Unrestrictive Aortopulmonary Window
Extreme Presentation as Non-Eisenmenger in a 30-Year-Old Patient

Patrick O. Myers, MD; Frédéric Lador, MD, PhD; Anne-Lise Hachulla, MD; Judith Bouchardy, MD; Stéphane Noble, MD; Marc Licker, MD; Jean-Claude Pache, MD; Dimitra Kalimanovaska-Ostric, MD; Milan Djkuc, MD; Afksendiyos Kalangos, MD, PhD; Maurice Beghetti, MD

A 30-year-old woman was referred for management of an aortopulmonary window. Her history was notable for chronic atrial fibrillation treated by amiodarone and Coumadin anticoagulation. Peripheral oxygen saturation was 95%. A chest x-ray film showed cardiomegaly, bilateral prominent hilar vascularization, and interstitial edema (Figure 1). The echocardiogram showed a large, 15-mm aortopulmonary (AP) window, ≈25 mm above the aortic valve annulus between the ascending aorta and main pulmonary artery (type I) with an unrestrictive, exclusively left-right shunt and a peak gradient of 30 mm Hg (Figure 2). There was no aortic coarctation or interruption of the aortic arch, and the main pulmonary artery (3.3 cm) was dilated to a size equal to the aortic root, with dilated branch pulmonary arteries (2.3 cm each). The left atrium was severely dilated (45 cm<sup>2</sup>), as was the left ventricle (end-diastolic diameter, 7.5 cm; indexed, 5.2 cm/m<sup>2</sup>). There was moderate mitral regurgitation (regurgitation volume, 26.6 mL; proximal isovelocity surface area convergence radius, 6.8 mm) attributable to annular dilatation. The biventricular systolic function was normal. The right ventricular systolic pressure was estimated at 63.3 mm Hg by the gradient of the tricuspid regurgitation jet. Cardiovascular MRI confirmed these findings (Figure 3). Cardiac catheterization showed a pulmonary systolic pressure 40% of systemic pressure (44/20 mm Hg; mean, 32), pulmonary capillary wedge pressure of 20 mm Hg, cardiac index of 3.04 L·min<sup>−1</sup>·m<sup>−2</sup>, pulmonary vascular resistance (PVR) of 1.79 Woods units (WU)/m<sup>2</sup>, and PVR:systemic vascular resistance (SVR) ratio of 0.21. After acute vasodilator testing with inhaled nitric oxide, the PVR decreased to 1.54 WU/m<sup>2</sup> with a PVR:SVR ratio of 0.12. Following pulmonary vasodilation with inhaled nitric oxide, the PVR decreased to 2.89 WU/m<sup>2</sup> with a PVR:SVR ratio of 0.12 and pulmonary capillary wedge pressure of 14 mm Hg (Figure 4). The patient was placed on sildenafil 20 mg 3 times per day. The 6-minute walk test showed a distance 64% of predicted, with a transcutaneous saturation decreasing from 98% to 91%. The lung biopsy (Figure 5) showed muscularization of the media of the intralobar and centrolobar arteries, with no intimal reaction, compatible with Heath and Edwards grade I pulmonary hypertension.1

At follow-up 13 months after repair and after discontinuing sildenafil, the patient denied any symptoms and had resumed full-time work. Her echocardiogram showed regression of left ventricular dilatation (end-diastolic diameter, 5.9 cm), mild systolic left ventricular dysfunction (ejection fraction, 50%), moderate mitral regurgitation, and estimated right ventricular systolic pressure of 35 mm Hg. Cardiac catheterization showed a pulmonary artery pressure of 31/8 mm Hg (mean, 18 mm Hg), a mean pulmonary capillary wedge pressure of 8 mm Hg, pulmonary vascular resistance of 1.6 WU/m<sup>2</sup>, with a PVR:SVR ratio of 0.03.

An AP window, a failure of septation between the ascending aorta and the pulmonary artery during fetal life because of nonfusion of the opposing conotruncal ridges...
responsible for separating the truncus arteriosus into the aorta and pulmonary artery, is a rare congenital cardiac anomaly. Patients usually present in the first week of life, because the lesion is usually large and nonrestrictive. Late or extremely late presentation is unusual, but has been reported in adults at the stage of cyanosis and Eisenmenger syndrome. We report the rare presentation of an AP window with an unrestrictive left-right shunt in a patient at 30 years of age, still at the stage of reactive pulmonary hypertension and who underwent successful surgical repair. Despite this positive short-term outcome, with an excellent functional result and normalization of hemodynamics just beyond 1 year from surgery, the long-term outcome remains to be seen. Not all patients follow the predicted development of irreversible pulmonary vascular lesion. This patient is of particular interest because AP windows are thought to develop rapid pulmonary vascular disease. Understanding why some patients seem to be protected from the development of pulmonary vascular disease would be of extreme importance for the treatment of pulmonary hypertension.

Disclosures
Dr. Beghetti has served as consultant, steering committee and/or advisory board member for Actelion Pharmaceuticals Ltd., Bayer-Healthcare, Eli Lilly, GlaxoSmithKline, and Pfizer. The other authors report no conflicts.

References

Figure 1. Preoperative anteroposterior chest x-ray.
Figure 2. Preoperative echocardiogram. **A**, Four-chamber view showing left atrial and ventricular dilatation and moderate mitral regurgitation. **B**, Long-axis view at the level of the aortopulmonary window (*).
Figure 4. Vasodilator testing 20 days after surgical repair. Pulmonary artery pressure at baseline (A) and after inhaled nitric oxide administration (B).

Figure 5. Lung biopsy histopathology. A small pulmonary artery demonstrates hypertrophy of the media (hematoxylin and eosin stain, original magnification ×400).
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