Noninvasive Assessment of Vascular Function and Hydraulic Power and Efficiency in Pediatric Fontan Patients

Kimberley A. Myers, MD, Mande T. Leung, PhD, M. Terri Potts, RDCS, James E. Potts, PhD, and George G. S. Sandor, MB, ChB, *Vancouver, British Columbia, Canada*

Background: Invasive studies have shown that children with Fontan palliation have abnormal arterial stiffness, impedance, and hydraulic power and efficiency. The aim of this study was to assess these indexes noninvasively in a cohort of children with Fontan circulation using Doppler echocardiography and compare their results with those of healthy peers.

Methods: This was a case-control study of 22 Fontan patients and 31 healthy control children. Using standard two-dimensional, M-mode, and Doppler echocardiographic imaging and carotid artery applanation tonometry, aortic flows, dimensions, and pulse-wave velocity were measured, and vascular impedance and arterial stiffness were calculated. Hydraulic power and efficiency were calculated from standard fluid dynamics formulae.

Results: The median age was similar between groups. Stroke volume index (39 vs 39 mL/min/m^2) and cardiac index (2.6 vs 2.5 L/min/m^2) were similar. Aortic cross-sectional area (3.3 vs 2.8 cm^2), peak aortic flow (302 vs 261 cm^3/sec), and myocardial performance index (0.47 vs 0.25) were higher and ejection fraction (50% vs 66%) was lower in Fontan patients. Input impedance (61 vs 83 dyne · sec/cm^5/m^2) was lower in Fontan patients. Pulse-wave velocity (488 vs 364 cm/sec), elastic pressure-strain modulus (305 vs 263 torr), and stiffness index (4.15 vs 3.04) were higher in Fontan patients. Total arterial compliance (1.29 vs 1.32 mL/torr/m^2) and mean power (606 vs 527 mW/m^2) were similar and total hydraulic power (716 vs 627 mW/m^2) was higher in Fontan patients. Efficiency and the power cost per unit of forward flow were similar.

Conclusions: Despite stiffer aortas, Fontan patients generate more hydraulic power associated with decreased ventricular function to achieve a similar hydraulic efficiency. In Fontan patients, therapy that is given to improve ventricular function may need to target vascular stiffness as well. This technique may be used to monitor the efficacy of therapeutic interventions. (J Am Soc Echocardiogr 2013;26:1221-7.)

Keywords: Fontan procedure, Pulse-wave velocity, Vascular impedance, Hydraulic power and efficiency

Long-term follow-up studies of patients with Fontan palliation have shown that many patients develop ventricular failure.[1] This may be due to a number of factors, including the type of ventricular geometry, disadvantageous myocardial fiber orientation, prior hypoxia, chronic volume loading, and myocardial bypass during surgery. In a group of younger children with Fontan palliation for single-ventricle physiology, invasive studies have shown decreased resting cardiac index (CI), reduced preload reserve, and increased systemic vascular resistance.[2,3] Until recently, the possible effects of abnormal vascular properties or the role of hydraulic power (W) and hydraulic efficiency (W_m/W_t) on ventricular function has received little attention.[3,4] Noninvasive methods of determining the biophysical properties of the aorta have been described.[5,6] These include pulse-wave velocity (PWV), arterial stiffness (elastic pressure-strain modulus [Ep] and stiffness index [β index]), and impedance (input impedance [Zi] and characteristic impedance [Zc]). The systemic ventricle can be considered to produce external work in the form of blood pressure and flow. Metabolic demands are met by increasing ventricular hydraulic power at the expense of lower ventricular efficiency. Total power (W_t) consists of steady (mean power [W_m]) and pulsatile components. The latter is a small component of the W_t generated. W_m describes the physiologically useful hydraulic power required to pump blood, pulsatile power describes energy lost in arterial pulsations, and the ratio of W_m to W_t has been used to describe ventricular-vascular coupling.[7,8] Noninvasive methods of assessing these indexes have been developed as research tools.[9]

In view of the problem of the long-term development of ventricular dysfunction, our hypothesis was that the biophysical properties of the aorta, hydraulic power, and efficiency would be abnormal in

From the Children's Heart Centre, British Columbia Children's Hospital, and the Department of Pediatrics, University of British Columbia, Vancouver, British Columbia, Canada.

Reprint requests: George G. S. Sandor, MB, ChB, Children's Heart Center, British Columbia Children's Hospital, 4480 Oak Street, Room 1C49, Vancouver, BC V6H 3V4, Canada (E-mail: gsandor@cw.bc.ca).

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Abbreviations

ACE = Angiotensin-converting enzyme

CI = Cardiac index

Ep = Elastic pressure-strain modulus

MPI = Myocardial performance index

PWV = Pulse-wave velocity

TAC = Total arterial compliance

W = Hydraulic power

W_m = Mean power

W_m/W_t = Hydraulic efficiency

W_t = Total power

W_t/CI = Power cost per unit of forward flow

Zc = Characteristic impedance

ZcF = Fourier-derived characteristic impedance

Zi = Input impedance

ZiF = Fourier-derived input impedance

a population of pediatric patients with Fontan circulation. Our objectives were to assess ventricular function, PWV, arterial stiffness, and impedance[5,6] and, for the first time, to describe a noninvasive method of measuring power and W_m/W_t in these children and compare them with controls.

