A Lesson About Pulmonary Artery Development From a Developing World Study

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In the current issue of the *Seminars*, Patel et al from India report outcomes in 25 patients with ventricular septal defect and pulmonary atresia (VSD/PA) who underwent valveless right ventricle to pulmonary artery (RV-PA) shunt as initial palliative surgery.[1] There was 1 operative and 1 late death. Among the remaining 23 patients, 17 achieved full repair while 6 were awaiting full repair. The distinctive aspect of this series is that the mean age of patients was 12.3 ± 3.2 years, much older than VSD/PA patients in the North America or Europe who typically receive surgery during infancy. The most remarkable finding of this series however was the fact that at a mean interstage follow-up of 8.3 ± 3.7 months, there was evidence of significant and symmetrical growth of the pulmonary arteries with an increase in Nakata index from 66 ± 24 to 186 ± 58 mm^2/m^2 and McGoon ratio from 0.9 ± 0.2 to 1.8 ± 0.4.

While the outcomes that the authors present are good, the complexity of this heterogeneous congenital cardiac defect makes it difficult to conclude a take home message from their experience. The patients presented in this study are obviously naturally selected and have survived to a mean age of 12 years without intervention. Early natural history studies have shown that 40% of VSD/PA patients die within a year from birth without intervention, and 60% die by 10 years of age, with endocarditis and pulmonary hemorrhage being major causes of mortality.[2,3] The risk of dying without intervention is inherently higher in patients without aortopulmonary collaterals (ductal-dependent) and is approximately 90% at 1 year from birth.[2,3] Therefore, the good results presented in this series from India cannot be used as an argument to delay surgery in those patients, and an intervention (repair or palliation) is expected to be necessary during early infancy. Another question is the optimal management of children with VSD/PA and some issues continue to be debated such as primary repair vs palliation in neonates with ductal-dependent VSD/PA, single-stage vs staged approach in children with aortopulmonary collaterals, and the management of those collaterals (unifocalization vs ligation).[4–6] The current series cannot address those controversies although it is very interesting to see that despite patients' old age (suggesting the presence of aortopulmonary collaterals), only 4 eventually underwent unifocalization.

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Central Message

Evidence of significant and symmetrical pulmonary artery growth following palliation of older patients with pulmonary atresia and ventricular septal defect using valveless RV-PA conduit stimulates reassessment of various management algorithms in patients having stalled progress due to limited pulmonary vascular bed.

The one interesting outcome in this report is the finding that there was a significant and symmetrical increase in the size of the pulmonary arteries at mean duration of 8.3 ± 3.7 months following initial placement of the valveless RV-PA shunt. This is in contrary to the common understanding that the greatest growth potential of pulmonary arteries only occurs during early years of life; a concept that has played an important role in the creation of many of our management strategies of various congenital heart defects. Not infrequently, we encounter in our practice patients who are failing their biventricular repairs or single-ventricle palliation surgeries due to small pulmonary arteries. The notion of conceivable rehabilitation of the pulmonary arteries at later age should stimulate physicians to explore

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approaches that might revert the patient to a shunt status with the hope to recuperate a better pulmonary vascular bed that would allow eventually a more successful repair or single-ventricle palliation. The regular and uniform growth in those older patients undergoing valveless RV-PA shunt is also a little dissimilar from the experience with aortopulmonary or RV-PA shunts in neonates and infants where distortion and disproportionate growth of the pulmonary arteries is not an uncommon encounter.[7] One possible explanation of the improved symmetry of pulmonary artery growth in the current series is the absence of compression by adjacent structures, for example, by the over-corrected neoaorta in patients undergoing Norwood operation. Another explanation might be the possibility that those older patients might be more resilient to develop pulmonary artery stretching, compression, or distortion by the shunt, heart, or aorta; consequently allowing constant and even growth of the pulmonary artery branches. This finding might therefore reaffirm that technical and mechanical factors play an important role in the evolution of pulmonary arteries following surgical intervention (shunt or repair) and should motivate surgeons to try to achieve flawlessness at time of initial surgery, and that can be accomplished with meticulous surgical technique, thoughtful choice of procedure (eg, length and type of shunt, shunt position in relation to the aorta, spatial relationship between the great vessels during neonatal reconstructions), and might be facilitated by preoperative planning (eg, virtual surgery and 3D reconstruction) and immediate postoperative confirmation of surgical adequacy (eg, intraoperative angiogram or magnetic resonance imaging).

The surgeons are always in a quest to improve outcomes, and that is not only limited to early survival but also to the performance of the optimal procedure that would provide the best long-term survival and the optimal functional status of their patients. While the findings from the study from India do not necessarily apply to the majority of patients with similar cardiac defects treated in developing countries, it does challenge our understanding of conceivable pulmonary artery growth in older children and that should kindle our curiosity to reassess our management strategies in patients with stalled progress following surgical repair or palliation due to hypoplastic and obstructed pulmonary arteries.

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