Radionuclide Angiography in Congenitally Corrected Transposition of the Great Vessels in an Adult

Rami Gal and Steven C. Port

Nuclear Cardiology Laboratory, Mount Sinai Medical Center, University of Wisconsin Medical School, Milwaukee Clinical Campus, Milwaukee, Wisconsin

The case of a 34-yr-old man with congenitally corrected transposition of the great vessels is described. Both first-pass and equilibrium radionuclide angiographic data were acquired and demonstrate the value of both studies in delineating the ventricular inversion and the transposed great vessels that are characteristic of this disorder. In addition to the anatomic information, the ejection fractions of the venous and systemic ventricles at rest and during exercise, the lack of any left to right shunt, and the presence of systemic A-V valve insufficiency can all be obtained from the scintigraphic data.

J Nucl Med 28:116–118, 1987

CASE REPORT

A 34-yr-old man presented to the emergency department complaining of severe lightheadedness. He was known to have congenital heart disease since approximately the eighth grade. Cardiac catheterization performed in 1979 had demonstrated congenitally corrected transposition of the great vessels (CCTGV). He had been asymptomatic until 1981 when he first experienced dizziness and palpitations. In 1982, he had a syncopal episode. No anti-arrhythmic therapy was prescribed. Recently he had been having increasing episodes of dizziness and a "funny feeling" in the stomach and it was the latter that prompted his visit to the emergency department, where a wide-QRS tachycardia at 210–250 bpm was documented. Neither lidocaine nor verapamil altered the rhythm and rate, and he subsequently converted to sinus rhythm spontaneously. At that time, the physical examination revealed a regular heart rhythm at 70 and a blood pressure of 120/80. Jugular venous and carotid pulses were normal. On cardiac auscultation, there was a grade 3/6 holosystolic murmur heard widely over the precordial area. The remainder of the examination was unremarkable.

The electrocardiogram taken in sinus rhythm showed a normal PR interval. There were deep Q waves in leads III and AVF and deep S waves in V_{2-3}. T wave inversions were present in leads I, AVL, V_{5-6}. The chest x-ray showed borderline cardiac enlargement but was otherwise normal; however, the only chest film available was a bedside portable.

The patient underwent electrophysiologic testing which showed that his arrhythmia was atrial fibrillation with rapid A-V conduction and ventricular aberration. There was no evidence of an accessory pathway. Because of the murmur, he was felt to have systemic A-V valve insufficiency and he was referred to the nuclear cardiology laboratory for evaluation of his ventricular function at rest and during exercise.

The patient was injected with 5 mg of stannous pyrophosphate and 15 min later, 25 mCi technetium-99m (^{99m}Tc) pertechnetate were injected during which a first-pass radionuclide angiogram (FPRNA) was acquired.* Subsequently, a three view, resting, equilibrium radionuclide angiogram (EQRNA) was acquired followed by an EQRNA† during supine exercise.

Figure 1 shows serial 1-sec images of the FPRNA acquired in the anterior projection in the patient with CCTGV. One can see that, during the "right" ventricular phase (series A), the venous ventricle has the typical morphology of a left ventricle. In addition, the great vessel arising from the venous ventricle courses more to the right than a typical pulmonary artery. During the systemic ventricular phase, the ventricle has a large triangular shape that is more typical of a morphologic right ventricle (series B). The great vessel that arises from the systemic ventricle is clearly displaced to the left compared with the normal situation. A time-activity curve from a region of interest around the systemic ventricle (Figure 2) shows a prolonged transit time in the systemic ventricle due to valvular insufficiency. The latter is consistent with either semilunar or A-V valve insufficiency. In this patient, atrial transit time was also prolonged supporting A-V valve insufficiency as the cause.

Figure 3 shows the end-diastolic and end-systolic frames of the resting EQRNA in the patient with CCTGV and in a normal patient. Optimum ventricular separation was found in a 25° left anterior oblique (LAO) projection and was never

Received Jan. 23, 1986; revision accepted July 17, 1986.

For reprints contact: Steven C. Port, MD, Mount Sinai Medical Center, P.O. Box 342, Milwaukee, WI 53201.

as good as obtained in typical normal patients. The morphology of the ventricles is not as obviously inverted as is seen in the first-pass study. Very striking, however, is the abnormal orientation of the great vessels (compare with the normal patient in Figure 3). In the CCTGV patient, the great vessels take an almost straight cephalad course. Note the leftward displacement of the aorta (solid arrows) and the rightward displacement of the pulmonary artery (open arrows) in the anterior view. In the LAO view, the great vessels do not cross each other as is seen in the normal situation where the ascending aorta courses anteriorly and to the right and the main pulmonary artery courses posteriorly and to the left.

Neither the images nor the lung curves indicated a shunt in this patient. The ejection fraction of the venous ventricle was 0.58 at rest as determined from the FPRNA, and 0.61 at rest using the EQRNA method (two regions of interest). The resting ejection fraction of the systemic ventricle was 0.43 on the FPRNA and 0.40 on the EQRNA. At peak exercise the EQRNA ejection fractions of the venous ventricle increased from 0.61 to 0.63 while the ejection fraction of the systemic ventricle decreased from 0.40 to 0.36.

