Right atrial mechanics provide useful insight in pediatric pulmonary hypertension

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Abstract
Right atrial (RA) mechanics have been studied infrequently in children in the past due to technical constraints. With the advent of strain imaging, RA physiology can now be studied in greater detail. The principal aim of this study was to describe functional changes in right heart mechanics of children with idiopathic pulmonary arterial hypertension (PAH), by using new applications of RA strain. In this retrospective study, we evaluated RA mechanics of 20 patients (age range = 3–23 years) with PAH and 18 control patients. RA longitudinal strain (RALS) and longitudinal displacement (LD) were calculated by speckle-tracking echocardiography. RALS was plotted against LD, producing a characteristic strain-displacement (S-D) loop. Standard indices of right heart function and right heart catheterization data were obtained. Patients were clinically subdivided into “compensated” and “decompensated” PAH. A chart review was performed to identify patients who subsequently developed adverse outcomes, including death, awaiting or received lung and/or heart transplantation. RALS was significantly lower in decompensated PAH compared with both controls and compensated PAH. Area enclosed by S-D loops differed significantly between the compensated and decompensated PAH subgroups (5.33 [3.90–9.44] versus 1.83 [1.17–2.36], \( P < 0.05 \)). S-D loop area and RALS possessed high sensitivity and specificity compared to other parameters for identifying children with PAH who subsequently developed adverse outcomes. In particular, their sensitivities and specificities were greatly superior compared to those of tricuspid annular plane systolic excursion (TAPSE). RALS may represent a useful metric for assessing right ventricular (RV) dysfunction. S-D loops, composed over an entire cardiac cycle, may present useful, composite information regarding both systolic and diastolic right heart function. RA mechanics may serve as useful tools for identifying patients with more severe PAH, who are at risk for future adverse outcomes associated with RV failure.

Keywords
pediatrics, pulmonary arterial hypertension, right atrial longitudinal strain, speckle tracking echocardiography, brain natriuretic peptide, six-minute walk, tricuspid annular systolic plane excursion, strain-displacement loop

Introduction
Standard and advanced echocardiographic techniques continue to represent essential elements in the evaluation of patients with pulmonary hypertension (PAH). This disease process is characterized by increased pulmonary vascular resistance resulting in chronic right ventricular (RV) pressure overload, myocardial remodeling, leading to RV hypertrophy, dilation, and eventual RV failure.1,2 The echocardiographic evaluation of a patient with suspected or established PAH may reveal characteristic findings such as tricuspid regurgitation (TR), right atrial (RA) and RV dilation, RV hypertrophy, abnormal shape and function of the interventricular septum, and dilation of the main pulmonary artery.3 However, many of these findings may not be present...
until quite late in the course of the disease, leading to the search for indices that may identify decompensation at earlier stages.

With the advent of speckle-tracking echocardiography (STE), it is now possible to quantify RA deformation over the course of the entire cardiac cycle from a single beat. Strain imaging has been applied in patients with PAH and right heart dysfunction. In adults with chronic systolic heart failure, RA peak longitudinal strain (RALS) showed a strong inverse correlation with invasively measured systolic pulmonary artery pressure (PAP) and may represent an adjunct to invasive means of hemodynamic assessment. In adults with PAH, RALS was noted to be low and it negatively correlated with RA pressure. However, in the pediatric population the RA represents the “forgotten” chamber of the heart and studies on RA physiology and function in children are quite limited. This is especially true in pediatric PAH. STE can generate both strain and displacement data from the same heartbeat, which can simultaneously assess regional and global myocardial deformation of a cardiac chamber, including mechanical dyssynchrony. Simultaneous measurement of strain and displacement have been studied in other chambers of the heart, including the pressure-overloaded RV in adults. Notably, these studies in adults have shown that measurement of RV displacement and strain may facilitate reliable and quantitative estimation of RV function. However, when the RV is significantly dilated, it may not be possible to include all the walls of the RV in the same echocardiographic acquisition window. The apex of the RV may often be truncated in acquisition windows, particularly in retrospective studies. In contrast, the RA is a simpler geometric structure and invariably well-visualized from an apical four-chamber view. Therefore, RA mechanics may provide a simpler and more reliable measure of RV decompensation. Based on these findings, we were enticed by the feasibility of describing a strain-displacement (S-D) relationship in the RA of children with PAH.

