

Scintigraphic imaging of a case of congenitally corrected transposition of the great vessels and an adult case of single atrium and single ventricle

Hitoko OGATA,* Tomoaki NAKATA,* Akita ENDOH,* Kazufumi TSUCHIHASHI,
Shuji YONEKURA,* Shigemichi TANAKA,* Takatoshi TSUDA,**
Masahiro KUBOTA,** and Osamu IIMURA

**The Second Department of Internal Medicine, and*

***Department of Radiology, Sapporo Medical College*

We report on the clinical utility of radionuclide angiography and gated blood pool single emission computed tomography (gated blood pool SPECT) in two patients having congenital heart disease. Both conventional equilibrium radionuclide angiography and gated blood pool SPECT demonstrated the connection of the great vessels with both ventricles in a 15-year-old male patient with a congenitally corrected transposition of the great vessels. In particular, the latter procedure could provide very useful information about the ventricular morphology and inversion which is important for diagnosing this disorder. The second case is an extremely rare 42-year-old female patient with a single atrium and single ventricle. She underwent first-pass and multiple gated blood pool angiography from the anterior, right and left oblique views. The combination of these scintigraphic techniques revealed an insufficiency in anatomical correlations among the single atrium, atrioventricular valve, single ventricle and the great vessels in addition to the connection of superior vena cava with the single atrium, and the atrioventricular valve. Thus, conventional equilibrated angiography from multiple views and gated blood pool SPECT seems to be very reliable not only for anatomical evaluation but also for clinical course observation in patients with complicated congenital heart disease.

Key words: Congenitally corrected transposition of great vessels, Single atrium and single ventricle, Radionuclide angiography, Gated blood pool tomography

INTRODUCTION

RADIONUCLIDE angiography seems an established method to diagnose various cardiac disorders and to evaluate cardiac function. However, there are few reports about scintigraphic images of congenital heart disease. Although congenitally corrected transposition of the great vessels was assessed by means of radionuclide techniques by some investigators,¹⁻⁵ there has not been a blood pool tomographic study

of this disorder. Furthermore, the scintigraphic imaging of single atrium and single ventricle, which is an unusual cardiac disease, is rarely found in adults. We encountered two adult patients with these cardiac malformations the scintigraphic analysis of which was very useful.

Case 1:

A 15-year-old male was admitted to our hospital for further examination of a heart murmur and electrocardiographic abnormalities. He had no symptoms and his physical development was found to be normal. However, he had developed a cardiac murmur at three years of age. On admission, physical examinations revealed a regular heart rhythm of 78/min and a blood pressure of 108/48 mmHg. A grade 3/6

Received April 8, 1988; revision accepted August 31, 1988.

For reprint contact: Hitoko Ogata, Second Department of Internal Medicine, Sapporo Medical College, S 1, W 16, Chio-ku, Sapporo 060, JAPAN.

systolic murmur was best heard at the left basilar area and the second heart sound was accentuated with normal respiratory splitting. The electrocardiogram showed a first-degree atrioventricular block, inverted T wave in leads I and aVL and the absence of Q wave in the left precordial leads. The chest X-ray showed that the right cardiac contour was enlarged. Two-dimensional echocardiography suggested that the aorta was the anterior leftward great artery which arose from the left ventricle with right ventricular morphology. The pulmonary artery was posterior and rightward to the aorta (Fig. 1). Radionuclide cardioangiography was carried out with 20 mCi of ^{99m}Tc red blood cells. Equilibrated radionuclide angiography was obtained from the left anterior oblique view with a gamma camera (LFOV, Seale). It showed that the aorta originated above the left-side ventricle which was morphologically suspected to be the right ventricle (Figure 2, top panel). ECG-gated data were acquired continually with a parallel-hole collimator. They were obtained for 60 seconds in every 10° from the left posterior oblique view to the right anterior oblique view by the 10 frame multigate method. The data were then filtered and back projected. Finally each view was obtained. The gated blood pool tomograms from the vertical long axis