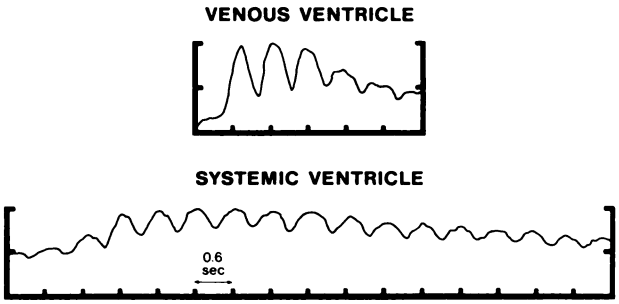

FIGURE 2
Time-activity curves from separate regions of interest for the venous and systemic ventricles. Note the normal transit time in the venous ventricle and the markedly prolonged transit time through the systemic ventricle. The latter is consistent with A-V valve insufficiency.

DISCUSSION

Congenitally corrected transposition of the great vessels has been clearly described in the angiographic (*1,2*), echocardiographic (*3,4*), and pathologic literature (*5,6*). However, descriptions of the entity in the scintigraphic literature are scarce. Hagan et al. (*7*) reported first-pass scintigraphic data from five subjects with CCTGV but four had shunts which precluded an accurate description of the ventricles and the one without a shunt was only 11 mo old. Attie et al. (*8*) presented FPRNA data from seven children with CCTGV in the anterior and left lateral views. Guit et al. (*9*) presented FPRNA data from one adult with CCTGV while Brendel et al. (*10*) showed both FPRNA and EQRNA images of a 5½ yr old with CCTGV. To our knowledge our patient is the first reported adult case with both FPRNA and EQRNA data available. Both techniques yielded valuable information. The first-pass study very clearly demonstrated the ventricular inversion. The left ventricular morphology of the venous ventricle and right ventricular morphology of the systemic ventricle were easily appreciated. In addition, the prolonged tracer transit time in the systemic ventricle suggested valvular insufficiency. The prolonged atrial transit time and the fact that patients with CCTGV often have systemic A-V valve insufficiency support the conclusion that the prolonged LV transit time was due to A-V valve rather than semilunar valve insufficiency. Although the abnormal positions of the great vessels could be appreciated on the FPRNA, the transposition of the great vessels was more strikingly apparent on the EQRNA when both great vessels were simultaneously imaged. The use of a very shallow left anterior oblique or straight anterior-

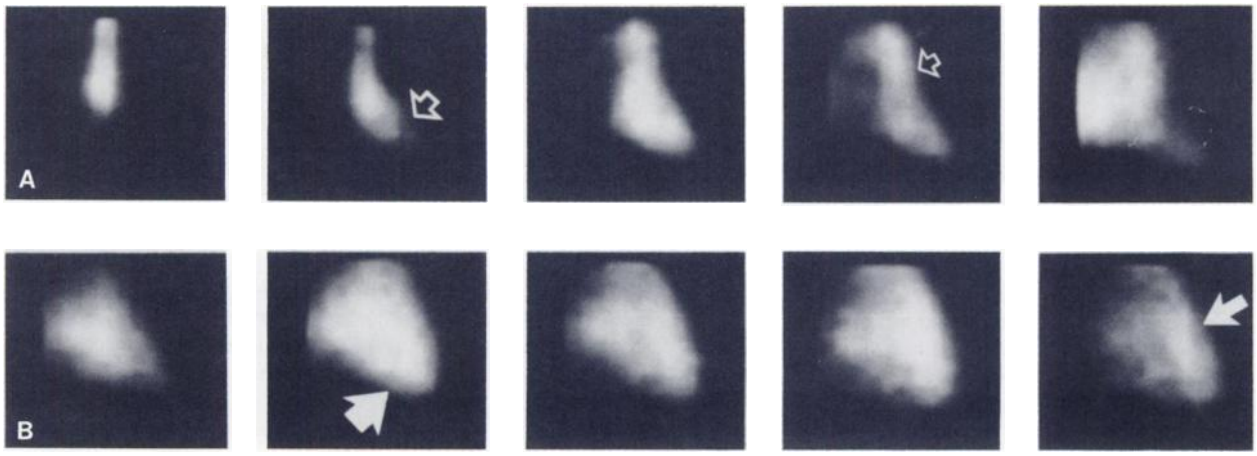

FIGURE 1
Serial 1-sec images from the first-pass study of the patient with CCTGV. Series A shows the radionuclide bolus entering from the superior vena cava into the venous ventricle (large open arrow) and then into the pulmonary artery (small open arrow). Series B shows the activity in the systemic ventricle (large closed arrow) and in the aorta (small closed arrow). See text for detailed explanation.

