

A TYPICAL CASE OF L-TRANSPOSITION OF THE GREAT ARTERIES INITIALLY PRESENTED AS COMPLETE ATRIOVENTRICULAR BLOCK IN MIDDLE-AGED MAN

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L-transposition of the great arteries (L-TGA) is a rare congenital anomaly and could cause complete atrioventricular (AV) block at relatively younger age. We present a case of 43-year-old male who complained of dizziness due to complete AV block. We confirmed L-TGA using transthoracic echocardiography and cardiac computed tomography. Permanent pacemaker was inserted without complications. No invasive treatment including corrective surgery was performed because patient's cardiac function was almost normal and the symptom was completely resolved after pacemaker insertion.

KEY WORDS: Congenital heart disease · L-transposition of the great arteries · Complete AV block.

INTRODUCTION

L-transposition of the great arteries (L-TGA) is a rare congenital anomaly representing approximately 0.5% of all congenital heart disease. In L-TGA, deoxygenated systemic blood flow from correctly located right atrium to discordant left ventricle [LV, functional right ventricle (RV)] through mitral valve (MV) and pumped to the discordant pulmonary artery (pulmonary circulation). Oxygenated blood from pulmonary veins flow from correctly located left atrium to discordant RV (functional LV) through tricuspid valve and pumped to discordant aorta (systemic circulation).¹⁾ It has various terms including congenitally or physiologically corrected transposition of the great arteries, L-transposition, double discordance [atrioventricular (AV), ventriculoarterial], ventricular inversion.²⁾ In patients with L-TGA, AV node and His bundle have an unusual position and course. Therefore, bundle is vulnerable to fibrosis with aging. This causes dysfunction of conduction system of L-TGA patients. Therefore L-TGA patients frequently have complete AV block, approximately 2% rate per year and recurrent reentry tachycardia.^{3,4)} Here, we report the typical case of L-TGA which was initially presented as

complete AV block in adult.

CASE

A 43-year-old male was transferred to our hospital with complaints of dizziness, dyspnea-on-exertion, and mild chest pain for 2 weeks. He had been diagnosed hypertension two years ago, but he did not take antihypertensives for two months. Otherwise he had no medical or surgical history. He smoked 2 packs per day for 20 years. He denied any previous history of syncope, palpitation and peripheral edema except dyspnea-on-exertion for 2 weeks. He also complained intermittent dizziness which was not related with specific situation or position. On admission, his blood pressure was 156/80 mm Hg and grade 2 systolic murmur was audible in pulmonic area. Electrocardiogram showed complete AV block with severe bradycardia, 45 beats/min. Complete AV block was confirmed by Holter monitoring (Fig. 1). Thyroid function test was normal (thyroid stimulating hormone 1.24 uIU/mL and free T4 1.21 ng/dL) and coronary angiography was followed to rule out coronary artery disease. There was no significant stenosis in left coronary arteries. Right coronary artery could not be examined

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due to engagement failure. Right-sided aortic arch was suspected during coronary angiography (Fig. 2). Thus, we had implanted a permanent pacemaker (Fig. 3). Complete AV block at relatively younger age, engagement failure of right coronary artery, right-sided aortic arch in coronary angiography could suggest congenital heart disease. Transthoracic echocardiography revealed coarse trabeculations of functional LV and fine trabeculations and smooth surfaced functional RV, which correspond with L-TGA. Left-sided systemic AV valve was displaced inferiorly closer to the cardiac apex than right-sided AV valve, which is consistent with tricuspid and mitral valve respectively. Estimated ejection fraction was 57%. In color Doppler study, there was no significant valvular dysfunction in right-sided mitral valve, aortic valve, and pulmonary valve except mild left-sided tricuspid regurgitation. Systolic murmur in pulmonic area could be explained by left-sided grade 2 tricuspid regurgitation (Fig. 4). Following cardiac computed tomography showed congenitally corrected transposition of great arteries with AV, ventriculoarterial discordance. Right-sided aortic arch with diverticulum of Kommerell was also detected (Fig. 5). After implanting permanent pacemaker, the patient's symptom was resolved and he was discharged with prescription of angiotensin converting enzyme (ACE) inhibitor without any complication.

