Commentary: Is there more than one way to skin a pulmonary artery aneurysm?

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Lung transplantation (LTX) has become an acceptable therapy for patients with end-stage lung disease. In this issue of the Journal, Schwarz and colleagues2 from the Vienna Lung Transplant Group report excellent outcomes using a technical solution in a rare subset of patients requiring LTX for pulmonary artery (PA) hypertension (PAH). Seven of 128 patients who underwent LTX for PAH had a pulmonary artery aneurysm (PAA), involving the main PA and often extending into the left and right PAs. This rare complication of PAH poses substantial problems for LTX and leads to the decision to perform heart-lung transplantation in some centers. We assume that these are patients with primary pulmonary hypertension. If so, this is a considerable and unique single-center experience in a country with a population of 9 million. In comparison, only 930 LTX procedures for primary PAH were performed in the United States (population >300 million) during the same time period (Figure 1).2 This discrepancy may be due to lung allocation policy differences and the ability of the Vienna group to choose recipients for donor lungs.

Their clever solution to the technical challenge of a PAA may also be feasible because of allocation policy in a country with a single LTX center. Their well described and illustrated technique involves using the entire PA of the donor when the heart was not recovered. Interestingly, these PAAs were not associated with pulmonary valve regurgitation and so did not involve the valve annulus. There was no association between the ascending thoracic aortic diameter and the presence of a PAA, implying that the pathogenesis of PAAs in the setting of PAH may differ.

Figure 1. Number of lung transplants (LTX) for primary pulmonary hypertension (PPH) in the United States for the same time period as in the study reported by Schwarz and colleagues (1996-2018). The increase in 2017 likely reflects US allocation policy changes for patients with pulmonary artery hypertension, and that in 2018 likely reflects the wider geographic distribution of donor lungs outside of the local donor service area to more patients with higher lung allocation scores.3
from that for ascending thoracic aortic aneurysms, which often involve the annulus and cause aortic insufficiency.

Given that the ascending aorta and PA develop from a common truncus arteriosus, it stands to reason that the tendency for the PA to dilate is translatable to the ascending aorta as well. Much of the enthusiasm for performance of the Ross procedure in adults with bicuspid aortic valve disease waned as a result of this finding in pulmonary autografts translocated to the aortic position. The patients in this group all have the equivalent of PAH. Recent basic science studies have shown that the resident cells in the ascending aorta (and hence likely in the PA as well) have mechanoreceptor “set points” that cause activation of cellular signaling and up-regulation of protease (in particular matrix metalloproteinase) production causing unopposed proteolysis and weakening of the arterial wall. Despite this, although hypertension is a risk factor for aortic aneurysm formation, most hypertensive patients never develop aortic aneurysms, indicating that other mechanisms are involved that require further investigation.

Schwarz and colleagues describe an elegant technique that works very well in this rare situation. When the entire PA is not available, Force and coworkers described another solution that used the donor thoracic aortic arch and descending thoracic aorta to replace the recipient PAA (Figure 2). Both techniques should be in the LTX surgeon’s toolbox to address this rare complication of PAH.

References