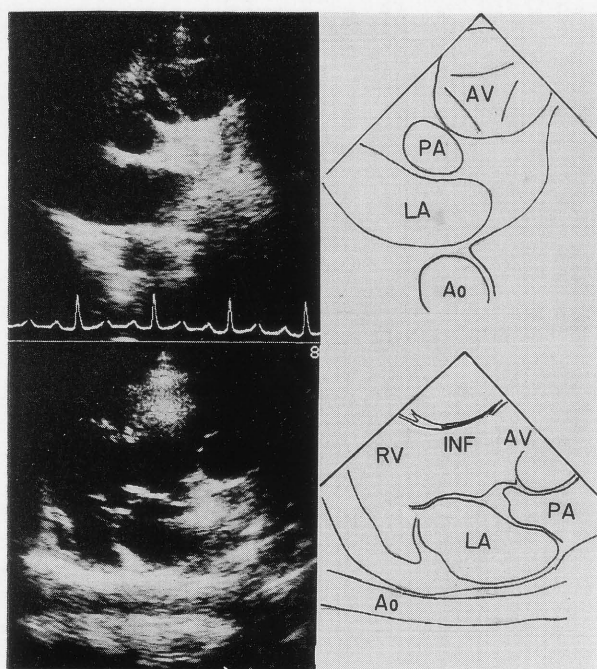


Fig. 1 Two-dimensional echocardiograms of case 1. Parasternal short axis (top panel) and long axis (bottom panel) images show the relation between the great vessels and cardiac chambers. The aortic valve (AV) is anterior and leftward to the pulmonary artery (PA). The aorta (AO) arises from the infundibulum (INF) of the left-side ventricle of the morphology of the right ventricle (RV). The ventricle is connected with the left atrium (LA).

more clearly demonstrated that the right-side ventricle had a left ventricular morphology and the left-side ventricle was anatomically a right ventricle with an infundibulum (Fig. 2, middle and bottom panels). No right-to-left shunt was shown by first-pass angiography. These results suggested that his cardiac malformation was congenitally corrected transposition of the great vessels (S.L.L.T.). The diagnosis was established morphologically by aorto- and ventriculography with contrast material. The results of the hemodynamic study revealed that the pressure of each chamber and artery was within the normal range. The end diastolic pressure of the left-side ventricle was 2 mmHg and that of the right side ventricle was 0 mmHg.

Case 2:

A 42-year-old female patient was admitted to our hospital because of severe cyanosis, dyspnea, and orthopnea. She had often suffered from these symptoms since childhood. The diagnosis of a single

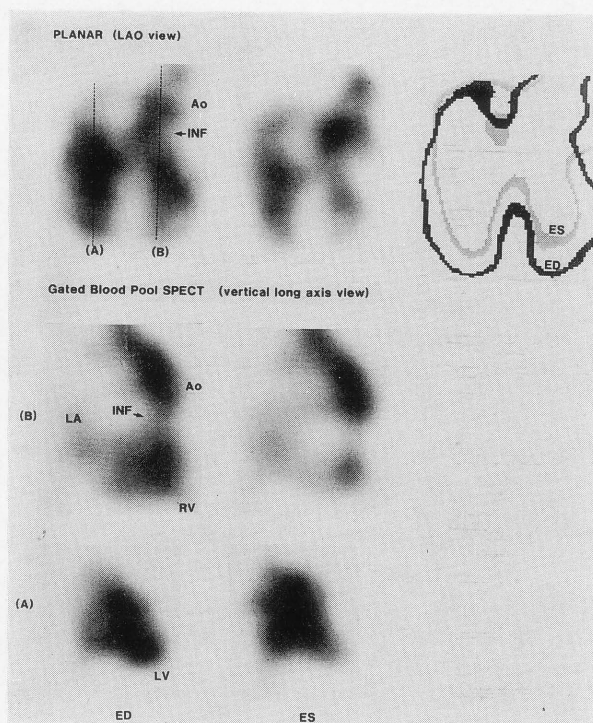


Fig. 2 Gated pool radionuclide angiograms of case 1. The left anterior oblique images indicates an abnormal connection of the great vessels with a ventricle (top panel). The vertical long axial tomograms are obtained by gated blood pool SPECT. The tomogram of the left-side ventricle sliced by the line "B" clearly shows that the aorta (AO) arises from the infundibulum (INF) of the ventricle which is, therefore, morphologically a right ventricle (RV). The tomogram of the right-side ventricle sliced by line "A" demonstrates that the shape and performance of the ventricle are those of the left ventricle (LV).