DISCUSSION

Most cases of L-TGA are associated with other cardiac anomalies such as large ventricular septal defect, pulmonary stenosis, Ebstein-like malformation, MV abnormalities, and these associated anomalies determine the signs and symptoms at presentation.⁵⁾ However, less than 20% of L-TGA patients without any other cardiac anomalies are usually asymptomatic and present later in life with signs and symptoms of arrhythmia and heart failure.^{6,7)} In such cases, heart failure may occur because morphological RV as a systemic pump becomes progressively dysfunctional, and concomitant tricuspid valve (systemic AV valve) regurgitation causes volume overload of the morphological RV. Complete AV block also frequently occurs because bundle branch are vulnerable to fibrosis in L-TGA.⁸⁾

In our case, the patient was incidentally diagnosed with L-TGA when implanting permanent pacemaker due to complete AV block at a relatively earlier age. With ventricular inversion, bundles are also inverted and this makes septal activation from right to left. This makes his electrocardiogram of Q-waves in lead II, III and absent Q waves in lead V5, 6 besides complete AV block. Therefore, electrocardiogram from L-TGA could be misinterpreted as inferior myocardial infarction.

During the coronary angiography, we could not examine right coronary artery due to engagement failure. In congenital heart disease, invasive coronary angiography could be replaced with advanced imaging technique such as computed tomography, magnetic resonance imaging, and nuclear imaging due to associated coronary anomaly.⁹⁾



Fig. 1. Electrocardiogram (A) and Holter (B) on admission. 2:1 atrioventricular (AV) block, Q wave in right precordial limb lead (V1, II, III), absent Q wave in left precordial lead (V5, V6) and left axis deviation (A), complete AV block (B) (arrow: P wave, arrowhead: QRS complex).

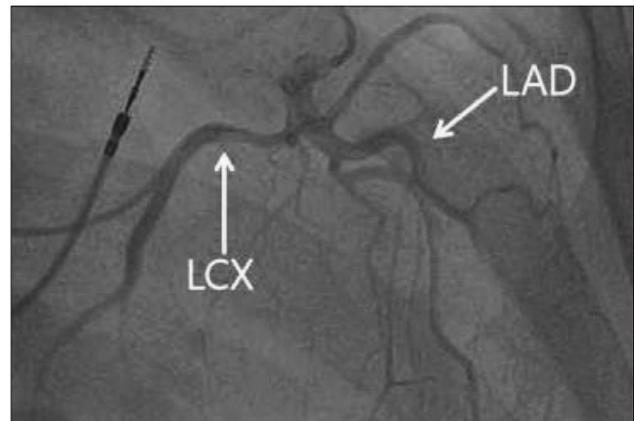


Fig. 2. Coronary angiography. There was no significant stenosis in left coronary arteries. Right coronary artery could not be examined due to failure of catheter engagement. LAD: left anterior descending artery, LCX: left circumflex artery.

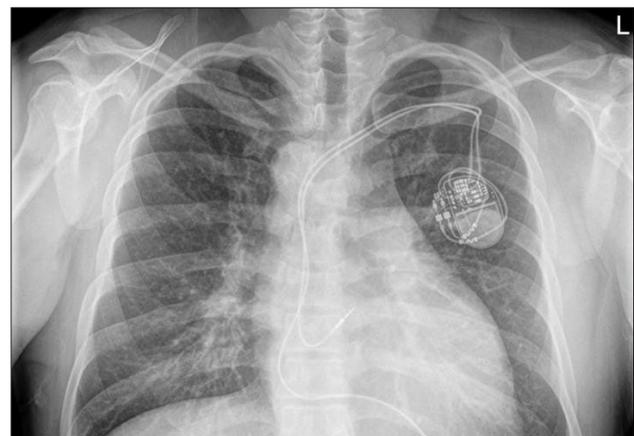


Fig. 3. Chest X-ray after permanent pacemaker insertion. A lead was positioned at RA, and V lead was positioned at functional RV (morphologic LV). Right sided aortic arch was also identified. RA: right atrium, RV: right ventricle, LV: left ventricle.

