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Pulmonary artery dilatation due to pressure or volume overload in congenital heart disease

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The morphology and pathophysiology of the pulmonary artery (PA) is an important topic, though is less frequently analyzed in the literature. Usually dilatation of the main pulmonary artery (MPA) is considered with a diameter above 29 – 30 mm (1) (**Figure 1**).

PA dilatation is mostly described only in association with pulmonary hypertension (PH) (2), other pathophysiological factors are less well known, though long-term severe PH leads to PA wall damage, weakness, or increased fragility and aneurysmal PA dilatation.

The most relevant clinical complications that may result from PA dilatation are (2,3): external compression of the main left coronary artery by the dilated PA, PA dissection; PA thrombosis; airway compression; recurrent laryngeal nerve compression. Clinical symptoms from these complications may be life-threatening for the patients. On the other hand, many of the patients are asymptomatic from PA dilatation for a long time, but at very high risk due to their underlying disease. There are no clear guidelines about the PA dilatation cut-off points for intervention, nor the optimal timing for this.

Aim of our study was to analyze the MPA size and its dilatation in 2 groups with different underlying chronic hemodynamic pathologies: 1. PA pressure overload (due to severe PH) and 2. pure PA volume overload (due to "free" pulmonary regurgitation); and compare this with age-matched healthy volunteers.

Patients and methods

Analyzed were: 60 patients with pulmonary arterial hypertension in congenital heart disease (PAH-CHD), 64 with repaired tetralogy of Fallot/pulmonary regurgitation (rTOF/PR), and 80 healthy (NORMAL). Measured were: main pulmonary artery (MPA) diameter and MPA/ascending aorta (Ao asc) ratio, by echocardiography (ECHO) and computer tomography or magnetic resonance imaging (CT/MRI).

Results

– MPA diameter: significant difference between PAH-CHD, rTOF/PR, and NORMAL were found (median): 37 *vs* 27 *vs* 21 mm (P < 0.0001) (**Figure 2A**); with significant increase with age in all 3 groups (**Figure 3A**).

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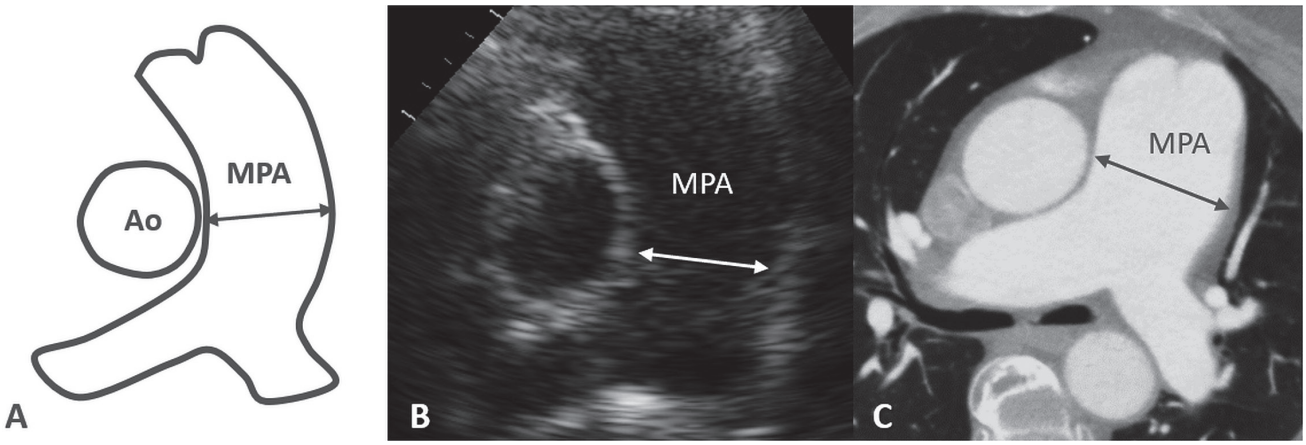

Figure 1 Pulmonary artery measurement: A. scheme; B. by echocardiography; C. by computer tomography
MPA – main pulmonary artery, Ao aorta

- Ao asc diameter: significant differences between PAH-CHD, rTOF/PR, and NORMAL were found (median): 29 *vs* 35 *vs* 26 mm (P < 0.0001) (**Figure 2B**); with significant increase with age in all 3 groups (**Figure 3B**).
- MPA/Ao asc ratio: difference found between PAH-CHD and NORMAL (median): 1.3 *vs* 0.8 (P < 0.0001) but not rTOF/PR and NORMAL: 0.74 *vs* 0.8 (P = 0.3) (**Figure 2C**); with no increase with age (**Figure 3C**).
- Significant MPA dilatation (> 40 mm): was present more frequently in patients with PAH-CHD (in 35% according to ECHO and in 76.9% according to CT/MRI); compared to rTOF/PR (in 3.1% according to ECHO and in 7.8% according to CT/MRI) – P < 0.0001. Severe MPA dilatation (> 50 mm) occurred only in PAH-CHD (in 16.7% according to ECHO and in 31.4% according to CT/MRI); while not in rTOF/PR – P < 0.0001.
- MPA/Ao asc ratio > 1: significant difference between PAH-CHD (in 88.1% patients according to ECHO; and in 100% according to CT/MRI); compared to rTOF/PR (in 10.9% according to ECHO, and in 34.5% according to CT/MRI) – P < 0.0001.
- There was a significant correlation between ECHO and CT/MRI measurements in overall evaluation of the parameters (MPA P < 0.0001; Ao asc diameter P < 0.0001; MPA/Ao asc ratio P < 0.0001), as well as in the analysis of MPA, Ao asc and MPA/Ao

