This study shows that median pulmonary arterial compliance in patients with shunt lesions and normal pulmonary artery pressures is 4.1 ml/mmHg (IQR 3.2). Pulmonary arterial compliance of 1.18 mmHg or less, etc., indicated inoperability. In this patient population, the RC time was not constant. This study has raised the possibility that an analysis of pulmonary arterial compliance in patients with shunt lesions might provide additional clinically meaningful data. Further studies seem warranted.

Supplementary material. To view supplementary material for this article, please visit https://doi.org/10.1017/S1047951122003341

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Conflicts of Interest. None.

References

1. Milnor WR, Jose AD, McGaff CJ. Pulmonary vascular volume, resistance, and compliance in man. Circulation [Internet] 1960; 22: 130–137.
2. van der Feen DE, Bartelds B, de Boer RA, Berger RMF. Assessment of reversibility in pulmonary arterial hypertension and congenital heart disease. Heart 2019; 105: 276–282.
3. Lopes AA, Oleary PW. Measurement, interpretation and use of haemodynamic parameters in pulmonary hypertension associated with congenital cardiac disease. Cardiol Young. 2009; 19: 431–435.
4. Slife DM, Latham RD, Sipkema P, Westerhof N. Pulmonary arterial compliance at rest and exercise in normal humans. Am J Physiol - Hear Circ Physiol 1990; 258: 27–26.
5. Basnet NB, Awa S, Hishi T, Yanagisawa M. Pulmonary arterial compliance in children with atrial and ventricular septal defect. Heart Vessels. 2000; 15: 61–69.
6. Reuben SR. Compliance of the human pulmonary arterial system in disease. Circ Res. 1971; 29: 40–50.
7. Kirby BJ. Pulmonary artery compliance in pulmonary heart disease. Progr Resp Res 1975; 9: 254–260.
8. LaFarge CG, Miettinen OS. The estimation of oxygen consumption. Cardiovasc Res [Internet] 1970; 4: 23–30.
9. Lopes AA, Barst RJ, Haworth SG, et al. Repair of congenital heart disease with associated pulmonary hypertension in children: what are the minimal investigative procedures? Consensus statement from the Congenital Heart Disease and Pediatric Task Forces, Pulmonary Vascular Research Institute (PVRI). Pulm Circ [Internet] 2014; 4: 330–341.
10. Yin FCP, Liu Z, Brin KP. Estimation of Arterial Compliance. In Ventricular/Vascular Coupling. Springer New York, New York, NY, 1987: 384–398.
11. Liu Z, Brin KP, Yin FC. Estimation of total arterial compliance: an improved method and evaluation of current methods. Am J Physiol Circ Physiol 1986; 251: H588–600.
12. Lankhaar JW, Westerhof N, Faes TJC, et al. Pulmonary vascular resistance and compliance stay inversely related during treatment of pulmonary hypertension. Eur Heart J. 2008; 29: 1688–1695.
13. Stergiopulos N, Meister JJ, Westerhof N. Simple and accurate way for estimating total and segmental arterial compliance: the pulse pressure method. Ann Biomed Eng 1994; 22: 392–397.
14. Saouti N, Westerhof N, Postmus PE, Vonk-Noordegraaf A. The arterial load in pulmonary hypertension. Eur Respir Rev. 2010; 19: 197–203.
15. Ghio S, Schirinzi S, Pica S. Pulmonary arterial compliance: how and why should we measure it? Glob Cardiol Sci Pract 2015; 2015: 58.
16. Stergiopulos N, Meister JJ, Westerhof N. Evaluation of methods for estimation of total arterial compliance. Am J Physiol - Hear Circ Physiol [Internet] 1995; 268: H1540–8. DOI 10.1152/ajpheart.1995.268.4.H1540
17. Saouti N, Westerhof N, Postmus PE, Vonk-Noordegraaf A. The arterial load in pulmonary hypertension. Eur Respir Rev 2010; 19: 197–203.
18. Tedford RJ. Determinants of right ventricular afterload (2013 Grover Conference series). Pulm Circ. 2014; 4: 211–219.
19. Vanden Eynden F, Bové T, Chirade ML, Van Nooten G, Segers P. Measuring pulmonary arterial compliance: mission impossible? Insights from a novel in vivo continuous-flow based experimental model. Pulm Circ 2018; 8: 1–12.
20. Iwaya Y, Muneuchi J, Sugitani Y, Watanabe M. Pulmonary vascular resistance and compliance in pulmonary blood flow alterations in children with congenital heart disease. Heart Vessels 2022; 37: 1283–1289.
21. Randall OS, van den Bos GC, Westerhof N. Systemic compliance: does it play a role in the genesis of essential hypertension? Cardiovasc Res 1984; 18: 455–462.
22. Galiè N, Channick RN, Frantz RP, et al. Risk stratification and medical therapy of pulmonary arterial hypertension. Eur Respir J 2019; 53: 1801889. DOI 10.1183/13993003.01889-2018.
23. Palecek T, Jansa P, Ambroz D, et al. Are pulmonary artery pulsatility indexes able to differentiate chronic pulmonary thromboembolism from pulmonary arterial hypertension? An echocardiographic and catheterization study. Heart Vessels 2011; 26: 176–182.
24. Kussmaul WG, Altschuler JA, Matthai WH, Laskey WK. Right ventricular-vascular interaction in congestive heart failure. Importance of low-frequency impedance. Circulation 1993; 88: 1010–1015.
25. Dragu R, Rispler S, Habib M, et al. Pulmonary arterial capacitance in patients with heart failure and reactive pulmonary hypertension. Eur J Heart Fail 2015; 17: 74–80.
26. Mahapatra S, Nishimura RA, Sorajja P, Cha S, McGoon MD. Relationship of pulmonary arterial capacitance and mortality in idiopathic pulmonary arterial hypertension. J Am Coll Cardiol. 2006; 47: 799–803.
27. Naeije R, Delcroix M. Is the time constant of the pulmonary circulation truly constant? Eur Respir J 2014; 43: 1541–1542.