Looks Do Matter! Aortic Arch Shape After Hypoplastic Left Heart Syndrome Palliation Correlates With Cavopulmonary Outcomes

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Background. **Aortic arch reconstruction after hypoplastic left heart syndrome (HLHS) palliation can vary widely in shape and dimensions between patients. Arch morphology alone may affect cardiac function and outcome. We sought to uncover the relationship of arch three-dimensional shape features with functional and short-term outcome data after total cavopulmonary connection (TCPC).**

Methods. **Aortic arch shape models of 37 patients with HLHS (age, 2.89 ± 0.99 years) were reconstructed from magnetic resonance data before TCPC completion. A novel, validated statistical shape analysis method was used to compute a three-dimensional anatomic mean shape from the cohort and calculate the deformation vectors of the mean shape toward each patient's specific anatomy. From these deformations, three-dimensional shape features most related to ventricular ejection fraction, indexed end-diastolic volume, and superior cavopulmonary pressure were extracted by partial least-square regression analysis. Shape patterns relating to intensive care unit and hospital lengths of stay after TCPC were assessed.**

Results. **Distinct deformation patterns, which result in an acutely mismatched aortic root and ascending aorta, and a gothic-like transverse arch, correlated with increased indexed end-diastolic volume and higher superior cavopulmonary pressure but not with ejection fraction. Specific arch morphology with pronounced transverse arch and descending aorta mismatch also correlated with longer intensive care unit and hospital lengths of stay after TCPC completion.**

Conclusions. **Independent of hemodynamically important arch obstruction, altered aortic morphology in HLHS patients appears to have important associations with higher superior cavopulmonary pressure and with short-term outcomes after TCPC completion as highlighted by statistical shape analysis, which could act as adjunct to risk assessment in HLHS.**

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Stage 1 palliation for hypoplastic left heart syndrome (HLHS) requires a modified Damus-Kay-Stansel procedure with reconstructive augmentation of the aortic arch, typically using a homograft patch of varying sizes and shapes. As a consequence, the resultant morphology of the neoascending aorta and aortic arch can be highly variable from patient to patient in addition to differing incidences of residual arch obstruction, dilatation, and tortuosity. Recently, late systemic hypertension in patients after successful aortic coarctation repair was found to relate to deranged aortic arch shape [1]. Although residual coarctation and recurrent arch obstruction are known to be associated with worse cardiac function and poorer outcomes in HLHS patients [2, 3], a recent study using wave intensity analysis has demonstrated that aortic arch shape features, such as grossly mismatched dimensions between the transverse and descending aorta, can also lead to maladaptive ventriculoarterial coupling and reduced ventricular ejection fraction [4].

In this study we analyzed in greater depth and detail arch shape features in patients who have undergone the stage 1 Norwood aortic arch reconstruction. Being widely variable in both shape and size, surgically reconstructed aortic arches in HLHS cannot be adequately analyzed by traditional morphometric methods using only two-dimensional (2D) measures, such as lengths and diameters, because these are insufficient to provide a comprehensive description of the multitude of

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*Modeling of Congenital Hearts Alliance (MOCHA) Collaborative Group participants are listed in the Appendix.

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morphologic permutations. We therefore applied a novel, validated three-dimensional (3D) statistical shape analysis method (SSM) that quantitatively evaluates the ascending aorta and arch morphology as a single, contiguous 3D unit, without the need for manually measuring the numerous dimensions [5–7]. We hypothesized that 3D arch shape features extracted using the SSM would be associated with the functional status of the stage 2 superior cavopulmonary circulation and with short-term clinical outcome after stage 3 total cavopulmonary connection (TCPC).

Patients and Methods

Patient Population

We retrospectively analyzed cardiovascular magnetic resonance (CMR) data of 37 patients (11 girls; mean age, 2.89 ± 0.99 years) with HLHS as the primary diagnosis who had undergone stage 1 Norwood-type aortic arch reconstruction (12 right ventricular–to–pulmonary arterial shunts, and the rest modified Blalock-Taussig shunts) and stage 2 superior cavopulmonary (bidirectional Glenn) palliation. In all patients, a standard Damus-Kay-Stansel aortic arch reconstruction was performed, followed by patch augmentation of the aortic arch using pulmonary arterial homograft, without coarctectomy. The homografts were fashioned per surgeon preference and expertise, but typically beginning with a triangular-shaped patch. Nine patients had undergone balloon dilatation owing to aortic recoarctation before, or concomitant aortic arch repair during, the stage 2 procedure.

