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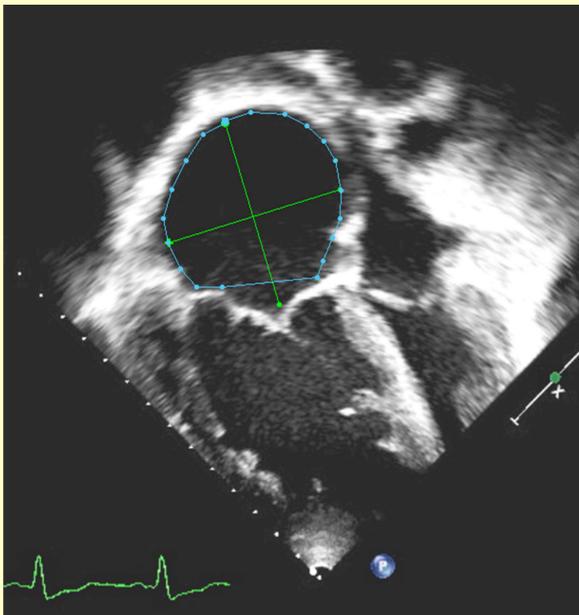
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Introduction:

The influence of growing age, body surface area (BSA), body length (BL), and body weight (BW) on RA dimensions have not been completely analyzed in healthy children and in children with volume or pressure overload. Before being able to judge RA values to be normal or abnormal in children with an atrial septal defect (ASD), or pulmonary hypertension secondary to congenital heart disease (PH-CHD), normative age-related RA size data are required. One aim of our study was to determine the normal z-score values for the RA variables end-systolic major-axis length, minor-axis length, and end-systolic area. We hypothesized that enlarged RA variables (defined as z-score > +2) can predict moderate to large secundum ASD or PH-CHD in children undergoing transthoracic echocardiography (Figure 1).

Methods:

A prospective study was conducted in a group of 516 healthy children (278 male; 238 female). The “normal echo” group encompassed neonates to adolescents (age: 1 day to 18.9 years; BW: 2.5 kg to 98 kg; BSA: 0.19 to 2.21 m²), and included 71 infants. The ASD study group consisted of 80 patients (median age: 5.1 years; range: 2 days – 18.1 years) with unrepaired isolated secundum type ASDs, moderate to large left-to-right shunting at the atrial level and signs of RA volume overload. The PH group consisted of 42 patients with PH-CHD (median age: 5.5; range 12 days to 18.2 years; BSA: 0.21 – 1.94 m²). The PH patients were all associated with CHDs according to the updated clinical classification of PH [Group 1.4.4], and included patients with post-tricuspid left-to right shunts such as VSD.



← **Figure 1:** RA measurements of the major-axis length, minor-axis length, and RA area performed in end-systole in the apical 4-chamber view. The maximal major-axis distance of the RA is from the center of the tricuspid annulus to the center of the superior RA wall, parallel to the interatrial septum. The RA major-axis dimension is represented by the longitudinal green line. The RA minor-axis length is defined from the mid-level of the RA free wall to the interatrial septum, perpendicular to the long axis. The RA minor-axis dimension is represented by the transverse green line. The RA area is traced from the lateral aspect of the tricuspid annulus to the septal aspect, excluding the area between the leaflets and annulus, following the RA endocardium, excluding the IVC and SVC and the RA appendage. The area is represented by the blue line. Abbreviations: RA, right atrium; IVC, inferior vena cava; SVC, superior vena cava.

→ **Figure 2:** Age specific z scores for (A) RA area, (B) major axis length and (C) minor axis length. Healthy subjects are given as gray circles, ASD patients as red diamonds, and PH patients as green squares. The cut off value of ± 2 z is given as broken line.

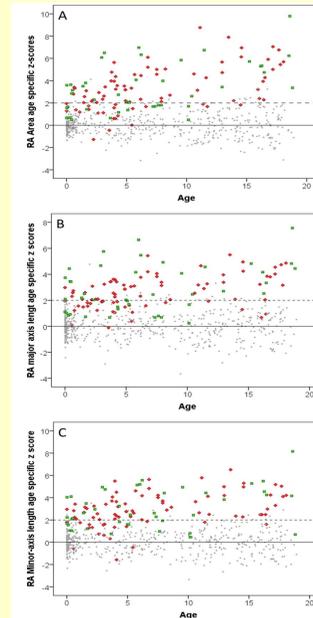


Table 1

	ASD			PHT		
	Cut off score	Sensitivity	Specificity	Cut off score	Sensitivity	Specificity
Age specific z-scores						
RA Area	2	73%	97%	2	64%	97%
RA major-axis length	2	66%	97%	2	64%	97%
RA minor-axis length	2	78%	97%	2	67%	97%
BW specific z-scores						
RA Area	2	69%	96%	2	74%	96%
RA major-axis length	2	64%	98%	2	69%	98%
RA minor-axis length	2	70%	97%	2	76%	97%
BL specific z-scores						
RA Area	2	64%	96%	2	69%	96%
RA major-axis length	2	65%	98%	2	64%	98%
RA minor-axis length	2	73%	98%	2	69%	98%
BSA specific z-scores						
RA Area	2	70%	96%	2	74%	96%
RA major-axis length	2	65%	98%	2	69%	98%
RA minor-axis length	2	74%	97%	2	76%	97%

Legends to Table 8: Impact of reference values on the detection of enlarged RAs in pediatric with ASDs and PH. For each RV dimension variables sensitivity and specificity of age-, BSA-, BW-, and BL-specific z-scores to detect ASD or PH with a cut off value of 2 are shown.

Results:

In healthy controls all RA variables, the end-systolic major-axis length, the minor-axis length, and the end-systolic area increased from neonates to adolescents in a nonlinear manner. Sixty-two out of 80 ASD patients were identified as having an enlarged RA by RA minor-axis length (sensitivity: 78 %, specificity: 97 %, with a negative predictive value of 97 % and a positive predictive value of 83 %), (Figure 2). Twenty-eight out of our 42 PH-CHD patients were identified as having an enlarged RA (sensitivity: 67 %, specificity: 97 %). Using RA major-axis length or RA area 27 out of 42 PH patients could be identified as having an enlarged RA (sensitivity: 64 %, specificity: 97 %), (Figure 2). When using a cutoff point of z-score > +2 for BSA, BW, or BL up to 74%, and 76 % of our ASD and PH-CHD patients, respectively, were identified as having enlarged RAs (Table 1).

Discussion:

We provide pediatric normative values for RA major-axis and minor-axis length and RA area. Based on these normative data, we could demonstrate an increased RA size and RA area in pediatric patients with RV volume overload (ASD) and pressure overload (PH-CHD). In our children with an ASD we found a significantly increased RA size and area when compared to healthy “normal echo” subjects. Our data demonstrate that the echocardiographic RA measurements can predict secundum ASD (64% to 78%), and maybe more importantly, RA measurements in the normal range make a significant pre-tricuspid left-to right shunt through an interatrial communication unlikely. In our pediatric PH-CHD patients significantly also increased RA size and area values compared to our healthy subjects were seen.

Conclusion:

We believe that this study is the first in children to present a correlation of normative values of RA size and area to dilated RAs in pediatric ASD and PH-CHD patients. Our study suggests that an echocardiographic evaluation a possibly enlarged RA size may be useful to guide decision making in ASD patients concerning the time point for transcatheter or surgical closure.