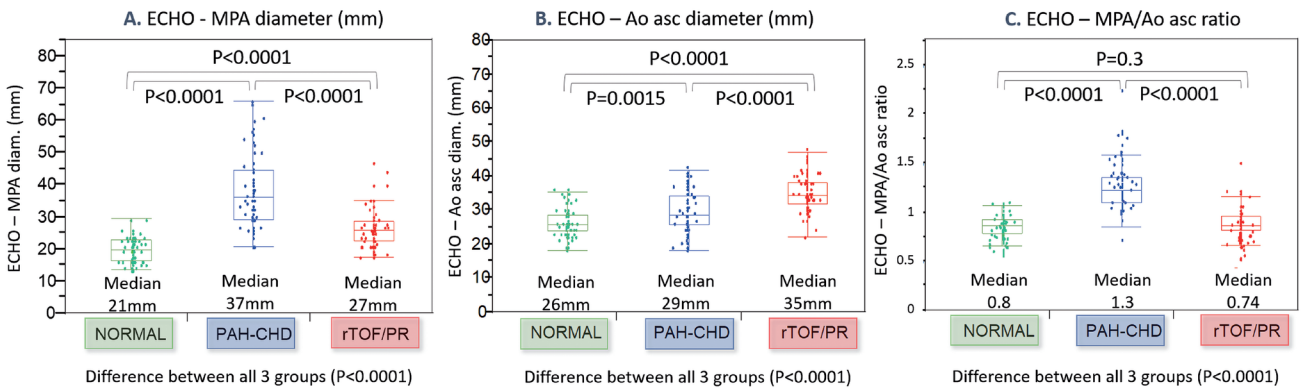

Figure 2 Parameters measured by ECHO, compared in groups (NORMAL, PAH-CHD and rTOF/PR): A. MPA diameter, B. Ao asc diameter, C. MPA/Ao asc ratio
NORMAL – control healthy subjects, PAH-CHD – pulmonary artery hypertension associated with congenital heart defects, rTOF/PR – repaired Tetralogy of Fallot with severe pulmonary regurgitation, ECHO – echocardiography, MPA – main pulmonary artery, Ao asc – ascending aorta

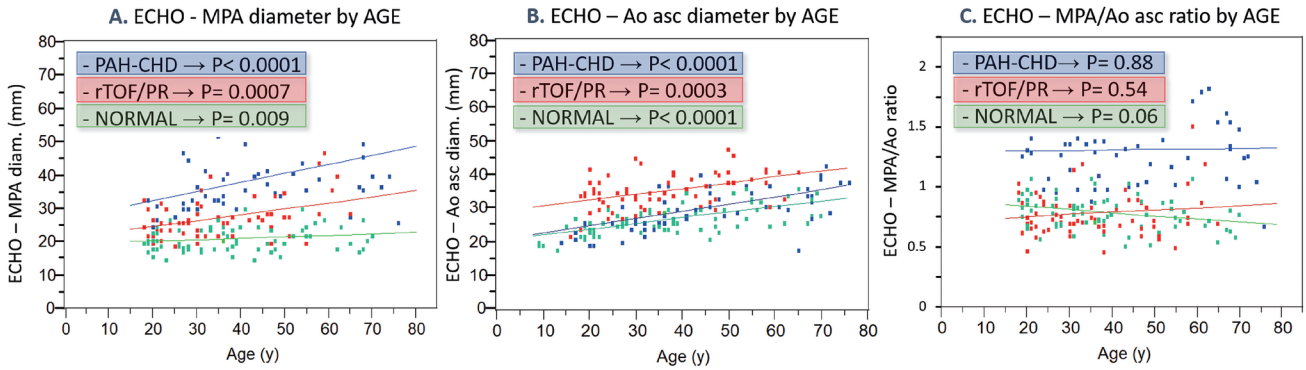

Figure 3 Correlation of parameters by age, measured by ECHO, compared in groups (NORMAL, PAH-CHD and rTOF/PR): A. MPA diameter, B. Ao asc diameter, C. MPA/Ao asc ratio

NORMAL – control healthy subjects, PAH-CHD – pulmonary artery hypertension associated with congenital heart defects, rTOF/PR – repaired Tetralogy of Fallot with severe pulmonary regurgitation, ECHO – echocardiography, MPA – main pulmonary artery, Ao asc – ascending aorta

asc ratio selectively in both patients´ groups. ECHO, though, significantly underestimated in comparison to CT/MRI, in all the analyzed parameters (**Table 1**).