The objective of our study is to characterize RA function throughout a single cardiac cycle in healthy children and those with PAH, specifically looking at differences in RA strain and the strain-displacement relationship. We hypothesize that evaluation of RA strain in combination with RA displacement may provide useful insight into the function of both the normal and failing RA in PAH. These indices may identify patients at risk for future adverse outcomes.

**Methods**

**Patient population**

Patients followed at The Children’s Hospital of Philadelphia with a diagnosis of PAH without post-tricuspid valve shunt were recruited for this study. Twenty children classified as having PAH due to idiopathic etiology (Nice Classification type 1.1), drug or toxin (Nice Classification type 1.3), congenital heart disease but without current shunt (Nice Classification type 1.4.4), or hematologic diagnosis (Nice Classification type 5.1) were included in this study. We excluded PAH patients with significant defects in the atrial septum and patients with left ventricular (LV) dysfunction or pulmonary vein obstruction. Control individuals in this study were otherwise healthy pediatric patients referred for echocardiography for indications such as noncardiac chest pain, vasodepressor syncope, and functional heart murmurs. All control participants were found to have structurally normal hearts without evidence of PAH by echocardiogram. This retrospective, descriptive study was approved by the Institutional Review Board of the Children’s Hospital of Philadelphia.

**Strain imaging**

Echocardiograms were obtained using Philips iE33 ultrasound machines (Philips Medical Systems, Andover, MA, USA). Two-dimensional apical four-chamber views were obtained and uploaded for analysis by a vendor-independent STE software (TomTec Cardiac Performance version 1.2.4.2, TomTec Imaging Systems, Munich, Germany). The frame rate of the acquired images exceeded 60 frames/s. Manual tracing along the RA endocardial border was performed with termination of the tracing 0.5 cm above the atrioventricular junction, to limit influence of tricuspid annular motion. The software divided the right atrium into six segments, with longitudinal strain and displacement obtained for each segment over a complete cardiac cycle. The global RA strain and longitudinal displacement (LD) for each patient were averages of the values obtained from these six segments. By convention, displacement away from the centroid of the cardiac chamber is indicated by negative values and may be considered as a surrogate for increase in RA volume.

**Right-heart function**

To better assess the utility of strain imaging in evaluation of right-sided function in individuals with PAH, the patients were subdivided into two groups: decompensated and compensated. “ Decompensated PAH” was defined as the subgroup of patients with functional and laboratory evidence of significant right heart dysfunction at time of the echocardiogram. Specifically, the participants in this subgroup were in World Health Organization (WHO) functional class III or IV. Based on adult and pediatric studies on brain natriuretic peptide (BNP), which showed strikingly decreased survival in idiopathic PAH, we chose a cut-off value for BNP of 180 pg/mL. In our study, all decompensated PAH patients exhibited a BNP value of > 180 pg/mL. “Compensated PAH” was defined as individuals in WHO functional class I or II or a BNP < 180 pg/mL, or both.
In addition to RA mechanics and strain values, more conventional indices such as RV peak strain, RV area change (RVAC), and tricuspid annular plane systolic excursion (TAPSE) were recorded and used as indices of RV function. To compare TAPSE in participants of different ages and sizes, the values were indexed to the body surface area (BSA). We also compared percent-predicted TAPSE values using established reference ranges. In individuals with PAH, BNP values obtained on or as close as possible to the date of the echocardiogram were evaluated for this study: BNP levels were obtained on 16 patients on the same day as the echocardiogram, the remaining four were obtained within three months.

