Reduced ascending aorta distensibility relates to adverse ventricular mechanics in patients with hypoplastic left heart syndrome: Noninvasive study using wave intensity analysis

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Objective: To evaluate the aortic arch elastic properties and ventriculoarterial coupling efficiency in patients with single ventricle physiology, with and without a surgically reconstructed arch.

Methods: We studied 21 children with single ventricle physiology after bidirectional superior cavopulmonary surgery: 10 with hypoplastic left heart syndrome, who underwent surgical arch reconstruction, and 11 with other types of single ventricle physiology but without arch reconstruction. All children underwent pre-Fontan magnetic resonance imaging. No patient exhibited aortic recoarctation. Data on aortic wave speed, aortic distensibility and wave intensity profiles were all extracted from the magnetic resonance imaging studies using an in-house–written plug-in for the Digital Imaging and Communications in Medicine viewer OsiriX.

Results: Children with hypoplastic left heart syndrome had significantly greater wave speed ($P = .002$), and both stiffer ($P = .004$) and larger ($P < .0001$) ascending aortas than the patients with a nonreconstructed arch. Aortic distensibility was not influenced by ventricular stroke volume but depended on a combination of increased aortic diameter and abnormal wall mechanical properties. Those with hypoplastic left heart syndrome had a lower peak wave intensity and reduced energy carried by the forward compression and the forward expansion waves, even after correction for stroke volume, suggesting an abnormal systolic and diastolic function. Lower wave energy was associated with an increased aortic diameter.

Conclusions: Using a novel, noninvasive technique based on image analysis, we have demonstrated that aortic arch reconstruction in children with hypoplastic left heart syndrome is associated with reduced aortic distensibility and unfavorable ventricular-vascular coupling compared with those with single ventricle physiology without aortic arch reconstruction. (J Thorac Cardiovasc Surg 2012;144:1307-14)

The first-stage palliative procedure for the treatment of hypoplastic left heart syndrome (HLHS) or Norwood procedure[1] requires extensive surgical reconstruction of the aortic arch with augmentation, typically using a pulmonary homograft patch.[2] Despite acceptable short-term outcomes,[3] long-term concern exists regarding the function of the right ventricle in a systemic position. Recent evidence suggests that Fontan patients with a systemic, single, right ventricle are at increased risk of ventricular dysfunction[4] and reduced exercise capacity[5] in the mid to long term. Although progressive attrition is thought to be related to abnormal ventricular structure, or even inherent to the Fontan physiology, concern exists that increased afterload could be the cause of premature ventricular dysfunction.[6] For example, in this setting, aortic coarctation is known to lead to ventricular dysfunction.[7] However, it is now becoming evident that ventricular afterload can be increased even by mechanisms different from vascular stenosis, such as increased aortic input and characteristic impedance and increased wave reflection.[8,9]

Additional surgical factors could play an important role in determining the increased ventricular afterload. Recent evidence has suggested that patients with HLHS have reduced distensibility of the ascending aorta[10-12] as a consequence of extensive reconstruction of the aortic arch at the Norwood operation. However, the relationship between the abnormal elastic properties of the proximal aorta and aortic arch and ventricular mechanics in this setting has not been assessed in full.

The aim of the present study was to noninvasively assess the mechanical properties of the ascending aorta in patients with single ventricle physiology after superior bidirectional cavopulmonary anastomosis and to evaluate the influence of

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Disclosures: Authors have nothing to disclose with regard to commercial support.

Read at the 92nd Annual Meeting of The American Association for Thoracic Surgery, San Francisco, California, April 28-May 2, 2012.

Received for publication April 11, 2012; revisions received Aug 1, 2012; accepted for publication Aug 7, 2012; available ahead of print Oct 1, 2012.

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0022-5223/$36.00

http://dx.doi.org/10.1016/j.jtcvs.2012.08.028

Abbreviations and Acronyms

FCW = forward compression wave
FEW = forward expansion wave
HLHS = hypoplastic left heart syndrome
LV = left ventricular
MRI = magnetic resonance imaging
SV = stroke volume

abnormal aortic mechanical properties on ventricular function and the efficiency of vascular-ventricular coupling.

METHODS

Patients

We selected 21 children (mean age, 4.0 ± 1.4 years) with single ventricle physiology who had undergone magnetic resonance imaging (MRI) under general anesthesia as a preparation for Fontan completion. Of the 21 children, 10 had HLHS and 11 had other forms of single ventricle physiology without systemic outflow obstruction or aortic coarctation (tricuspid atresia with ventriculoarterial concordance in 4, pulmonary atresia in 4, double-outlet right ventricle in 2, and double inlet left ventricle in 1). All patients had received a bidirectional superior cavopulmonary anastomosis as a part of their staged surgical management at the time of the study. All patients with HLHS had undergone standard surgical reconstruction of the aortic arch with a homograft patch at the Norwood procedure, with all procedures performed by only 2 surgeons. None of the other 11 children with other forms of single ventricle physiology had undergone any ascending aorta or aortic arch surgery, and they represented the control group.