atrium and single ventricle was made when the second surgical repair was attempted sixteen years after the first surgical procedure which was a Blalock-Taussig's shunt operation on the diagnosis of Tetralogy of Fallot at the age of 14. After that, despite conservative therapy she gradually became worse. On admission, her exercise tolerance was a grade 4/4 of the NYHA classification. The physical examinations revealed a regular heart rhythm of 90/min, blood pressure of 132/76 mmHg and respiratory rate of 17/min with severe cyanosis and clubbed fingers. There was severe hepato-splenomegaly. The jugular vein was markedly dilated. A grade 5/6 harsh systolic murmur was audible at the right basilar area and a grade 3/6 holosystolic murmur was present at the right mammary area. The second heart sound was single and the third heart sound was heard. The chest X-ray showed dextrocardia and was compatible with severe pulmonary hypertension (Fig. 3). The electrocardiogram revealed an incomplete left bundle branch block and T wave inversion in leads V5R and V6R. The laboratory data showed marked hypoxemia, polycythemia and renal dysfunction.

Radionuclide cardiography was carried out with 20 mCi of ^{99m}Tc red blood cells. First-pass angiograms from the anterior projection demonstrated a connection of the superior vena cava with the left-side cardiac chamber and the left-to-right shunt (Fig. 4). In addition, the retention of radioactive blood pool seemed to show the presence of valvular insufficiency. Gated blood pool scintigraphy was performed from three different angles (Fig. 5). The wall motion analysis revealed that the left-side chamber had the atrial performance and the right-side chamber showed a ventricular contraction during the systole. Although the two chambers seemed

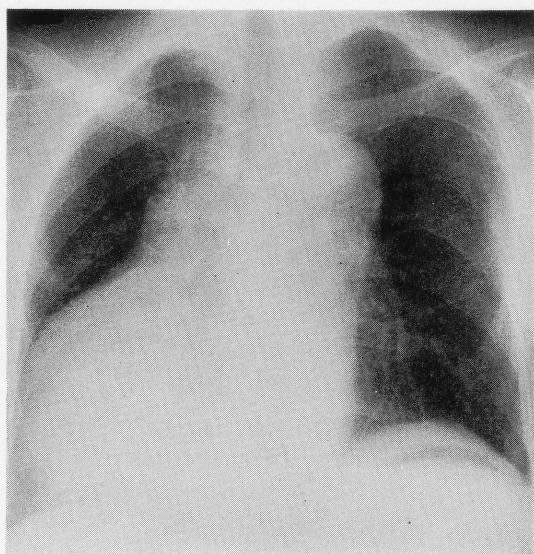


Fig. 3 The chest X-ray of isolated dextrocardia of case 2.



Fig. 4 Serial first-pass angiograms per second from the anterior projection of case 2. The retention of a blood pool in the left-side chamber which communicates with the superior vena cava shows the presence of valvular insufficiency. Early visualization of the aorta suggests the left-to-right shunt.

to be well separated, no other cardiac cavities were detected by any imagings. The pulmonary trunk was visualized at the back of the aorta. These scintigraphic findings suggested that this patient had a single atrium and single ventricle, where the great vessels originated, and atrio-ventricular valve insufficiency.

The diagnosis was confirmed by contrast angiography (Fig. 6). The single ventricle had well developed trabeculae. The aorta arose in front of the pulmonary artery with a subaortic conus. The pulmonary valve was markedly stenotic. In spite of the recovery from congestive heart failure, she suddenly died. An autopsy supported these findings and revealed that the walls of the single ventricle were markedly hypertrophied with well developed trabeculae but no residual septum. The atrioventricular valve was tricuspidal and insufficient. The Blalock-Taussig's shunt was occluded and pulmonary stenosis was evident.

DISCUSSION

In almost all cases of congenitally corrected transposition of the great vessels (CCTGV), operative repair is not necessary. Its diagnosis and clinical course observation is very important because of the possibility of advanced A-V blocks. Several studies of scintigraphic imaging of CCTGV have been reported.¹⁻⁵ Recently, however, Gal R. et al. (3)