Six-minute walk distances (6MWD) were obtained on or as close as possible to the date of the echocardiogram. Of the 20 PAH participants, 19 had 6MWD measurements recorded. Twelve measurements took place on the same day as the echocardiogram and four underwent the 6-min walking test (6MWT) within three months of their echocardiograms. For the remaining three individuals, the 6MWT was performed approximately six months after their echocardiograms. To determine a percent-predicted 6MWT value for each participant, the observed 6MWD was divided by the distance expected for their sex and age. Several reference ranges are available for pediatric 6MWT times based on their ages. Using the appropriate reference ranges, we calculated predicted values for 6MWTs in our cohort.

**Strain-displacement loops**

Construction of S-D loops was undertaken by plotting RALS along the y-axis and LD along the x-axis. A typical S-D loop is illustrated in Fig. 2. The area enclosed by the S-D loop was calculated using the trapezoidal area of polygon formula, where the loop was broken into mini-trapezoids and the area of the loop was equal to the sum of all the trapezoids. The total area of the loop was the sum of \[x(i+1)-x(i)][y(i)+y(i+1)]/2\] for \(i\) points of a polygon, where \(x(1), y(1)=x(i+1), y(i+1)\). The unit for this measurement is % mm/cm and is simply referred to as unit (U) in this paper. In order to compare the right atria of children of different ages, the LD was indexed to the BSA. We used an additional method of indexing LD by dividing it with the largest RA dimension with the tricuspid valve closed.

**Cardiac catheterization data**

We evaluated right heart catheterization data that were obtained within six months of the echocardiogram. Mean pulmonary artery pressure (mPAP) and indexed pulmonary vascular resistance (PVRi) were recorded for each individual.

**Adverse outcomes**

Based on > 2-year follow-up data, we identified participants who later developed adverse outcomes associated with their PAH. Adverse outcomes included death, awaiting or receiving lung and/or heart transplantation.

**Statistical analysis**

Continuous variables were non-normally distributed and are reported as median with interquartile ranges (IQR) noted parenthetically. Comparisons between the variables were performed using the Mann–Whitney U test. Non-parametric receiver operating characteristic (ROC) analysis was performed to test the sensitivity and specificity of the different echocardiographic indices, for the outcome described above. ROC curves were plotted and the areas under the curves were measured. For all significance testing, a difference was considered significant if \(P\) value < 0.05. The statistical analysis was performed using Stata software (StataCorp, version 13, College Station, TX, USA). Intra-class correlation (ICC) was used to assess intra- and inter-observer variability. For this purpose, eight patients were selected randomly and the analysis was repeated de novo by two investigators. For intra-observer variability, one observer repeated the measurements after two weeks.

**Results**

**Demographics**

A total of 38 children were included in this retrospective study, with 18 control individuals and 20 patients with PAH. Demographic data are summarized in Table 1. Control participants did not differ significantly from PAH participants with regards to age or BSA. Of the 20 PAH individuals, 15 were classified as idiopathic primary PAH (Nice Classification type 1.1), two were classified as having PAH arising due to connective tissue disease (Nice Classification type 1.4.1), one was classified as having PAH arising due to congenital heart disease (closed VSD) (Nice Classification type 1.4.4), one was classified as...
having PAH arising from drug or toxin (Nice Classification type 1.3), and one was classified as having PAH arising from hematologic etiology (Nice Classification type 5.1). None of the 20 individuals with PAH included in this study were found to have a post-tricuspid valve shunt. A small patent foramen ovale with bidirectional shunting was noted in two patients.

Comparison of control and PAH population

The control group differed significantly from the PAH group in several RV parameters, including percent-predicted TAPSE, and RV to LV ratio in diastole. RV longitudinal strain was lower in the PAH group versus controls (–15.7 ± 3.6% vs –23.3 ± 3.8%, \( P < 0.05 \); data expressed as mean ± standard deviation). Several indices of RA function were abnormal in the PAH group. Notably, RALS was significantly decreased in the PAH group compared to controls (31.9 [21.6–48.1] versus 57.8 [52.9–65.9], \( P < 0.05 \)). RALS also showed a moderately significant inverse correlation with mPAP in patients undergoing catheterization within six months (\( r = 0.51, P < 0.05 \)) (Fig. 1). The above findings are summarized in Table 2.

