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LETTER TO THE EDITOR

Arch Hypoplasia and Aneurysm After Aortic Coarctation Repair

Abnormal Flow May Be the Link

Aortic arch hypoplasia has been linked to aneurysm formation after coarctation repair, with abnormal blood flow proposed as a mechanism (1). Time-resolved, 3-dimensional phase-contrast magnetic resonance imaging (4-dimensional flow) allowed dynamic visualization of flow and computation of vectorial wall shear stress in 5 patients after coarctation repair, 4 with arch hypoplasia (2). Symmetrical flow and wall shear stress were demonstrated in the case with normal arch geometry, whereas asymmetrically elevated systolic blood flow and wall shear stress were shown in the cases with arch hypoplasia. These findings suggest that aberrant blood flow may contribute to aneurysm formation in this context.

Four-dimensional flow was used to visualize systolic blood flow and calculate wall shear stress in 5 patients after coarctation repair, 4 with arch hypoplasia. Maximum intensity projection magnetic resonance angiography is provided to demonstrate aortic arch geometry in all cases. Peak systolic velocity–coded streamlines show localized flow velocity throughout the aortic arch (Fig. 1), and cross sections show velocity and wall sheer stress at the coarctation repair site; green bars represent the magnitude of shear stress vectors. Normal systolic streamlines and symmetrical wall shear stress were demonstrated in a patient with normal arch geometry

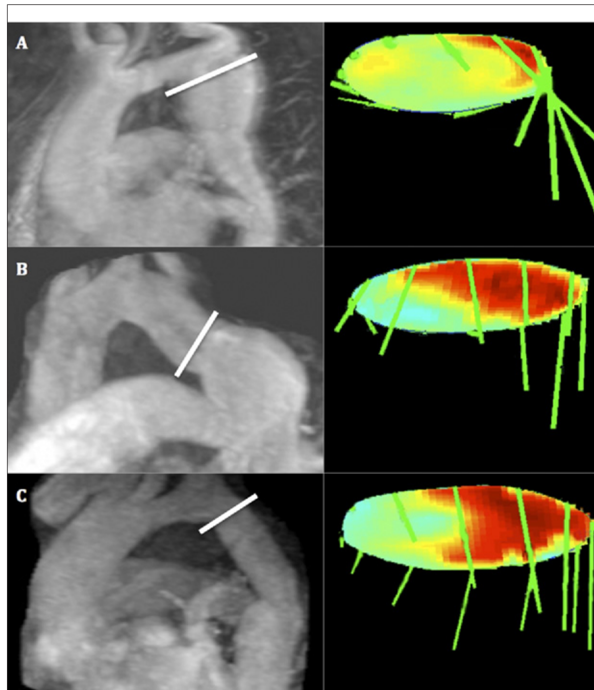

Figure 2. Wall Shear Stress Downstream of Hypoplastic Arch After Aortic Coarctation Repair

Magnetic resonance angiography with corresponding cross-sectional analysis at the planes indicated demonstrate eccentric systolic flow resulting in focally elevated wall shear stress in 3 patients with arch hypoplasia after coarctation repair, 2 with repair site aneurysms **(A, B)**. Dynamic visualization of blood flow with a particle trace for case A can be found as Online Video 1.

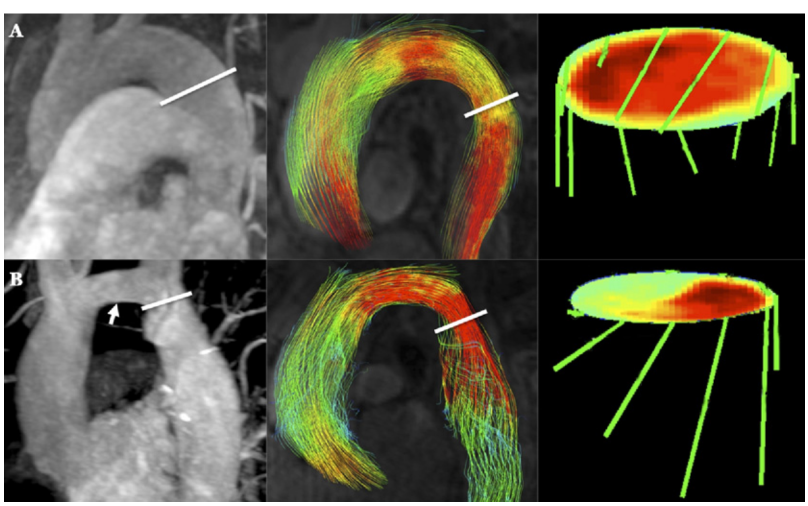

Figure 1. Aortic Blood Flow After Coarctation Repair With Normal and Hypoplastic Aortic Arch