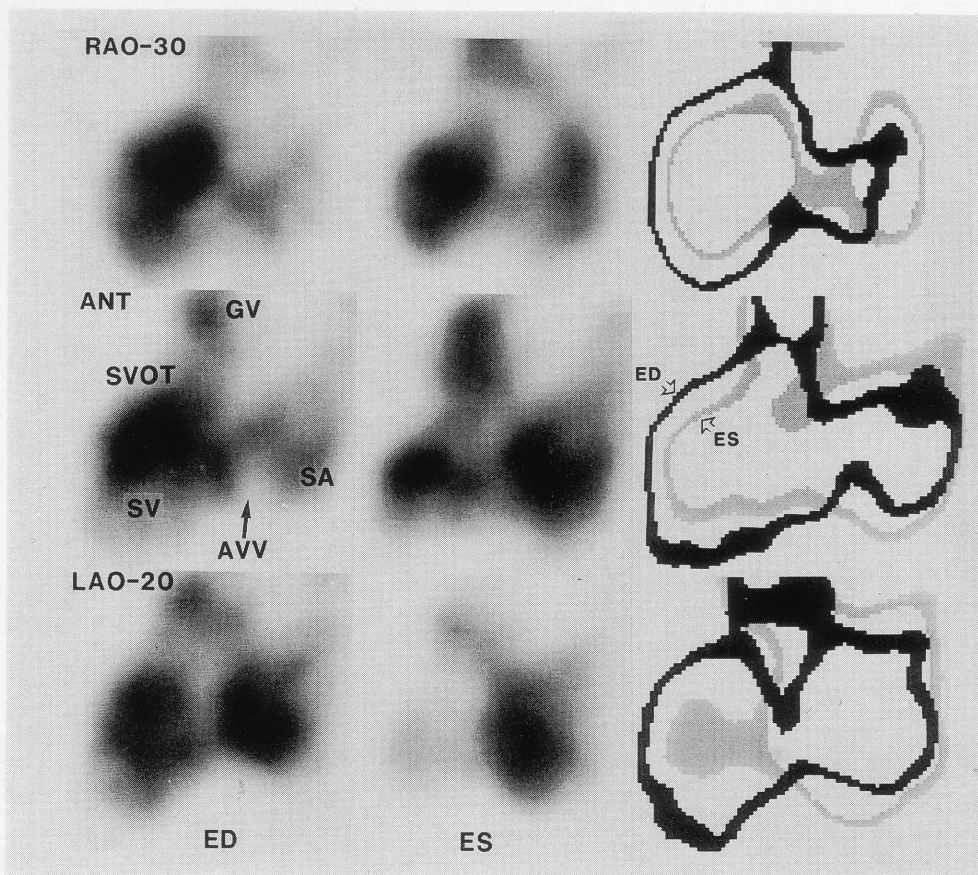


Fig. 5 Gated blood pool images obtained from RAO 30 (top), anterior (middle) and LAO 20 (bottom) in case 2. Only two chambers and the great vessel(s) can be detected. The wall motion analysis demonstrates that the left chamber is an atrium and the right one is a ventricle where the great vessels arise. Accordingly, atrioventricular valve insufficiency is suspected. GV=Great vessels, SV=single ventricle, SA=single atrium, SVOT=single ventricle outflow tract.

showed the usefulness of first-pass and equilibrated radionuclide angiography in CCTGV. The conventional planar angiography does not provide all the precise anatomical information including the morphological assessment of the ventricles. On this point, gated blood pool single photon emission computed tomography (gated blood pool SPECT) appears to be useful in obtaining an accurate delineation of the cardiovascular system and separating the other blood pool from the cardiac chambers.⁶⁻⁸ However, there have been no scintigraphic reports of CCTGV revealed by this particular method. Our study shows that gated blood pool SPECT can contribute to the detailed observation of the morphology of the right and left ventricles and, therefore, is very useful for diagnosing CCTGV.

We experienced a very rare adult case with a single atrium and single ventricle.⁹⁻¹² No scintigraphic assessment of single atrium and single ventricle (SA-SV) has been reported. Two-dimensional echo-

cardiographic and scintigraphic examinations were very useful in diagnosing it, in particular under the condition of congestive heart failure. First-pass angiography revealed dextrocardia, correlation of the superior vena cava with the single atrium and A-V insufficiency. Gated blood pool angiography from several angles could more clearly demonstrate the anatomical connection of the single atrium, single ventricle and great vessels. The wall motion analysis contributed to the differentiation of an atrium from a ventricle. These scintigraphic observations were compatible with angiographic findings with contrast material and confirmed by autopsy. From these clinical data, we speculate that she had been able to survive till the age of 42 because of pulmonary stenosis and the marked hypertrophy of the single ventricle which was a morphologically the right ventricle despite occlusion of the Blalock-Taussig's shunt. Concerning the long-term compensation of this single ventricle, it cannot always be

