A Novel Case of L-transposition with a Right-dominant Double Aortic Arch

David J. Goldberg, MD, Jenifer Glatz, MD, Paul M. Weinberg, MD, and Matthew J. Gillespie, MD

The Children's Hospital of Philadelphia, Division of Cardiology, Philadelphia, Penn, USA

ABSTRACT

We describe a case of congenitally corrected transposition with a double aortic arch. This unique combination of lesions highlights the importance of a complete anatomic assessment prior to referral for surgery.

Key Words. Transposition; Vascular Ring

Congenitally corrected transposition of the great arteries, the most common of which is transposition of the great arteries {S, L, L},* also known as L-transposition (L-TGA), is a rare congenital anomaly making up <1% of all cases of congenital heart disease.[1] The major associated cardiac defects include ventricular septal defect, pulmonary outflow tract obstruction, and tricuspid valve dysplasia, including Ebstein anomaly.[1–7] Coarctation of the aorta and interruption of the aortic arch have also been found.[4–6] We report a case of congenitally corrected transposition with a ventricular septal defect (VSD) and a right-dominant double aortic arch.

Case Presentation

A four-day-old Hispanic male was referred to the Cardiology Clinic at The Children's Hospital of Philadelphia for evaluation of a cardiac murmur. He was born at full term via caesarian section done for breech presentation and a history of a previous caesarian section. The pregnancy was complicated by gestational diabetes that was controlled by diet alteration and insulin therapy. During his nursery stay, he was noted to have a cardiac murmur. He was in no distress and had normal pulse oximetry, so he was referred for an outpatient exam on the day of discharge. Aside from his cardiac murmur, he had an uneventful nursery course.

On arrival to cardiology clinic, physical exam revealed a well-appearing, well-developed newborn male in no distress. His length was 60 cm and his weight was 4.3 kg. His pulse rate was 142 and his respiratory rate was 80. The blood pressure in his right arm was 63/41, with no gradient to his lower extremities. The oxygen saturation was 95% in his right great toe. His lungs were clear to auscultation. He had a normal S_1 and a single S_2. He had a low-pitched, grade 2/6 systolic murmur along the sternal border. His abdomen was soft with no organomegaly.

The infant had an ECG performed at the time of his visit that showed normal sinus rhythm with voltage criteria for biventricular hypertrophy. There was no q-wave in V1 and there was a small q-wave in V6. Unsedated echocardiography was performed that revealed the diagnosis of transposition of the great arteries {S, L, L} with a possible double aortic arch (Figures 1 and 2).

The infant was referred for cardiac catheterization to assess his suitability for a double switch (a Senning procedure and an arterial switch operation[8]). In the cath lab, the diagnosis of congenitally corrected transposition with a ventricular septal

*This means situs solitus or normal arrangement of the atria; ventricular L-loop, sometimes called ventricular inversion, where the morphological right ventricle (RV) is left-sided and the left ventricle (LV) is right-sided; and L-position of the great arteries, where the aorta arises from the morphological RV to the left of the pulmonary artery, which arises from the LV. Another form of congenitally corrected transposition is situs inversus with D-loop and D-position of the great arteries {I, D, D}. In both, aside from associated anomalies, systemic venous blood goes to the pulmonary artery and pulmonary venous blood to the aorta, hence corrected physiology despite anatomical transposition.

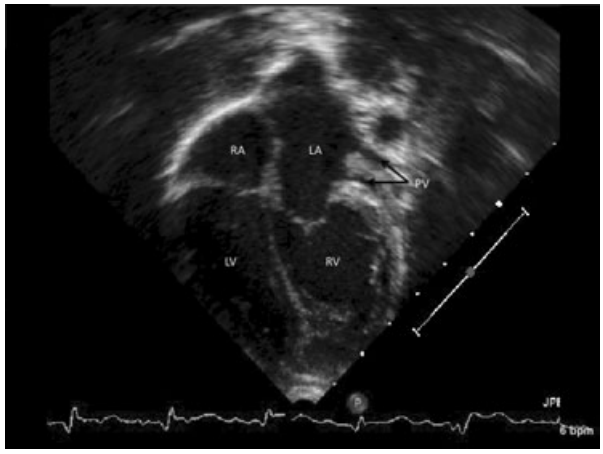

Figure 1. A two-dimensional echocardiographic image demonstrating congenitally corrected transposition {S, L, L}. The septal surface of the right-sided ventricle (LV) is smooth, consistent with a morphologic left ventricle. The left-sided ventricle (RV) is heavily trabeculated and appears to have chordal attachments to the septal surface, consistent with a morphologic right ventricle. Pulmonary veins (PV) can be seen entering the left-sided atrium (LA), consistent with a morphologic left atrium.