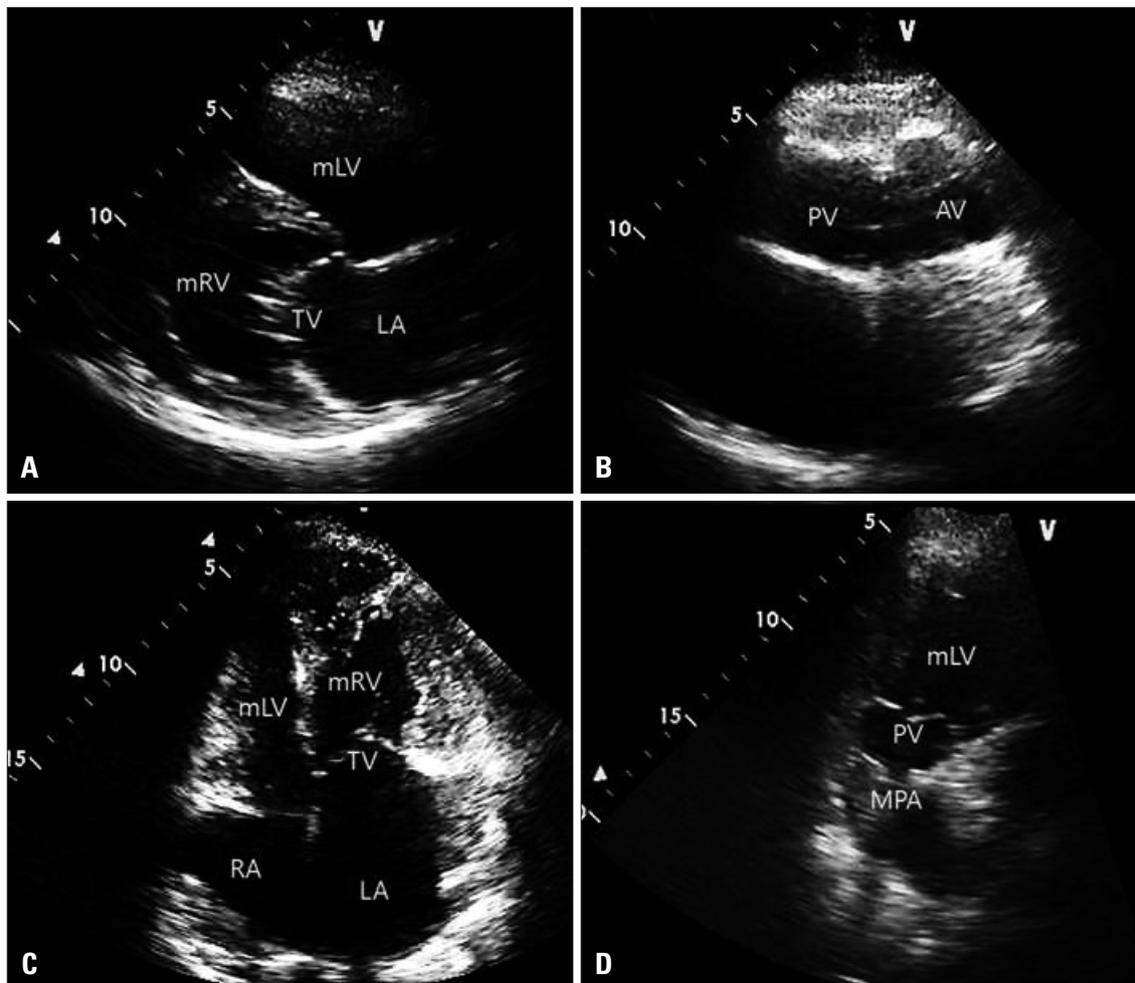


Fig. 4. Transthoracic echocardiography. Parasternal long axis view (A), parasternal short axis view (B), apical four chamber view (C), right ventricular outflow view (D). mRV was located at systemic location and mLV at pulmonary circulation. Note PV and AV location was inverted at short axis view (B). Left-sided atrioventricular valve is inferiorly located closer to the cardiac apex than right-sided atrioventricular valve (C). AV: aortic valve, LA: left atrium, mLV: morphologic left ventricle, MPA: main pulmonary artery, mRV: morphologic right ventricle, PV: pulmonary valve, RA: right atrium, TV: tricuspid valve.