Comparison of compensated and decompensated PAH groups

The entire PAH population was divided into compensated and decompensated groups to determine if RA indices such as RALS and S-D loops varied between these subgroups and whether a point of decline in RA function could be detected in these individuals. The data are summarized in

### Table 2. Right heart data.

|                | Control | All PAH | Compensated PAH | Decompensated PAH |
|----------------|---------|---------|-----------------|-------------------|
| N              | 18      | 20      | 13              | 7                 |
| RALS (%)       | 57.84 (52.94–65.89) | 31.93 (21.61–48.19)* | 46.38 (34.71–50.94)* | 21.04 (17.17–22.53)*† |
| Loop area\( \text{BSA} \) (U) | 25.31 (15.07–29.53) | 12.95 (10.44–32.19) | 19.30 (13.68–38.21) | 10.22 (5.41–11.60)*† |
| Loop area\( \text{length} \) (U) | 7.88 (5.94–9.88) | 3.85 (1.92–8.51)* | 5.33 (3.90–9.44) | 1.83 (1.17–2.36)*† |
| TAPSE\( \text{BSA} \) (cm) | 1.45 (1.30–1.83) | 1.26 (1.16–1.69) | 1.23 (1.17–1.56) | 1.49 (0.75–1.69) |
| \( \% \text{p TAPSE} \) (%) | 91.24 (84.97–102.69) | 81.71 (68.59–85.76)* | 83.79 (76.97–85.88)* | 73.02 (61.85–83.67)*† |
| RV:LV\( \text{diastole} \) | 1.88 (1.66–2.05) | 1.52 (0.98–1.94)* | 1.73 (1.1–2.19) | 0.84 (0.61–1.52)*† |
| BNP (pg/mL)    | 60.5 (19.95–300.85) | 29.7 (15.8–52.7) | 434.5 (190.5–509.5) | 434.5 (190.5–509.5)† |
| 6MWD (m)       | 548 (412–595) | 574.5 (486–617.5) | 440 (336–560) | 440 (336–560) |
| \( \% \text{p 6MWD} \) (%) | 89.2 (75.09–96.59) | 94.17 (90.69–101.71) | 75.09 (66.89–81.81) | 75.09 (66.89–81.81)† |
| mPAP (mmHg)    | 56 (42–68) | 54.5 (40–58.5) | 68 (53–75) | 68 (53–75) |
| PVRI (Wood Units. m\(^2\)) | 9.2 (9.1–21.6) | 13.9 (8.4–18.5) | 18.1 (14.1–25.5) |

Data are expressed as median with first and third interquartile range in parenthesis.

* \( P < 0.05 \) when compared with control.
† \( P < 0.05 \) when compared with compensated PAH.

RALS, right atrial longitudinal strain; Loop area\( \text{BSA} \), S-D loop area indexed for body surface area; Loop area\( \text{length} \), S-D loop area indexed for right atrial length; TAPSE\( \text{BSA} \), tricuspid annular plane systolic excursion, indexed for body surface area; \( \% \text{p TAPSE} \), percent-predicted TAPSE; RV:LV\( \text{diastole} \), ratio of right ventricular to left ventricular diameter at end-diastole; BNP, brain natriuretic peptide; 6MWD, 6-min walk distance; \( \% \text{p 6MWD} \), percent-predicted 6-min walk distance; mPAP, mean pulmonary artery pressure.
Table 2. Regarding the more conventional data, it is noteworthy that the BNP values exceeded 190 pg/mL in all patients in the decompensated group (median = 434.5 pg/mL). The decompensated PAH participants differed significantly from the compensated PAH group with regards to several measures of RA mechanics (RALS, S-D loop area indexed for either BSA or RA length), RV mechanics (RV to LV ratio in diastole), laboratory markers, and clinical function (percent-predicted 6MWT) as outlined in Table 2. Both the compensated and decompensated PAH populations exhibited elevated mPAPs. Despite a trend of observing higher mPAP and PVRi in the decompensated group, there was no statistical difference in mPAP or PVRi between the two subgroups.

