Right Aortic Arch: Demonstration by First-Pass and Multigated Radionuclide Ventriculography

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Right aortic arch is a rare congenital anomaly associated with abnormal development of the paired embryological aortic arches. While various abnormalities of the great vessels have been described using both first-pass and multigated radionuclide ventriculographic studies, diagnosis of a right-sided aortic arch has typically required a radiographic contrast technique. We present a case of a patient with a suspected right-sided aortic arch diagnosed by radionuclide methods.

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Right aortic arch is a rare congenital anomaly associated with abnormal development of the embryological aortic arches (*1*). Radiographic features are well described, with the diagnosis often being made from a standard postero-anterior (PA) and lateral chest film (*2*). In these cases, the aortic knob protrudes from the right side of the mediastinum, the trachea is deviated to the left, and the descending aorta is seen along the right side of the spine. Opacification of the esophagus with swallowed barium often demonstrates an indentation produced by the right-sided aorta (*2*). In order to differentiate the various types of great vessel anomalies, contrast angiography and/or computed tomography (CT) are often necessary (*3,4*). Digital subtraction angiography (DSA) has also been shown to be useful (*5*) and the efficacy of echocardiography (*6*) and magnetic resonance imaging (MRI) (*7*) have been more recently described.

Radionuclide ventriculography employing both first-pass and multigated methods has most commonly been used to assess ventricular function, chamber size, and the presence of circulatory shunts. Various abnormalities of the great vessels have been visualized by radionuclide methods (*9–13*). We report a case of an adult patient in whom radionuclide ventriculography was used to confirm the presence of a suspected right-sided aortic arch.

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CASE REPORT

A 44-yr-old male was referred for cardiac catheterization to evaluate intermittent chest pain after a stress test was suggestive of exercise-induced ischemia. Physical exam was completely normal. Resting electrocardiography revealed an intraventricular conduction delay with nonspecific T-wave abnormalities. A chest radiograph from the referring hospital was reported to be normal except for a calcified granuloma in the left upper lobe.

The patient underwent cardiac catheterization via the right femoral artery. Abnormal passage of the catheter along the right side of the thoracic spine was noted, raising the possibility of a right-sided aortic arch. Coronary angiography was normal. In order to minimize the amount of dye used, left ventriculography and aortography were not performed. Following the procedure, a chest radiograph revealed an abnormal mediastinal structure (Fig. 1), which was felt to be either a mass or a right-sided aortic arch. Radionuclide ventriculography was performed to further clarify the patient's aortic anatomy.

Using an in vivo red blood cell labeling technique, a first-pass angiocardiographic study was performed with a 300-mm field of view gamma camera (with a low-energy all purpose collimator) in the 30° right anterior oblique projection (RAO). Images were acquired in a 64 × 64 matrix at one frame per second. Standard multigated radionuclide blood-pool ventriculography (MUGA) was then performed using the same camera and collimator with 16 frames per R-R interval acquired in a 64 × 64 matrix. In addition to standard anterior, left anterior oblique, and left posterior oblique views, imaging was performed in the 30° RAO projection. Both the first-pass study and resting MUGA confirmed the right-sided aortic arch with otherwise normal cardiac anatomy and function (Figs. 2 and 3).

DISCUSSION

There are two types of right-sided aortic arch, type I and type II, each resulting from abnormalities in the complex embryologic development of the aorta. At an early fetal stage (prior to the 30-mm stage) there are six paired aortic arches (*1*). The first three arches are involved in formation of the carotid arteries. Normally the ventral portion of the right fourth arch forms the right subclavian and innominate arteries, while the dorsal portion disappears. The left-sided fourth arch persists in its entirety to form the adult aortic arch. If the dorsal portion of the left fourth aortic arch disappears (rather than the right), a right-sided aortic arch is formed which is a mirror image of the usual left-