We thought that the chest pain and dyspnea on exertion was developed due to complete AV block, because both symptoms were resolved after pacemaker insertion, although it also could be explained as myocardial ischemia due to reduced coronary flow reserve in L-TGA.¹⁰

In case of permanent pacemaker insertion, ventricular lead was positioned at morphologic LV. This could negatively impact systemic RV function due to interventricular dyssynchrony. It was reported that cardiac resynchronization therapy could help severe systemic ventricular dysfunction in L-TGA cases.²

There are two treatment options for L-TGA patients. One is periodic follow-up with 2 dimensional-echocardiography for worsening ventricular function or tricuspid regurgitation, and the other is corrective surgery.¹¹⁻¹³ Our patient had relatively normal ejection fraction (57%), mild tricuspid regurgitation (grade 2) and his sign and symptom was improved after implanting permanent pacemaker. Therefore the patient was followed up with medical treatment rather than surgical repair. In

case of medical follow-up, there is a paucity of evidence on proper medication. ACE inhibitor or beta blocker has not been studied in the L-TGA population. Therefore most of patients received conventional systemic LV protection strategies.^{2,5} ACE inhibitor was prescribed for his hypertension and ventricular dysfunction protection.

When considering treatment options including periodic follow-up and corrective surgery, morphologic RV dysfunction is important. In L-TGA, ejection fraction measurement could be inaccurate by 2 dimensional-echocardiography because morphologic RV has multiple coarse trabeculations.⁹ Therefore cardiac magnetic resonance imaging is considered as an additional tool for measurement of ejection fraction in patients with L-TGA.

In conclusion, as adult congenital cardiac diseases grow exponentially, congenital anomalies such as L-TGA should be considered when complete AV block is developed at a relatively younger age.