We wish to point out that the area enclosed by the S-D loop was similar between the control and compensated

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**Fig. 2.** Typical RA S-D loop. The S-D loop is generated by plotting the RA LD along the x-axis and the longitudinal strain along the y-axis. This results in a characteristic elliptical loop in the counter-clockwise direction, starting with tricuspid valve closure (TVC). The area of the S-D loop was constituted by reservoir, conduit, and contractile phases of RA function. The reservoir phase continues from TVC to tricuspid valve opening (TVO), after which starts the conduit phase, followed by a brief contractile phase. The contractile phase in children is not as pronounced as in adults and therefore a distinct incisura is not well-detected in the waveforms of pediatric patients.
groups, but significantly smaller in the decompensated PAH patients compared with both control and compensated PAH individuals (Fig. 3, Table 2).

**Outcomes in PAH populations**

Six out of seven decompensated and one out of 13 compensated participants experienced an adverse outcome. To determine the sensitivity and specificity in identifying the individuals who later went on to develop these outcomes, ROC curves were constructed for RALS, S-D loop area indexed for BSA, S-D loop area indexed for RA length, BNP, percent-predicted TAPSE, and TAPSE indexed for BSA (Table 3). ROC curves for S-D loop indexed for RA length and BSA, and RALS all demonstrated excellent AUC (0.92, 0.90, 0.92, respectively), whereas the AUC for TAPSE indexed for BSA was quite poor (0.54) (Fig. 4).

**Intra-observer and inter-observer variability**

We found excellent reproducibility for RALS and LD measurements. ICC values for inter-observer variability were 0.88
Table 3. ROC curve data for measures of right heart function and adverse outcomes.

| ROC analysis                  | Cutpoint | Sensitivity (%) | Specificity (%) | AUC     | 95% CI       |
|------------------------------|----------|-----------------|-----------------|---------|--------------|
| Loop area<sup>length</sup> vs. Outcome | 2.36 U   | 100             | 92.3            | 0.92    | 0.77–1.0     |
| Loop area<sup>BSA</sup> vs. Outcome | 11.6 U   | 100             | 84.6            | 0.90    | 0.75–1.0     |
| RALS vs. Outcome           | 28.74%   | 100             | 84.6            | 0.92    | 0.79–1.0     |
| BNP vs. Outcome            | 190.4 pg/mL | 85.7           | 92.3            | 0.84    | 0.58–1.0     |
| Percent-predicted TAPSE vs. outcome | 83.7%   | 85.71           | 53.85           | 0.64    | 0.35–0.92    |
| TAPSE indexed for BSA vs. outcome | 1.27 cm | 71.4            | 61.5            | 0.54    | 0.23–0.84    |

AUC, area under ROC curve; Loop area<sup>length</sup>, S-D loop area indexed for right atrial length; Loop area<sup>BSA</sup>, S-D loop area indexed for body surface area; RALS, right atrial longitudinal strain; BNP, brain natriuretic peptide; CI, confidence interval.

Fig. 4. ROC curves comparing (a) S-D loop area indexed for RA length vs outcome, (b) S-D loop area indexed for BSA vs outcome, (c) RALS vs outcome, (d) BNP vs outcome, (e) percent-predicted TAPSE vs outcome, and (f) TAPSE indexed for BSA vs outcome. Additional data for each ROC curve (cut-point, sensitivity, specificity, area under curve [AUC]) can be found in Table 3. In all ROC curves, cut-points are delineated by triangles.
for RALS and 0.94 for LD, whereas ICC values for intra-
observer variability were 0.9 for RALS and 0.92 for LD, respectively.