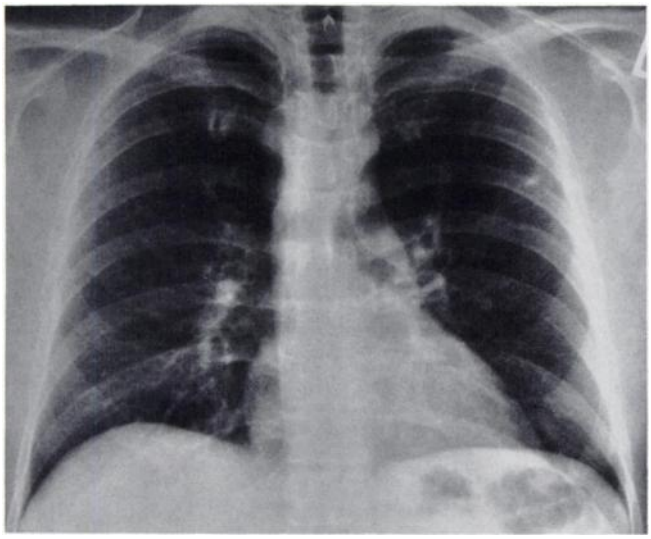

FIGURE 1. PA chest radiograph demonstrating protrusion of the aortic knob from the right side of the upper mediastinum along with leftward tracheal deviation. The descending aorta is seen along the right side of the thoracic spine.

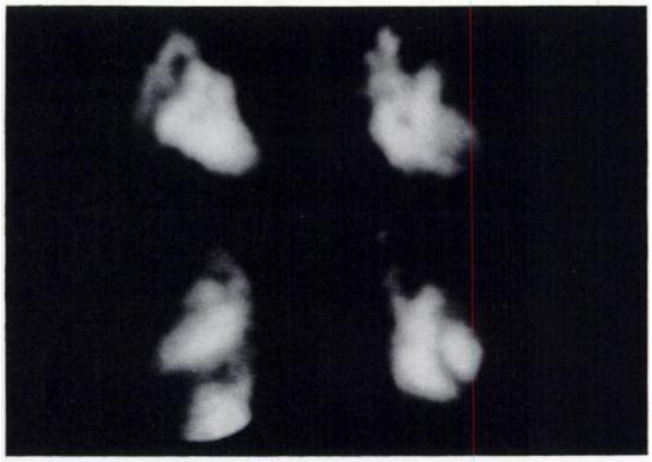

FIGURE 3. End-diastolic frames of the multigated blood-pool ventriculogram. Clockwise from the left upper corner, these images represent 30° RAO, anterior, maximal-separation LAO, and LPO views, respectively. The abnormal right-sided aortic arch is best seen in the RAO projection. The other three views demonstrate absence of the normal left-sided aortic arch and descending aorta.

sided arch. This is referred to as a type I or "mirror image" right aortic arch. In 90% of cases, this type I right-sided arch is associated with other cardiac anomalies, most commonly Tetralogy of Fallot (*1*,*8*).

A type II right aortic arch is formed when the dorsal portions remain intact, but a break occurs in the ventral portion of the embryological left fourth arch (*1*). In this condition, there is no innominate artery and the left subclavian artery arises from a diverticulum extending behind the esophagus off the distal right aortic arch. The type II arch is the one seen most commonly in adults and is not associated with an increased incidence of congenital cardiac anomalies (*2*). While the two types appear the same on standard chest radiographs, differentiation is possible when a barium-filled esophagus is seen to bow anteriorly due to pressure from the diverticulum of a type II right-sided arch (*1*,*2*). It should be noted that in virtually all cases of right-sided arch the descending thoracic aorta crosses back to the left side and enters the diaphragm at its normal position (*2*). The level at which the aorta crosses over to the left side is variable and has no diagnostic significance.

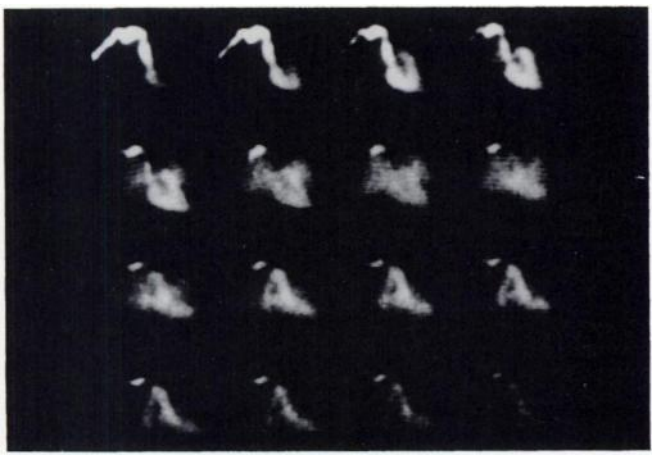

FIGURE 2. First-pass angiocardiographic study displayed at 1 sec per frame was acquired with the gamma camera positioned in a 30° RAO projection. The bolus of [^{99m}Tc]pertechnetate is noted to pass normally from the right subclavian vein and superior vena cava into the right-sided cardiac chambers. After normal pulmonary circulation, the left ventricle is visualized. Radiotracer is then noted to exit the left ventricle through an abnormal right-sided aortic arch. There was no visual evidence of abnormal left-to-right or right-to-left shunt, and the great vessels appear otherwise normal.