METHODS

Patients

Twenty-two patients with Fontan circulation followed at the Children's Heart Centre at British Columbia Children's Hospital were assessed prospectively between January 2007 and July 2008. To minimize confounding factors in this initial study, patients with hypoplastic left heart syndrome or surgical revision of the aorta, including the Damus-Kaye-Stansel procedure, were excluded from the study. The median age at completion of the Fontan procedure was 3.1 years (range, 2.2–11.5 years). The cardiac diagnoses are listed in Table 1. None of the patients had significant aortic or atrioventricular valve regurgitation. A list of current medications was compiled for the Fontan group. The control group comprised 31 age-matched healthy children, all with normal results on cardiac examination and on echocardiography, selected from our data pool of volunteers who were tested between August 2005 and July 2010.

Data Acquisition

Standard two-dimensional, M-mode, and Doppler echocardiography were performed on all patients. The techniques that we used to measure PWV, arterial stiffness, and impedance have been described previously.[5,6] To acquire the data to calculate W_t, W_m, and W_m/W_t, carotid pressure waveforms were recorded with applanation tonometry using a Millar pulse transducer (model SPT-301; Millar Instruments, Inc., Houston, TX) connected via a control box (model SD-640; Millar Instruments, Inc.) to a GE Vivid 7 Pro ultrasound machine (GE Healthcare, Wauwatosa, WI). The carotid pressure waveforms were obtained simultaneously with pulse-wave Doppler waveforms in the ascending aorta and sphygmomanometric measurement of the left brachial artery blood pressure in the supine position. Care was taken to obtain Doppler envelopes with minimal spectral broadening, and patients with turbulent flow were excluded. Three pressure waveform beats of the highest quality were selected and averaged (Figure 1). In calibrating the tonometry pressure, diastolic and mean pressure were assumed to be the same at the carotid artery and at the brachial artery.[10] Fourier analysis of the pressure and flow data derived from the aortic and carotid waveforms was used to calculate vascular impedance and left ventricular W_t, W_m, and W_m/W_t.

Table 1 Anatomic diagnoses of the Fontan patients

Diagnosis	*n*
Tricuspid atresia	8
Pulmonary atresia, intact ventricular septum	4
Unbalanced atrioventricular septal defect	6
Univentricular heart of left ventricular morphology	2
Univentricular heart of right ventricular morphology	2

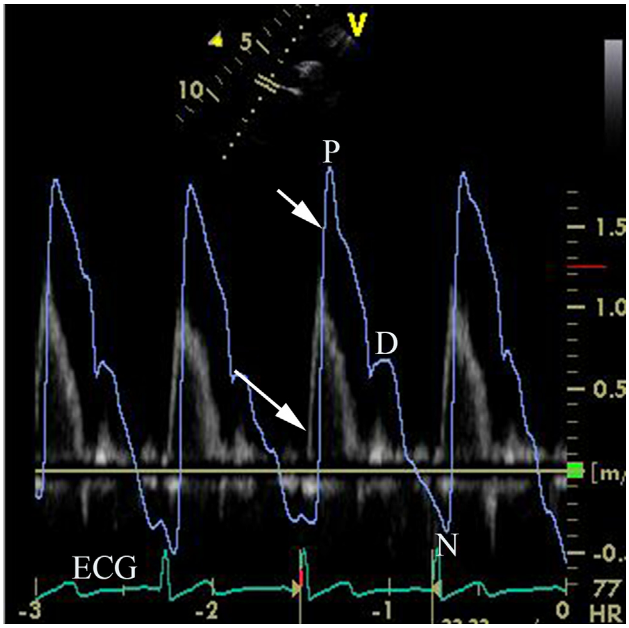

Figure 1 An example of a carotid pulse tracing and ascending aortic Doppler flow taken from the suprasternal notch during echocardiography in a control subject. The sample volume is placed centrally in the ascending aorta just distal to the aortic valve. The *short arrow points* to the carotid pulse tracing. The magnification of the tracing is adjusted to fit on the screen. The *longer arrow points* to the onset of Doppler flow. *D*, Dicrotic notch; *ECG*, electrocardiogram; *N*, nadir of the pressure tracing; *P*, peak of the pressure tracing.

The ventricular ejection fraction was calculated using echocardiography, with end-systolic and end-diastolic volumes estimated using Simpson's method for biplane ejection fraction.

The myocardial performance index (MPI) was calculated using pulse Doppler, as described by Tei *et al.*[11]

Data Analysis

The formulas for the calculations of the following are listed in the Appendix: transit time, PWV, pulse pressure, peak aortic flow, Ep, β index, Zi, Zc, mean brachial artery pressure, and total arterial compliance (TAC).

The aortic flow waveform, aortic cross-sectional area, aortic impedance spectrum, Fourier-derived Zi (ZiF) and Fourier-derived Zc (ZcF), W_t and W_m, W_m/W_t, and the power cost per unit of forward flow (W_t/CI) were calculated from the carotid pressure and flow waveforms. TAC, ZiF, ZcF, W_t, and W_m were normalized to body surface area.

Patient and control data were measured separately by two investigators (K.A.M., G.G.S.S.). ZiF, ZcF, W_t, W_m, W_m/W_t, Wt/CI, and TAC