Discussion

Although the RA is often regarded as the “forgotten cham-
ber” of the heart, RA mechanics may serve as an important
bometer of right heart dysfunction. Strain imaging repre-
sents an exciting new tool for evaluating RA mechanics.
This single-center, pilot study represents one of the first
efforts to use this tool in the evaluation of pediatric PAH.
Additionally, in this study we have proposed a new concept,
the S-D loop, that may provide useful discriminatory in-
formation regarding compensated and decompensated states
of PAH in children.

Right atrial longitudinal strain

Measurement of RA strain may represent an exciting
new tool in the assessment of RA physiology that was not
available in the past. Previous research into pediatric RA
strain have demonstrated abnormal indices in single ven-
tricle patients22 and in patients with repaired tetralogy of
Fallot.23 Adult studies have demonstrated a decreased
RALS in participants with PAH compared with controls6,7
and this finding is reflected in our pediatric data. Of interest,
RALS was also significantly different between compensated
and decompensated subgroups of PAH.

Patients with chronic right heart pressure overload dem-
onstrate diastolic dysfunction before systolic dysfunction.24
The gradual increase in RA strain is reflected by the ascend-
ing limb and peak of the RA strain curve (Fig. 2). This phase
occurs during atrial filling, referred to as reservoir function
of the RA. Atrial filling, in turn, occurs during ventricular
diastole. Therefore, the lower RALS in patients with PAH
may reflect the abnormal myocardial relaxation and dia-
stolic dysfunction of the RV.

Atrial strain-displacement loops

To gain insights into RA function and mechanics over the
entire cardiac cycle, we propose a novel approach, the S-D
loop. Plotting atrial longitudinal strain against LD pro-
duced a characteristic elliptical loop, with turning points
corresponding to tricuspid valve closing and opening
(Fig. 2). Strain represents local deformation of a small myo-
cardial segment, whereas displacement evaluates the move-
ment of the whole chamber. Moreover, displacement may
serve as a surrogate for change in RA volume. The S-D
loops may serve as a helpful index, comparing atrial segmen-
tal deformation against the overall movement of the atrial
wall and may provide a useful “composite view” of atrial
filling (reservoir phase) and emptying (conduit + contractile
phases). The concept of simultaneous measurement of lon-
gitudinal strain and LD has been applied in adults with
constrictive pericarditis,25 RV pressure overload,9,10 LV
pressure overload in aortic stenosis;26 and in the evaluation
of carotid artery stiffness.27 Longitudinal strain and LD
have been used to describe the elasticity of the dilated
aorta. Elasticity of the aorta incorporates both the property
of dilating and the property of recoiling to its initial shape.28
Similarly, the atrial S-D relationship may provide indirect
evidence of the elasticity of the RA tissue. We also speculate
that during the conduit phase (when tricuspid valve remains
open), the RA S-D loop may also reflect RV diastolic stiff-
ness. The concept of the S-D loop is quite new. It provides
simultaneous information regarding the regional shortening
and global movement of the atrial wall during the entire
cardiac cycle. The concept was derived from torsion-
displacement loop described by us and others.28,29 To
describe the area enclosed by torsion-volume loops,
Notomi et al. coined the term “Filling Work.” They pro-
posed that the area enclosed by the loops gives a measure of
“work” being performed by the chamber and may provide a
measure of the energy stored in the cardiac chamber.30
Although a theoretical analogy is being drawn, we cannot
extrapolate these theoretical concepts to our study, as ours
is a pilot study that aimed to show the feasibility of con-
structing strain versus displacement loops in normal and
diseased RA. Clearly, further work is needed to determine
the physiologic significance of the area enclosed by the S-D
loop. Based on other studies that have used strain and dis-
placement simultaneously, we can merely speculate that the
reduced area of the S-D loop may provide a visual display of
the decreased elasticity of the atrial tissue. It is a first step in
visually describing the RA mechanics during an entire car-
diac cycle. This was nicely demonstrated in patients with
decompensated PAH, where the S-D loops were notably
“skinnier” (Fig. 3, lower panel).

