Commentary: Unifocalization revision: Patches, patches, and more patches...

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Central Message

An integrated approach is needed to manage recurrent branch pulmonary artery stenosis in patients with pulmonary atresia and major aortopulmonary collateral arteries after previous unifocalization.

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Pulmonary atresia with ventricular septal defect and major aortopulmonary collateral arteries represents a real challenge to the congenital cardiac surgical community. Unifocalization is the process of changing this abnormal multicompartamental pulmonary vascular bed into a normal single compartment through surgical reconstruction. The aim is to unite all these multiple different sources of pulmonary blood flow into a uniform compartment, with the goal of having the lowest possible pulmonary arterial (PA) to right ventricular (RV) pressure ratio to ensure satisfactory long-term outcomes. Several surgical strategies have been proposed during the last 2 decades, ranging from staged repair¹ to single-stage midline repair, as proposed by Hanley and colleagues.² The debate has continued between PA branch rehabilitation³ and unifocalization. The fate of this unifocalized vascular bed is not clearly known; however, reoperations are the rule, and it is well-known that these beds require repeat interventions, whether by repeat operations or by cardiac catheterization with ballooning or stenting.

In their article in this issue of the *Journal*, Mainwaring and colleagues⁴ review 254 patients who underwent single-stage midline complete unifocalization and described their experience with reoperation for recurrent branch stenoses that required revision (48 patients; 18%). Follow-up data were available for 85% of these patients, but there were no long-term data. This is another important addition from a leading group to the treatment of this challenging anomaly, and Mainwaring and colleagues⁴ are to be congratulated on their excellent outcomes.

In my mind, the initial unifocalization procedure is the most important one, because it sets up the path for what is happening next. The future of the patient's pulmonary vascular bed will depend on how it is managed in the first operation. We can see that with proper initial repair, most of these patients will do well. But we also have to realize that some of these patients will never reach complete repair. This group is luckily the smallest, and the question is how to identify this group early on to avoid unnecessary

interventions. There is no doubt that unifocalization is a complex and long operation, and it is not without postoperative morbidities. Several dilemmas remain:

1. How do we identify those patients who are at risk for recurrent branch PA stenosis? These patients will need a different follow-up protocol. As an example, those who initially had unifocalization to a shunt appear to have shorter interval before the development of branch PA stenosis.
2. Is unifocalization of major aortopulmonary collateral arteries needed in all patients? Clearly, some patients will never have complete repair despite extensive unifocalization, and if we can identify this subset early on, an alternate strategy such as PA branch rehabilitation with a shunt or RV-PA conduit will perhaps be enough.
3. For recurrent branch stenoses, when do we pull the trigger for percutaneous interventions with balloon dilation or stenting? In the study of Mainwaring and colleagues,⁴ 25% of patients who underwent revision had only mild elevation in their ratio of PA pressure to aortic pressure. Although the lower the right ventricular pressure, the better the long-term outcomes, repeat operation is not without risks.

In conclusion, pulmonary atresia with ventricular septal defect and major aortopulmonary collateral arteries requires

an integrated approach with proper initial unifocalization as the first step toward the formation of a healthy pulmonary vascular bed. Closer follow-up is mandatory, and a combination of interventional catheterization and repeat revision will be needed by most of these patients.

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