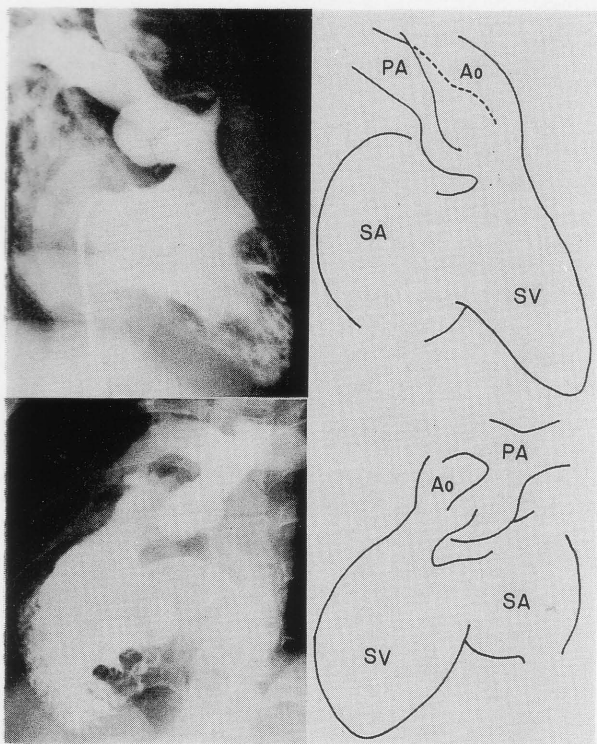


Fig. 6 Contrast angiograms of case 2. Both the RAO (top) and LAO (bottom) images reveal the infundibulum and well developed trabeculae of the ventricle where the great vessels originate. Furthermore, the aorta is anterior and leftward to the pulmonary artery of the stenotic valve. PA=pulmonary artery, Ao=aorta, SA=single atrium, SV=single ventricle.

said that its origin is the right ventricle even if its morphology is like that.

In summary, radionuclide angiography can be performed and repeated in patients with severe clinical symptoms without any deleterious side effects. The combination of the first-pass technique, equilibrium radionuclide angiography and gated blood pool SPECT seems to be clinically very useful in diagnosing complicated congenital heart disease and estimating its clinical condition.

REFERENCES

1. Attie F, Cuaron A, Marquez M, et al: Quantitative radionuclide angiography in the diagnosis of corrected transposition. *AHJ* 97: 141-146, 1979
2. Brendil AJ, Wynchank S, Choussat A, et al: Radionuclide angiography in the diagnosis of congenitally corrected transposition of the great arteries. *Clin Nucl Med* 9: 383-388, 1984
3. Gal R, Port SC: Radionuclide angiography in congenitally corrected transposition of the great vessels in an adult. *J Nucl Med* 28: 116-118, 1987
4. Hagan AD, Friedman WF, Ashburn WL, et al: Further application of scintillation scanning techniques to the diagnosis and management of infants and children with congenital heart disease. *Circulation* 45: 858-868, 1972
5. Guit GL, Kroon HM, Chin JG, et al: Radionuclide angiography in the clinical evaluation of cardiac malformations in situs solitus in adults. *J Nucl Med* 27: 484-490, 1986
6. Moore MD, Murphy PH, Burdine JA: ECG-gated emission computed tomography of the cardiac blood pool. *Radiology* 134: 233-235, 1980
7. Barat JL, Brendel AJ, Colle JP, et al: Quantitative analysis of left ventricular function using gated single photon emission tomography. *J Nucl Med* 25: 1167-1174, 1984
8. Nakata T, Murakami H, Inoue M, et al: Qualitative determination of infarct segment by Fourier analysis using gated cardiac pool emission computed tomography. *J Cardiatr* 16: 873-884, 1986 (in Japanese)
9. Goldberg HL, Sniderman K, Devereux R: Prolonged survival (62 years) with single ventricle. *Am J Cardiol* 52: 214-215, 1983
10. Elliott LP, Bream PK, Gessner IH: Single and common ventricle, In *Heart disease in Infants, Children and Adolescents*, 2nd ed, Moss AJ, Adams FH, Emmanouilides GC (ed), Baltimore, The Williams & Wilkins Co., 1978, pp 381-393
11. Moodie DS, Ritter DG, Tajik AJ: Long-term follow-up in the unoperated univentricular heart. *Am J Cardiol* 53: 1124-1128, 1984
12. Feigenbaum H: Single ventricle, In *Echocardiography*, 4th ed, Philadelphia, Lea & Febiger, 1986, pp 430-437