Compliance and Anatomy of the Neo-Aorta in Children with Hypoplastic Left Heart Syndrome (HLHS)

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Introduction

Hypoplastic left heart syndrome (HLHS) is a fatal disease of the newborn if left untreated. Surgical treatment of HLHS includes a staged reconstruction. The first of three stages of surgical palliation includes formation of a neo-aorta using aortic homograft or other graft material. This so called Norwood operation is nowadays the standard procedure in these patients, but little is known about the anatomy and compliance of the neo-aorta in later life. Echocardiography and cardiac catheterization have been the standard imaging modalities for long term follow-up. However, it has been shown that the sensitivity of echocardiography in detection stenosis of the neo-aorta is low [1] and that cardiac catheterization is associated with considerable morbidity in these patients. Cardiac magnetic resonance imaging (MRI) is increasingly used for evaluation of the cardiovascular system in congenital heart disease and has been recently established as an exact non-invasive tool for assessment of aortic compliance [2].

The purpose of this study was to assess aortic anatomy and aortic compliance in children with HLHS using MRI at 3 Tesla.

Materials and Methods

27 children (mean age 66.2 ± 37.6 months, age range 2 month - 14 years) with HLHS after second or third stage operation (creation of bidirectional cavopulmonary connection and total cavapulmonary connection, respectively) underwent a MRI study at 3 Tesla (Philips Achieva). Cine MRI and high resolution contrast enhanced time-resolved MR-angiography using a 3D keyhole technique with centric re-ordering were performed in all patients. Imaging parameters were as follows:

Cine MRI: FOV 280x224 mm, measured voxel size 1.88x1.94 mm, slice thickness 6 mm, 12 axial slices, TR / TE 4.4 / 2.5 ms, scan duration 3:50 min.

MR-angiography: FOV 380x380 mm, 70 slices, keyhole percentage 20%, 20 dynamics, scan time per volume1.7 s, TR / TE 2.4 / 0.93 ms, scan duration 0:40 min.

The 3D images were used for measuring diameters of the reconstructed aorta at five selected segments (Figure 1). The cine loops were used for determination of the aortic compliance at four selected segments (Figure 2).

Non-invasive blood pressure measurements (BP) were performed eight times during the whole examination by means of an MRI compatible sphygmomanometer cuff (Precess™, Serie 3610, Invivo Germany GmbH). The average of the BP measurements was used for compliance estimates.

The largest and smallest cross-sectional areas at selected locations of the neo-aorta were determined by manual tracing. Compliance (C) was calculated according to the following standard formula [3]:

$C\ (10^{-3} mmHg^{-1}) = (A_{max} - A_{min})/[A_{min} \times (P_{max} - P_{min})]$

where A_{max} and A_{min} are the maximal and minimal cross-sectional lumen areas (in millimetres squared) of the neo-aorta, and P_{max} and P_{min} are the average systolic and diastolic blood pressures (mm Hg).

We compared the measured diameters to normal values for healthy children generated by echocardiography and cardiac catheterisation [4, 5], neo-aortic compliance was compared to normal values acquired by MRI [6].

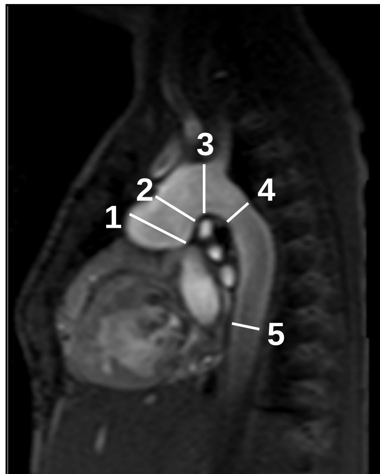

Figure 1: Selected segments for measuring diameters of the neo-aorta: Aortic root (1), ascending aorta (2), aortic arch (3), aortic isthmus (4) and descending aorta (5).

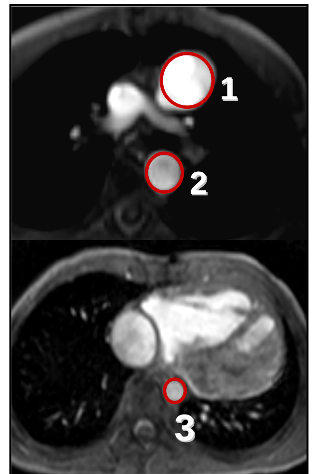

Figure 2: Selected segments for determination of aortic compliance: Ascending aorta (1), aortic arch (not shown) and at two levels of the descending aorta (2 and 3).

Variables	Patients (n= 27)	Normal values	P
Diameters			
aortic root	25.0 ± 4.7	19.4 ± 3.2	< 0.01
ascending aorta	21.6 ± 4.3	16.4 ± 2.8	< 0.01
aortic arch	27.3 ± 4.1	12.3 ± 2.1	< 0.01
aortic isthmus	9.5 ± 2.8	11.2 ± 1.8	< 0.01
descending aorta	12.5 ± 4.0	10.9 ± 2.0	0.09
Compliance			
ascending aorta	4.5 ± 3.0	8.61 ± 2.22	
aortic arch	5.1 ± 3.9	-	
descending aorta, segment 1	10.0 ± 6.2	8.66 ± 3.85	
descending aorta, segment 2	13.5 ± 7.2	8.73 ± 2.39	

Table 1: Diameters (upper part) and compliance (lower part) of selected segments in HLHS patients compared to normal values (mean values shown in mm and $10^{-3}mmHg^{-1}$ respectively, ± 1 SD).

Results and Discussion

The diameters of the aortic root, the ascending aorta and the aortic arch were significantly increased in HLHS patients compared to normal values generated by echocardiography and cardiac catheterization (Table 1, upper part).

On the other hand we found a significant smaller aortic isthmus in children with HLHS, whereas the dimensions of the descending aorta were not significantly changed. Aortic compliance was decreased in the ascending aorta as well as the aortic arch in comparison to values for healthy subjects (Table 1, lower part). We have demonstrated the feasibility of using cine MRI and 3D angiography at 3T without steady state free precession to assess aortic dimensions and aortic compliance.

References

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