CMR examination was routine in all patients in preparation for stage 3 TCPC completion. At the time of CMR and TCPC completion, none of the 37 patients had hemodynamically significant residual aortic arch obstruction requiring revision, as determined by Doppler echocardiographic interrogation, done as part of the routine pre-TCPC assessment. Ethical approval was obtained for the use of image data for research purposes, and all parents/legal guardians gave informed consent for research use of the data.

CMR Imaging and Processing

CMR data were acquired during middiastolic rest using a 3D balanced, steady-state free precession whole-heart sequence on a 1.5 T Avanto MR scanner (Siemens

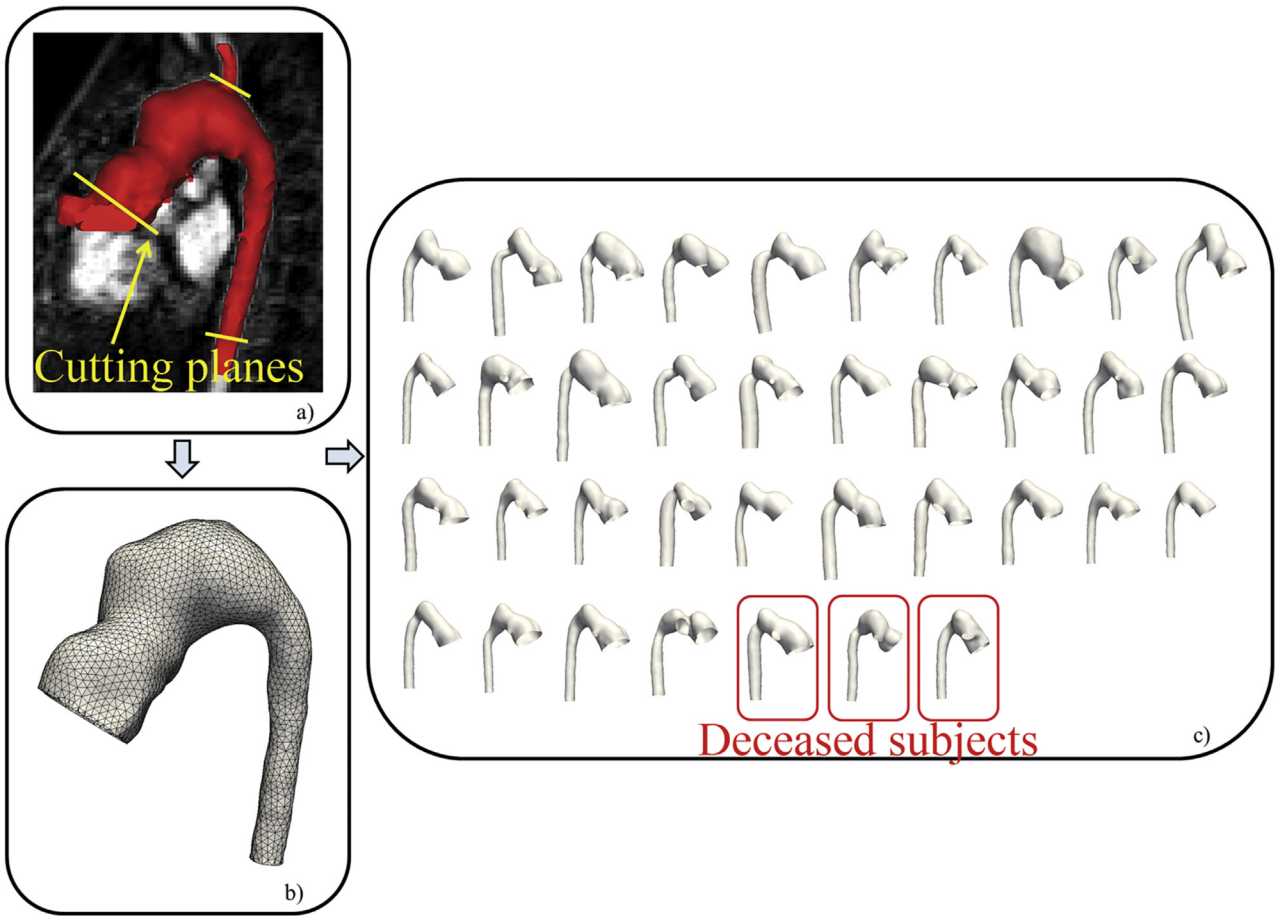

Fig 1. (a) Aortic arch volumes were segmented from cardiac magnetic resonance data and cut at the subannular plane and at the level of the diaphragm. Head and neck, coronary vessels, as well as the native aorta were cut off as close as possible to the arch. (b) A three-dimensional (3D) surface mesh. (c) The 3D surface meshes of all 37 patients constituted the input for the statistical shape model. The 3 patients who died are marked in red.