Comparison with current means of assessing right heart
mechanics in PAH

Non-invasive identification of impending RV decompen-
sation is challenging in children with PAH, since the 6MWD
and the degree of TR do not deteriorate with increase in
disease severity at the same rate as in adult PAH.
Moreover, in the absence of sufficient TR, current echocar-
diographic indices are poor predictors of disease severity.
In this scenario, RA mechanics may provide a better insight
into the state of RV decompensation. Both RALS and S-D
loops may serve as upgrades to the clinician’s toolkit. It is
important to recognize the best use of each tool available for
the evaluation of PAH in children. It may be that a com-
bination of tools will yield the most complete picture of right
heart mechanics at a given point in time. This study and
others in adults6,7 have demonstrated that the decrease in
RALS with increasing disease severity makes RALS a useful
measure of RV diastolic dysfunction. Since diastolic dys-
function of the RV precedes systolic dysfunction in chronic
right heart pressure overload,22 RALS may be a useful index
to monitor serial diastolic changes in patients with PAH, similar to findings in serum BNP. In contrast, TAPSE and RV strain provide serial evaluation of RV during systole, while the RA S-D loop may provide a composite insight into both systolic and diastolic function of RV. Our study showed that the RA indices had greater sensitivity and specificity (and larger AUCs) compared to the more conventional indices like BNP and TAPSE. In our study, out of all the indices evaluated, the S-D loop indexed for RA length showed the best sensitivity (100%) and specificity (92.3%). This points to great potential value of this index but also warrants further validation. In particular, serial monitoring of these indices after initiating therapy for PAH would be of utmost interest to researchers and clinicians alike.

**Clinical implications**

Established clinical criteria used in the evaluation of adult PAH may be less useful in the pediatric population. For example, the 6MWT can be difficult to carry out according to established protocols in younger children and does not always reflect the degree of right heart dysfunction. Children often present with lower RA pressures, better compensation of the RV to the increased afterload, and higher mixed venous oxygen saturations.2,13 The study of RA mechanics may therefore be of value in this population. The S-D loop provides us with a composite view of the reservoir, conduit, and contractile phases of RA function. The mathematical calculations for measuring these new indices can be incorporated into calculation packages of commercial echocardiography machines. Automation of RALS and S-D loops into commercial ultrasound machines may encourage more use of these indices in busy clinical settings and not limit them to the research arena only.

**Study limitations**

Due to small sample size, further subgroup analyses were not feasible. Furthermore, statistical evaluations determining degree of association between the laboratory variables and adverse outcomes were not performed because of small sample size. We excluded individuals with ongoing lung parenchymal lesions, significant atrial level shunts, pulmonary vein stenoses, and LV dysfunction. In this fashion, we present a relatively homogeneous population which should have narrow limits for test results and clinical outcomes. Moreover, even in a tertiary referral center, idiopathic PAH remains an uncommon disease. As our primary aim in this study was to perform research on a new echocardiographic index, we pursued a statistically sufficient number of patients needed to provide meaningful results, at the same time limiting heterogeneity in the sample. This study is a single-center, pilot study that may pave the way for larger, multicenter studies. The primary focus of this study was on the RA physiology. Therefore, we did not focus significantly on the RV physiology.

**Conclusions**

RA mechanics may serve as novel metrics in the assessment of right heart dysfunction in children with PAH. Identification of impending RV decompensation is difficult in pediatrics, where sub-maximal exercise testing and the degree of TR are often better than expected for the degree of elevated mPAP. RA strain and S-D loops may serve as useful tools for identifying patients at risk for future adverse outcomes from progressive RV dysfunction.

**Conflict of interest**

The author(s) declare that there is no conflict of interest.

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