Commentary: Never Argue with Success, Unless…

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Central Message: We congratulate the authors for a successful one-year outcome in a patient with right atrial isomerism, anomalous pulmonary venous connection, and ventriculo-arterial discordance.

Central Figure Legend: Ryan G. McQueen, BS and Ronald K. Woods, MD, PhD

Commentary

Right atrial isomerism, total anomalous pulmonary veins, and ventriculo-arterial discordance – not a simple hand to be dealt. Careful judgment and execution ultimately led to a biventricular circulation with a systemic morphologic left ventricle with encouraging results at one year of follow-up.¹ Since this is so far a success, we wholeheartedly congratulate the authors.

We would typically not argue with success. But we have to write something, otherwise our commentary is done. So, let’s consider an alternative approach in management from the outset. In so doing, we trust the authors and readership will understand we do this with no intent of criticizing the authors’ decisions and actions.

At the outset we know there are two good ventricles, two good atrioventricular valves, and two good semilunar valves. There is also a good pulmonary venous confluence and additional extension of pulmonary venous tissue heading up to the left superior vena cava (SVC) – enough pulmonary venous confluence positioned nicely behind the left sided atrium (at least by computed tomography). Let’s accept the premise (although some may not), that regardless of
whatever else we do, we will initially leave the atrium widely un-septated. What follows,
however, is based on the presumption that an anatomic repair in this particular case is preferable
to physiologic repair, a presumption with which some may disagree.

So, as an alternative approach, what about doing the arterial switch concomitant with the veins
repair? We acknowledge this prolongs the surgery; however, in a straightforward manner the
heart can be reperfused after the veins repair before another period of cardioplegia, which seems
to be better tolerated than one prolonged ischemic period. The payoff is avoiding the very tricky
calibration of the pulmonary band, not to mention the potential impact of the band on subsequent
neo-aortic valve function. It also permits the coronary mobilization and transfer to be done in a
non-redo operative field.

Let’s assume this all goes well (not a trivial assumption). We have the luxury of waiting as there
is no threat of outgrowing a band – simply an atrial septal defect and mixing physiology. And in
either overall approach, we are intensely hoping the pulmonary vein repair remains durable. It’s
entirely unclear to us what happened to the right superior vena cava, but it’s gone. So, whenever
we declare the time right, we bring the patient back for atrial septation. While the current two-
patch baffle seems to be working well, we are mildly concerned by the cross-sectional imaging
appearance of the superior limb. A Glenn, perhaps with a left pulmonary artery band to the right
of the Glenn to reduce the pressure in the superior vena cava, would preclude the need for the
superior patch, but we acknowledge this is not a clearly superior alternative. No doubt the
authors will employ further follow-up imaging studies and manage their patient accordingly.
Unique cases such as this enhance both the challenge and fulfillment inherent to our specialty. We trust the authors agree with this sentiment. We congratulate them for thoughtful and carefully executed management.

**References**