MRI Data

All patients underwent MRI (1.5-T Avanto; Siemens Medical Solutions, Erlangen, Germany) before total cavopulmonary connection Fontan completion. Phase-contrast MRI was used for flow quantification. Through-plane flow data (about 30 ms temporal resolution, interpolated to 30 frames per heart beat) were acquired with the use of retrospective cardiac gating. Also, retrospectively gated, balanced, steady-state free precession cine images of the heart were acquired in the vertical long-axis, 4-chamber, and short-axis view covering the entirety of both ventricles (9-12 slices). Post-processing was performed using OsiriX (Pixmeo, Geneva, Switzerland) with in-house–written plug-ins. Single ventricle end-diastolic and end-systolic volumes were measured. The stroke volume (SV), ejection fraction, and cardiac output were calculated from these measurements. The ventricular volumes and cardiac output were indexed for body surface area. During the cardiac MRI scan, the blood pressure was measured every 5 to 10 minutes noninvasively with a cuff of appropriate size placed around the right arm or right leg. The systolic and diastolic blood pressure at the beginning of the scan was recorded, and the pulse pressure was calculated as the systolic minus the diastolic blood pressure.

Care was taken to ensure that, in the HLHS group, the imaging plane was always above the aortic–pulmonary anastomosis to ensure that the reconstructed aorta was captured. In all cases, the ascending aortic data was acquired just above the Damus-Kaye-Stansel anastomosis using a standard method, according to our institutional pre-Fontan completion protocol.

Institutional ethical approval for the retrospective use of the MRI data was obtained, and all patients' parents gave informed consent for research use of the imaging data.

Calculation of Aortic Distensibility

All calculations were semiautomated by the use of an in-house–written OsiriX plug-in.[13]

Aortic distensibility (D) was derived directly from knowledge of wave speed (c), according to the Bramwell-Hill equation:

$$D = \frac{1}{\rho c^2} \tag{1}$$

where ρ is the blood density. The calculation of wave speed (c) was determined from the area (A) and velocity (U) information, directly derived from the MRI data. The ascending aortic MRI flow data were segmented using a validated semiautomatic registration-based segmentation algorithm,[14] allowing extraction of the U and A signals. Changes in velocity (dU) and fractional changes in area (dlnA) were related using the water hammer equation:

$$dU_{\pm} = \pm c d\ln A_{\pm} \tag{2}$$

According to this equation, the U-lnA relationship should have a linear slope yielding the wave speed in early systole, when no backward waves are expected. This holds true for noncoronary arteries. The aortic U-lnA relationship is represented by a loop, similar to the pressure-velocity loop[15] and diameter-velocity loop[16] methods for wave speed calculation. All analyses were performed by the same operator.

Distensibility was also assessed in terms of stress (σ) and strain (ε) to evaluate the effect of the presence of the patch. Variations in diameter, d (Δd) over the diastolic diameter (d_{dias}) yielded an indication of circumferential strain ($\varepsilon = \Delta d/d_{dias}$), and circumferential stress was defined as follows:

$$\sigma = \frac{Pd}{2h} \tag{3}$$

where P is the pulse pressure, and h is the wall thickness.

Wave Intensity Analysis

Although the formulation of wave intensity is traditionally determined from the pressure and velocity[17] and, more recently, from diameter and velocity,[18] we have proposed a formulation using area and velocity.[13] Using the area and velocity, the net wave intensity (dI_A) and separated wave intensity ($dI_{A\pm}$) are respectively defined as follows:

$$dI_A = dU d\ln A \tag{4}$$

and

$$dI_{A\pm} = \pm\frac{c}{4}\left(d\ln A \pm \frac{1}{c}dU\right)^2 \tag{5}$$

Having obtained a dI_A pattern, the waves can be identified as compression ($d\ln A > 0$) or expansion ($d\ln A < 0$) waves. Depending on whether they are traveling away or toward the heart, the waves can also be defined as forward-traveling or backward-traveling, respectively. Traditionally, left ventricular (LV) ejection is described by a forward compression wave (FCW), and LV relaxation is described by a forward expansion wave (FEW). It has been shown in normal subjects that FCW correlates significantly with a maximum rate of pressure rise (dP/dt) and that the FEW correlates significantly with the LV relaxation time constant.[19] On this basis, the peak intensity of, and the energy carried by, the FCW and FEW were calculated. Wave energy (I) was derived from the area under the wave intensity curve, as follows:

$$I = \int_{t1}^{t2} dI_A dt \tag{6}$$

where t_1 and t_2 indicate, respectively, the onset and arrival of each wave. The energy carried by the FCW and FEW was also indexed for the SV, as an indication of the energy carried by a single unit of ejected volume.

It should be noted that the units of wave intensity and wave energy are not the traditional [W/m^2] and [J/m^2] but rather [m/s] and [m], respectively.