Discussion

– The normal size of the MPA: most studies consider as normal MPA size < 28.9 mm for males and < 26.9 mm for females based on the Framingham Heart Study (1); though significant differences are found according to the measurement method and technique used. Most frequently CT studies established the upper normal limits up to 29 – 30 mm or even more (2-5). ECHO studies measuring MPA are very rare but the normal upper limits measured here were usually 25 – 26 mm (6,7). According to the differences and inconsistency in the measurement techniques, this topic should be addressed in larger multicentric studies and unified in future imaging guidelines.

– Correlation of the MPA diameter with the patient´s body constitution (weight, height, body surface area) is well-known (4,8); though, in our study, we have found also a significant correlation (progression) with age (in all analyzed groups).

– MPA dilatation according to the underlying etiology: the presence of MPA dilatation in PH is described almost invariably and is even considered diagnostic for the presence of PH (2,9,10). In our study a significant MPA dilatation was found both in PAH-CHD as well as in rTOF/PR, though the MPA dilatation was more frequent and more prominent in PAH-CHD patients. Understanding the pathological hemodynamics and its role in MPA dilatation is crucial. The mechanism of MPA dilatation in PH is quite well understood, the influence of PA

Table 1 Comparison of ECHO and CT/MRI – MPA and MPA/Ao asc ratio measurements

	Overall			PAH-CHD			rTOF/PR		
Analyzed parameter	ECHO Median	CT/MRI Median	P	ECHO Median	CT/MRI Median	P	ECHO Median	CT/MRI Median	P
MPA (mm)	26	33	< 0.0001	37	46	< 0.0001	27	32	< 0.0001
Ao asc (mm)	30	33	0.01	29	31.5	0.01	32	35	0.002
MPA / Ao asc ratio	0.86	1.12	< 0.0001	1.3	1.5	0.002	0.74	0.94	< 0.0001

PAH-CHD – pulmonary artery hypertension associated with congenital heart defects, rTOF/PR – repaired Tetralogy of Fallot with severe pulmonary regurgitation, ECHO – echocardiography, CT/MRI – computer tomography / magnetic resonance imaging, MPA – main pulmonary artery, Ao asc – ascending aorta

volume overload is less studied. In severe PR, the PA wall shear stress is most probably less severe than expected, and is instead influencing the RV more (11,12).

– Diagnostic assessment of the MPA diameter: in our study we confirmed a statistically good correlation between ECHO and CT/MRI measurements, though ECHO underestimated in all measured parameters.
– MPA / Ao asc ratio is often used for establishing the presence of MPA dilatation. Usually considered as pathological the MPA / Ao asc ratio > 0.9 (1,2,13), sometimes > 1.05 – 1.1 (7,14). We confirmed the usefulness of this parameter in PAH-CHD but in rTOF/PR this showed itself to be absolutely unreliable. This is due to the embryologically disproportional development of the great arteries in TOF, often with hypoplastic PA and dilated Ao (15).
– High-risk MPA dilatation: there are no clear cut-off points for establishing the severity of MPA dilatation. Usually MPA aneurysm is considered when the MPA diameter exceeds 40 mm, or 43 mm in males (7,16). On the other hand, case reports presenting severe complications related to MPA dilatations are usually in association with a bigger MPA diameter (> 50 – 55 mm) (17-20), or with a fast progression of PA dilatation (> 2 mm/year) (21). Usually the presence of PH in these cases is also reported. Close follow-up of these patients is mandatory.
– Management of patients with severe PA dilatation: there are no guidelines for indication criteria for reintervention, with or without symptoms. Patients with PH are often high-risk due to their underlying pathology, therefore the decision for reintervention (especially in an asymptomatic patient) might be very difficult. On the other hand, once symptoms appear, they very often represent a life-threatening situation (17-20).

Conclusions

- Both pressure and volume overload often lead to MPA dilatation; though in PAH-CHD (pressure overload) the MPA dilatation is usually more frequent and much more severe compared to rTOF/PR (volume overload).
- No clear MPA diameter cut-off point for the risk of severe complications (dissection, thrombosis, coronary artery compression) is established; but in patients with MPA diameter > 50 mm and/or progressive MPA dilatation (> 2 mm/year) caution and close follow-up is required, regardless of the etiology of dilatation.
- MPA/ Ao asc ratio in MPA dilatation diagnosis is useful only in PAH-CHD but not in rTOF/PR.
- It is important to differentiate the MPA measurement method; ECHO and CT/MRI correlate well but ECHO underestimates.

The authors declare no conflict of interest.

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