Coronary–Cameral and Coronary Arteriovenous Fistulae in a Transplanted Heart

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A 32-year-old man underwent routine elective left heart catheterization 1 year after orthotopic cardiac transplantation for advanced congestive cardiac failure secondary to familial dilated cardiomyopathy. At the time of transplantation, the donor heart appeared normal macroscopically. During the following year, a total of 12 protocol endomyocardial biopsies were performed, all of which were negative for acute rejection. On the patient’s latest follow-up visit, his cardiac examination was normal and no murmur was present. An ECG demonstrated right bundle-branch block and left anterior hemiblock, both abnormalities being present since the time of transplantation.

As part of left heart catheterization, left ventriculography was performed, during which the pulmonary artery unexpectedly opacified with contrast (Figure I and Movie I in the online-only Data Supplement). Selective coronary angiography of his left main coronary artery revealed the presence of a coronary–cameral fistula between the fourth septal perforating branch of the left anterior descending artery and the right ventricle as well as a coronary arteriovenous fistula between the proximal left anterior descending artery and the main pulmonary artery (Figure II in the online-only Data Supplement). These fistulae were not the only cause of pulmonary artery opacification during ventriculography; selective coronary angiography of his right coronary artery revealed a third coronary arteriovenous fistula from the proximal right coronary artery to the main pulmonary artery (Figure III in the online-only Data Supplement). Echocardiography showed the presence of diastolic turbulent flow in the right ventricle originating from the interventricular septum on color Doppler (Figure I and Movie IV in the online-only Data Supplement), which was present even in studies performed before the patient’s first endomyocardial biopsy. Echocardiography also demonstrated normal systolic function of both the left and right ventricles (left ventricular ejection fraction, 78%; Figure II in the online-only Data Supplement). The left ventricular outflow tract velocity time integral was not elevated (Figure IV in the online-only Data Supplement). Calculation of the left-to-right shunt using echocardiographic parameters (left ventricular outflow tract diameter, 2.1 cm; left ventricular outflow tract velocity time integral, 17 cm; right ventricular outflow tract diameter, 2.4 cm; right ventricular outflow tract velocity time integral, 14.5 cm; Figure V in the online-only Data Supplement) found this shunt to be insignificant physiologically (QP:QS, 1.1:1). Similar QP:QS values were obtained during a nuclear medicine 99m-Tc-diethylene triamine pentaacetic acid shunt study (QP:QS, 1.18:1) and right heart catheterization (Fick pulmonary cardiac output, 5.4 L/min; Fick systemic cardiac output, 5.0 L/min; QP:QS, 1.08:1). Right heart catheterization also demonstrated no elevation in right-sided filling pressures (right atrium: 1/1 mm Hg; mean, 1 mm Hg; right ventricle: 19/1 mm Hg; mean, 4 mm Hg; pulmonary artery: 17/4 mm Hg; mean, 10 mm Hg; pulmonary capillary wedge, 12/4 mm Hg; mean, 9 mm Hg). Further investigation discovered that the cause of death of the donor was massive intracranial hemorrhage secondary to cerebral arteriovenous malformation.

Coronary artery fistulae occur as congenital anomalies in 0.2% of the general population, but are more common among recipients of orthotopic cardiac transplantation, in which the incidence is approximately 8%. This higher prevalence, particularly of fistulae that drain into the right ventricle, is thought to be related to repeated endomyocardial biopsies, which are usually taken with a biopette from the interventricular septal portion of the right ventricle. Another possible reason for the higher prevalence of fistulae among heart transplant recipients is that people with multiple arteriovenous malformations, particularly those with cerebral arteriovenous malformations, may be at increased risk of catastrophic neurological events and are therefore more likely to become organ donors. Iatrogenic injury from endomyocardial biopsy is a potential cause of this patient’s coronary–cameral fistula, particularly because it involves a perforating septal branch in the interventricular septum and drains into the right ventricle. However, the presence of 2 other coronary artery fistulae in the transplanted...

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The online-only Data Supplement is available with this article at http://circ.ahajournals.org/lookup/suppl/10.1161/CIRCULATIONAHA.112.109199/-/DC1.

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(Circulation. 2012;126:2018-2019.)

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Circulation is available at http://circ.ahajournals.org DOI: 10.1161/CIRCULATIONAHA.112.109199

2018
heart, multiple cerebral arteriovenous malformations in the donor, as well as the fact that the left anterior descending septal branch associated with the fistula is significantly greater in caliber than nearby septal branches are consistent with the fistula involving the septal perforator being congenital in origin. The presence of bifascicular block in the transplanted heart may also be a consequence of a congenital abnormality involving the interventricular septum.

Coronary artery fistulae are associated with complications such as myocardial ischemia secondary to a steal phenomenon, bacterial endocarditis, and sudden cardiac death, all of which may potentially reduce life expectancy. The presence of left-to-right shunting during diastole may potentially increase the risk of future right ventricular overload, pulmonary hypertension, and congestive cardiac failure. In patients with significant shunting, elective percutaneous coil or balloon embolization has been shown to be safe and effective treatment strategies. In our patient, echocardiography, right heart catheterization, and a nuclear medicine shunt study have proved the left-to-right shunt to be insignificant. From a functional viewpoint, this patient has experienced marked improvement after transplantation and has returned to full-time work. He is being monitored regularly for signs of pulmonary hypertension and has been advised to take antibiotic prophylaxis prior to surgical or dental procedures.

Disclosures

None.

References

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doi: 10.1161/CIRCULATIONAHA.112.109199
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/126/16/2018

Data Supplement (unedited) at:
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SUPPLEMENTAL MATERIAL
Supplemental Figure Legends

Supplemental Figure 1: Transthoracic echocardiography performed from a parasternal short axis transducer position, demonstrating turbulent flow into the right ventricle arising from the interventricular septum which is more marked during diastole (A) than systole (B).

Supplemental Figure 2: Calculation of the patient’s left ventricular ejection fraction.

Supplemental Figure 3: The velocity time integral recorded in the left ventricular outflow tract from an apical transducer position (pulse waveform).

Supplemental Figure 4: Calculation of the aortic valve mean gradient from an apical 5-chamber transducer position.

Supplemental Figure 5: The velocity time integral recorded in the right ventricular outflow tract from a parasternal short axis transducer position.
Supplemental Figure 1: Transthoracic echocardiography performed from a parasternal short axis transducer position, demonstrating turbulent flow into the right ventricle arising from the interventricular septum which is more marked during diastole (A) than systole (B). Although this flow is continuous, it is much more marked during diastole, which is consistent with the presence of a fistula between the coronary circulation and the right ventricle.
Supplemental Figure 2: Calculation of the patient’s left ventricular ejection fraction.
Supplemental Figure 3: The velocity time integral recorded in the left ventricular outflow tract from an apical transducer position (pulse waveform).
**Supplemental Figure 4:** Calculation of the aortic valve mean gradient from an apical 5-chamber transducer position.
Supplemental Figure 5: The velocity time integral recorded in the right ventricular outflow tract from a parasternal short axis transducer position.