We can only conjecture as to the type of right aortic arch that this patient had. There was no evidence of an associated cardiac anomaly. This apparently healthy young male is thus most likely to have had a type II right-sided arch, with a posterior diverticulum giving rise to the left subclavian artery.

Although not typically used to assess the great vessels, radionuclide ventriculography and radionuclide angiography have been used to visualize various anomalies in development of the great arteries. Kriss et al. reviewed the findings in first-pass studies of Tetralogy of Fallot, pulmonic stenosis, and other congenital cardiac disorders (*9*, *10*). Wesselhoeft et al. described the radionuclide angiocardiographic appearance of pulmonary atresia and truncus arteriosus (*11*) and Long et al. demonstrated a case of hemitruncus (*12*).

In adult patients, acquired abnormalities of the proximal aorta are more commonly encountered than congenital anomalies and many such aberrations have been described by radionuclide methods. Sussman et al. reviewed several examples of abnormalities of the ascending aorta detected

on multigated blood-pool images, including aortic root dilatation as well as an ascending aortic aneurysm (*13*).

Although radionuclide ventriculography is not the optimal procedure for precise anatomical definition, this technique may be quite helpful in patients unable to tolerate standard intravenous radiographic contrast agents (thus precluding the use of contrast angiography or CT), those who cannot be placed in an MRI unit, or those in whom optimal echocardiography cannot be performed. Evaluation of the great vessels is an important part of cardiac blood-pool imaging. Congenital anomalies of the heart and great vessels should be considered when unusual shapes and/or structures are seen on radionuclide studies performed for other purposes.

REFERENCES

1. Felson B, Palayew MJ. The two types of right aortic arch. *Radiology* 1963;81:745–759.
2. Baron MG. Right aortic arch. *Circulation* 1971;44:1137–1145.
3. Backer CL, Ilbawi MN, Idriss FS, DeLeon SY. Vascular anomalies causing tracheoesophageal compression: review of experience in children. *J Thoracic Cardiovasc Surg* 1989;97:725–731.
4. Vallette RC, Arensman RM, Falterman KW, Ochsner JL. Tracheoesophageal compression syndromes related to vascular ring. *South Med J* 1989;82:338–340.
5. Otero-Cagide M, Moodie DS, Sterba R, Gill CC. Digital subtraction angiography in the diagnosis of vascular rings. *Am Heart J* 1986;112:1304–1308.
6. Kveselis DA, Snider AR, Dick M, Rocchini AP. Echocardiographic diagnosis of right aortic arch with a retroesophageal segment and left descending aorta. *Am J Cardiol* 1986;57:1198–1199.
7. Kersting-Sommerhoff BA, Sechtem UP, Fisher MR, Higgins CB. MR imaging of congenital anomalies of the aortic arch. *Am J Roentgenol* 1987;149:9–13.
8. Stewart JR, Kincaid OW, Titus JL. Right aortic arch: plain film diagnosis and significance. *Am J Roentgenol* 1966;97:377.
9. Kriss JP, Enright LP, Hayden WG, Wexler L, Shumway NE. Radioisotopic angiocardiography: wide scope of applicability in diagnosis and evaluation of therapy in diseases of the heart and great vessels. *Circulation* 1971;43:792–808.
10. Kriss JP, Enright LP, Hayden WG, Wexler L, Shumway NE. Radioisotopic angiocardiography: findings in congenital heart disease. *J Nucl Med.* 1971;13:31–40.
11. Wesselhoeft H, Hurley PJ, Wagner HN. Nuclear angiocardiography in the differential diagnosis of congenital heart disease in infants [Abstract]. *J Nucl Med* 1971;12:406.
12. Long WA, Perry R, Henry GW. Radionuclide diagnosis of anomalous origin of the right pulmonary artery from the ascending aorta (so-called hemitruncus). *Int J Cardiol* 1985;8:492–496.
13. Sussman MH, Horowitz SF, Navarro JL, Machac J. Abnormalities of the ascending aorta detected by gated blood-pool imaging. *Clin Nucl Med* 1986;11:198–202.