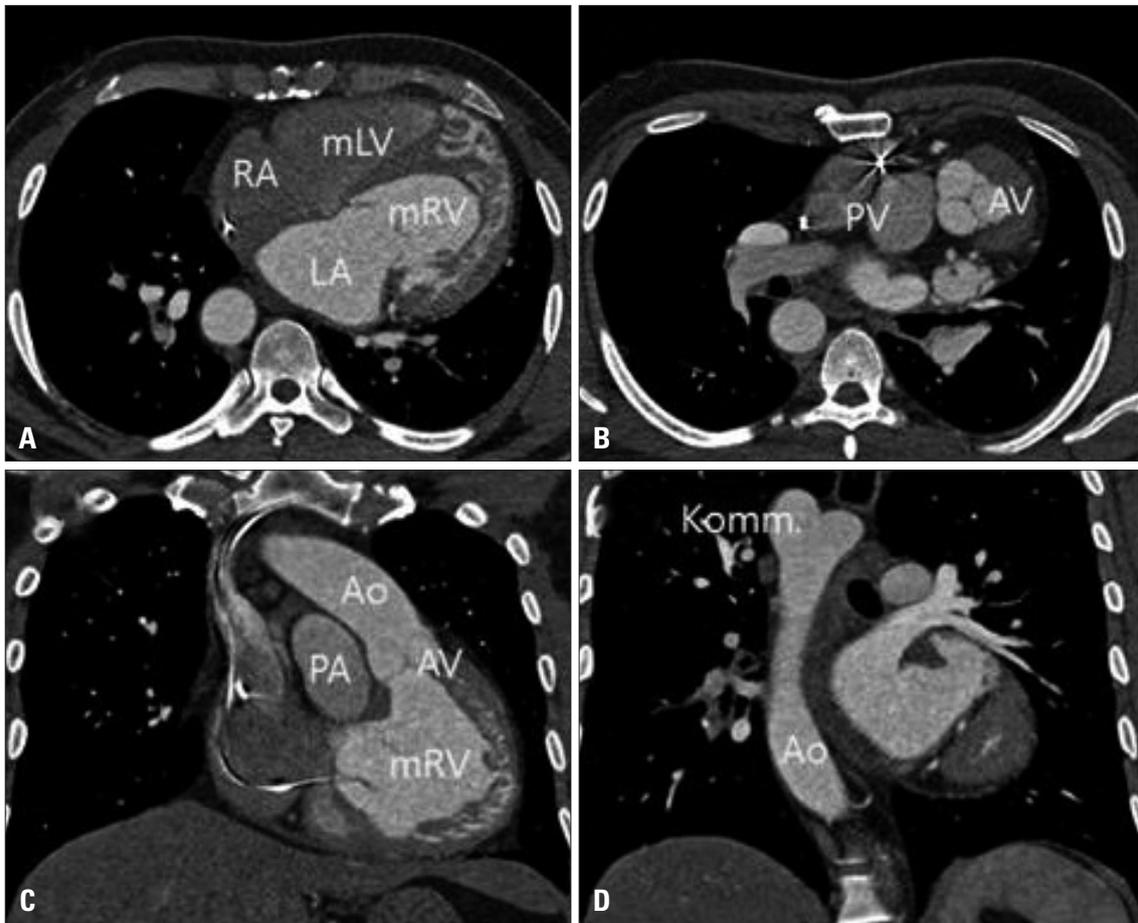


Fig. 5. Cardiac computed tomography, left-sided mRV has moderator band, coarse trabeculations (A). Aortic valve was abnormally located anterior, superior, and to the left of the pulmonary valve. Aorta arises anterior and leftward of pulmonary artery from the mRV (B and C). Right sided descending thoracic aorta with diverticulum of Kommerell (D). Ao: aorta, AV: aortic valve, Komm: diverticulum of Kommerell, LA: left atrium, mLV: morphologic left ventricle, mRV: morphologic right ventricle, PA: pulmonary artery, RA: right atrium.

REFERENCES

1. Warnes CA. *Transposition of the great arteries.* *Circulation* 2006;114:2699-709.
2. Hornung TS, Calder L. *Congenitally corrected transposition of the great arteries.* *Heart* 2010;96:1154-61.
3. Anderson RH, Becker AE, Arnold R, Wilkinson JL. *The conducting tissues in congenitally corrected transposition.* *Circulation* 1974;50:911-23.
4. Wilkinson JL, Smith A, Lincoln C, Anderson RH. *Conducting tissues in congenitally corrected transposition with situs inversus.* *Br Heart J* 1978;40:41-8.
5. Graham TP Jr, Bernard YD, Mellen BG, Celermajer D, Baumgartner H, Cetta F, Connolly HM, Davidson WR, Dellborg M, Foster E, Gersony WM, Gessner IH, Hurwitz RA, Kaemmerer H, Kugler JD, Murphy DJ, Noonan JA, Morris C, Perloff JK, Sanders SP, Sutherland JL. *Long-term outcome in congenitally corrected transposition of the great arteries: a multi-institutional study.* *J Am Coll Cardiol* 2000;36:255-61.
6. Presbitero P, Somerville J, Rabajoli F, Stone S, Conte MR. *Corrected transposition of the great arteries without associated defects in adult patients: clinical profile and follow up.* *Br Heart J* 1995;74:57-9.
7. Beauchesne LM, Warnes CA, Connolly HM, Ammash NM, Tajik AJ, Danielson GK. *Outcome of the unoperated adult who presents with congenitally corrected transposition of the great arteries.* *J Am Coll Cardiol* 2002;40:285-90.
8. Prieto LR, Hordof AJ, Secic M, Rosenbaum MS, Gersony WM. *Progressive tricuspid valve disease in patients with congenitally corrected transposition of the great arteries.* *Circulation* 1998;98:997-1005.
9. Kilner PJ. *Imaging congenital heart disease in adults.* *Br J Radiol* 2011;84 Spec No 3:S258-68.
10. Hauser M, Bengel FM, Hager A, Kuehn A, Nekolla SG, Kaemmerer H, Schwaiger M, Hess J. *Impaired myocardial blood flow and coronary flow reserve of the anatomical right systemic ventricle in patients with congenitally corrected transposition of the great arteries.* *Heart* 2003;89:1231-5.
11. Murtuza B, Barron DJ, Stumper O, Stickley J, Eaton D, Jones TJ, Brawn WJ. *Anatomic repair for congenitally corrected transposition of the great arteries: a single-institution 19-year experience.* *J Thorac Cardiovasc Surg* 2011;142:1348-57.e1.
12. Hraska V, Duncan BW, Mayer JE Jr, Freed M, del Nido PJ, Jonas RA. *Long-term outcome of surgically treated patients with corrected transposition of the great arteries.* *J Thorac Cardiovasc Surg* 2005;129:182-91.
13. 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;23:137-45.