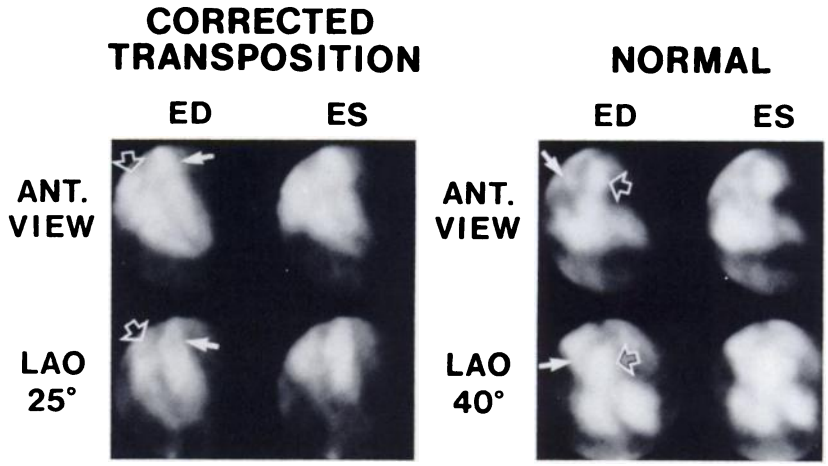

FIGURE 3
End-diastolic and end-systolic images from the anterior and LAO acquisitions in the patient with CCTGV and in a normal patient for comparison. Solid arrows indicate the aorta and open arrows the pulmonary artery in both patients. See text for explanation.

posterior projection for equilibrium imaging in patients with CCTGV has been emphasized previously by Parrish et al. (*11*) and was helpful in delineating the anomalous position of the great vessels and in separating the systemic and venous ventricles for calculation of ejection fractions in our patient. Similar to the findings of Parrish et al., the ejection fraction of the venous ventricle was normal at rest and during exercise in our patient. The ejection fraction of the systemic ventricle was mildly reduced at rest and decreased during exercise. Parrish et al. also reported a drop in ejection fraction during exercise for the systemic ventricle. The older age and chronic systemic A-V valve insufficiency may have contributed to the abnormal resting ejection fraction of the systemic ventricle in our patient.

In summary, both first-pass and equilibrium radionuclide angiography can be of value in identifying the rare congenitally corrected transposition of the great vessels. The combination of the two techniques seems to provide all the anatomic information necessary for the diagnosis. Detection of shunts or valvular insufficiency can also be accomplished and longitudinal follow-up of systemic ventricular function at rest and during exercise can be readily performed.

NOTES

* Baird Corporation, (Scinticor), Bedford, MA.
† Elscint Inc., (Apex 215), Boston, MA.

REFERENCES

1. Graham TP, Parrish MD, Boucek RJ, et al. Assessment of ventricular size and function in congenitally corrected transposition of the great arteries. *Am J Cardiol* 1983; 51:244–251.
2. Freedom RM, Harrington DP, White RI Jr. The differential diagnosis of levo-transposed or malposed aorta: an angiocardiographic study. *Circulation* 1974; 50:1040–1046.
3. Feigenbaum H. Echocardiography. 3rd ed. Philadelphia: Lea & Febiger, 1981:387–388.
4. Hagler DJ, Tajik AJ, Seward JB, et al. Atrioventricular and ventriculoarterial discordance (corrected transposition of the great arteries): wide-angle two-dimensional echocardiographic assessment of ventricular morphology. *Mayo Clin Proc* 1981; 56:591–600.
5. Allwork SP, Bentall HH, Becker AE, et al. Congenitally corrected transposition of the great arteries: morphologic study of 32 cases. *Am J Cardiol* 1976; 38:910–923.
6. VanPraagh R, Durnin RE, Jockin H, et al. Anatomically corrected malposition of the great arteries [S, D, L]. *Circulation* 1975; 51:20–31.
7. Hagan AD, Friedman WF, Ashburn WL, et al. Further applications of scintillation scanning technics to the diagnosis and management of infants and children with congenital heart disease. *Circulation* 1972; 45:858–868.
8. Attie F, Cuaron A, Marquez M, et al. Qualitative radionuclide angiocardiography in the diagnosis of corrected transposition. *AHJ* 1979; 97:141–146.
9. Guit GL, Kroon HM, Chin JG, et al. Radionuclide angiocardiography in the clinical evaluation of cardiac malpositions in situs solitus in adults. *J Nucl Med* 1986; 27(4):484–490.
10. Brendel AJ, Wynchank S, Choussat A, et al. Radionuclide angiocardiography in the diagnosis of congenitally corrected transposition of the great arteries. *Clin Nucl Med* 1984; 9(7):383–388.
11. Parrish MD, Graham TP, Bender HW, et al. Radionuclide angiographic evaluation of right and left ventricular function during exercise after repair of transposition of the great arteries: Comparison with normal subjects and patients with congenitally corrected transposition. *Circulation* 1983; 67:178–183.