(A) From left to right: Magnetic resonance angiography, systolic streamlines and cross-sectional analysis at the plane indicated, with relative shear stress represented by **green bars**, exhibit normal flow in a patient with normal arch geometry after coarctation repair. **(B)** Identical analysis reveals a hypoplastic aortic arch **(arrow)** after coarctation repair resulting in eccentric flow and asymmetric and elevated wall shear stress.

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(Fig. 1A). However, with arch hypoplasia (Fig. 1B), accelerated flow through the narrowed arch resulted in elevated and asymmetrical shear stress in the distal arch (Fig. 1B).

Asymmetrically elevated wall shear stress on the posterior wall of the aorta downstream of a hypoplastic arch after coarctation repair is further demonstrated in 3 patients, 2 with an aneurysm (Fig. 2). Time-resolved evaluation of the case depicted in Figure 2A reveals that accelerated flow through the hypoplastic arch impacts the posterior wall and then spins in a large helix within a repair site aneurysm (Online Video 1).

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ARVC/D Task Force Imaging Criteria

It Is Difficult to Get Along With the Guidelines

In a recent study published in *iJACC*, Vermes et al. (1) concluded that the revision of the Arrhythmogenic Right Ventricular Cardiomyopathy/Dysplasia (ARVC/D) Task Force imaging criteria significantly reduced the overall prevalence of major and minor criteria in cardiac magnetic resonance (CMR) studies (2). The study was accompanied by a very interesting editorial by Bluemke (3), who concluded that the quantitative CMR metrics proposed in the modified task force criteria are useful only "if the CMR laboratory develops its own standards for normal subjects" and if these are the same as previously reported for the MESA (Multi-Ethnic Study of Atherosclerosis) study, which served as a normal reference for comparison with the ARVC/D population.

However, matters become complicated when new echocardiographic ARVC/D Task Force criteria are considered. The new task force recommendations propose as major ARVC/D diagnostic criteria cutoff values of ≥32 mm for diastolic right ventricular outflow tract (RVOT) diameter in the parasternal long-axis (PLAX) view and ≥36 mm for diastolic RVOT diameter in the short-axis view. The respective cutoff values for minor criteria are ≥29 to <32 mm for the diastolic RVOT PLAX view and ≥32 to <36 mm for the diastolic RVOT short-axis view (2). Sensitivity and specificity for the proposed echocardiographic criteria are provided, but contrary to CMR criteria, the task force document does not refer to any specific study (2).

The echocardiographic cutoff values follow neither those proposed by Yoerger et al. (4) nor those proposed by the North American Multidisciplinary Study (5) (both of which are cited in the ARVC/D Task Force document): the RVOT PLAX cutoff value and fractional area change (FAC) proposed by Yoerger et al. (4) are >30 mm and <32%, respectively, and the FAC cutoff value cited in the North American Multidisciplinary Study is ≤26%. FAC cutoff values proposed by the task force are ≤33% and ≤40% (2).

Confusion grows when one refers to the recent American Society of Echocardiography (ASE) guidelines for the echocardiographic assessment of the right heart in adults (6). The cutoff value for the diastolic RVOT PLAX view in these guidelines is ≥32 mm; hence, the revised ARVC/D Task Force minor criteria for the diagnosis of ARVC/D (≥29 to <32 mm) are well within the normal limits recommended by the ASE. To further complicate the situation, the RVOT PLAX measurement techniques proposed by Yoerger et al. (4) and the ASE (6) are not perfectly compatible. Moreover, abnormal FAC is defined as <35% by the ASE and as ≤40% (minor criteria) and <33% (major criteria) by the task force; many normal ASE measurements fulfill the minor ARVC/D diagnostic criteria (2,6).

In conclusion, there is an urgent need to unify the 2 important guideline documents (2,6). Until that happens, because of major discrepancies, we shall not be able to "get along with the guidelines."

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