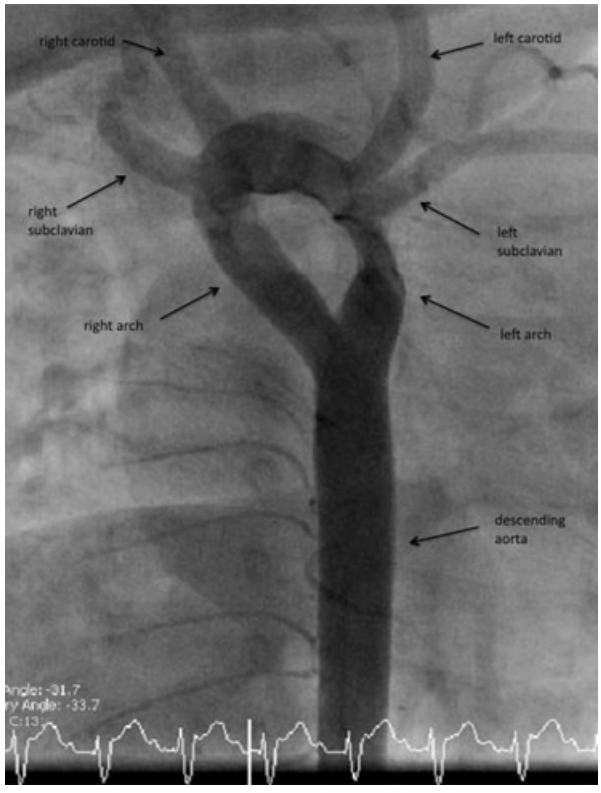

Figure 2. An angiogram with right anterior oblique and caudal angulation demonstrating a right-dominant double aortic arch. The right subclavian and the right carotid come off of the right arch while the left subclavian and the left carotid come off the left arch. The ascending aorta is not well opacified in this projection.

defect and a right-dominant double aortic arch was confirmed. Hemodynamic evaluation suggested suitability for a double switch, and he was referred for cardiovascular surgery. A double switch procedure with a VSD closure and division of a double aortic arch was performed at 3 months of age. The procedure was complicated by transient third-degree heart block that required temporary pacing.

On most recent follow-up, the child, now 2 years old, had normal growth and development and no symptoms related to his cardiovascular system. He was not on any cardiac medications. He was in normal sinus rhythm though his ECG did demonstrate a left bundle branch block.

Discussion

While intracardiac anomalies are frequently associated with congenitally corrected transposition, associated anomalies of the great vessels are considerably less frequent. In a review of 121 patients with L-TGA and 2 good-sized ventricles, Rutledge found that only 9 (7.4%) had no associated anomalies. Of the remainder with associated anomalies, only 16 had aortic arch involvement.[5] All of those patients had obstructive abnormalities (e.g., coarctation or interruption of the aortic arch) and none had a double aortic arch. Anderson and Schiebler reported smaller series; and both found that the majority of patients had associated intracardiac anomalies but very few had anomalies of the great vessels.[3,6]

Unlike L-TGA, double aortic arch is often found in isolation, most often with right-sided dominance. In a review of vascular rings, Backer found 7% prevalence of cardiac lesions associated with double aortic arch.[9] Alsenaidi reported a prevalence of 18% in his review.[10] Both numbers are significantly lower than the frequency of cardiac defects found in association with congenitally corrected transposition.

While aortic arch anomalies have previously been described in patients with transposition of the great arteries,[11,12] we believe that this is the first description of congenitally corrected transposition of the great vessels with a double aortic arch. It is unknown whether this combination of lesions is related to fetal hemodynamics or whether there is an underlying genetic abnormality that predisposes to this novel anatomy. In this case, the patient had a double switch procedure as well as division of his double aortic arch and is currently doing well and symptom free. Our case highlights

the importance of adequate imaging to assess the entirety of a patient's anatomy.

Corresponding Author: David J. Goldberg, MD, Division of Cardiology, The Children's Hospital of Philadelphia, 34th and Civic Center Boulevard, Philadelphia, PA, 19104, USA. Tel: (+1) 267-426-3058; Fax: (+1) 215-590-5825; E-mail: goldbergda@email.chop.edu

Conflict of interest: None.

Accepted in final form: August 6, 2008.

References

1 Freedom RM. Conditions with double discordance (congenitally corrected transposition of the great arteries). In: Freedom R, ed. *The Natural and Modified History of Congenital Heart Disease*. Elmsford, NY: Blackwell Publishing, Inc. / Futura Division; 2004:356–365.
2 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.
3 Anderson RC, Lillehei CW, Lester RG. Corrected transposition of the great vessels of the heart: a review of 17 cases. *Pediatrics*. 1957;20:626–646.
4 Lundstrom U, Bull C, Wyse RK, Somerville J. The natural and "unnatural" history of congenitally corrected transposition. *Am J Cardiol*. 1990;65: 1222–1229.
5 Rutledge JM, Nihill MR, Fraser CD, Smith OE, McMahon CJ, Bezold LI. Outcome of 121 patients with congenitally corrected transposition of the great arteries. *Pediatr Cardiol*. 2002;23:137–145.
6 Schiebler GL, Edwards JE, Burchell HB, Dushane JW, Ongley PA, Wood ED. Congenital corrected transposition of the great vessels: a study of 33 cases. *Pediatrics*. 1961;27(suppl 5):849–888.
7 Van Praagh R, Papagiannis J, Grunenfelder J, Bartram U, Martanovic P. Pathologic anatomy of corrected transposition of the great arteries: medical and surgical implications. *Am Heart J*. 1998;135 (5, pt 1):772–785.
8 Yagihara T, Kishimoto H, Isobe F. Double switch operation in cardiac anomalies with atrioventricular and ventriculoarterial discordance. *J Thorac Cardiovasc Surg*. 1994;107:351–358.
9 Backer CL, Mavroudis C, Rigsby CK, Holinger LD. Trends in vascular ring surgery. *J Thorac Cardiovasc Surg*. 2005;129:1339–1347.
10 Alsenaidi K, Gurofsky R, Karamlou T, Williams WG, McCrindle BW. Management and outcomes of double aortic arch in 81 patients. *Pediatrics*. 2006; 118:e1336–e1341.
11 Feingold B, O'Sullivan B, del Nido P, Pollack P. Situs inversus totalis and corrected transposition of the great arteries [I,D,D] in association with a previously unreported vascular ring. *Pediatr Cardiol*. 2001;22:338–342.
12 Higashino SM, Ruttenberg HD. Double aortic arch associated with complete transposition of the great vessels. *Br Heart J*. 1968;30:579–581.