Cardiology in the Young

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Transposition of the great arteries with an intact ventricular septum in older children

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Brief Report

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Abstract

Complete transposition of the great arteries is a common life-threatening complex cyanotic congenital heart disease in infants, resulting in the operation usually performed about one week after birth. However, little is known about the surgical strategy and experience of transposition of the great arteries with an intact ventricular septum in older patients. Herein, we present an abandoned 7-year-old boy with severe cyanosis with clubbed fingers and toes and then diagnosed with transposition of the great arteries with an intact ventricular septum, atrial septal defect, patent ductus arteriosus, and pulmonary hypertension. The patient underwent a two-staged procedure: an aortopulmonary shunt and pulmonary artery banding were performed at the first stage, followed by the Switch operation, defect repair, and patent ductus arteriosus ligation, all of which were successfully performed. The patient was discharged on the 15th day after the operation, and the arterial oxygen saturation returned to normal level (99%). The illustrative report highlights the essence of raising awareness and developing accurate treatment strategy of transposition of the great arteries, especially in remote rural areas of eastern countries, where the level of health care and services is relatively underdeveloped.

Introduction

Complete transposition of the great arteries, a common life-threatening complex cyanotic congenital heart disease in infants, is a kind of artery malformation with the aorta arising anterior to the pulmonary artery and from the right ventricle, while the pulmonary artery lies posterior to the aorta and arises from the left ventricle.[1,2] Transposition of the great arteries can be divided into dextro-transposition of the great arteries and levo-transposition of the great arteries (Figure 1).[1,3] The most common type of transposition of the great arteries is called dextro-transposition of the great arteries, which is characterised by the right ventricle on the right side of the left ventricle and the aorta on the front and right side of the pulmonary artery. The lesser-known form of levo-transposition of the great arteries, also known as corrected transposition of the great arteries, is characterised by the left ventricle located on the right side of the right ventricle. The main pulmonary artery and aorta are anatomically correct. However, due to ventricular inversion, the aorta merges with the right ventricle, and the main pulmonary artery merges with the left ventricle.[1] Cyanosis usually occurs within 30 days after birth, so patent ductus arteriosus and ventricular septal defect are needed to mix oxygen-rich and oxygen-deficient blood.[4] The initial treatment of transposition of the great arteries in patients is to ensure adequate oxygenation.[5] Once the haemodynamics of the patients are stable, corrective surgery can be performed.[6] The surgical repair of transposition of the great arteries is usually carried out about one week after birth, which can be divided into two types: one-stage radical operation and staged radical operation according to the number of operations, such as arterial switch operation, Rastelli procedure, the Mustard and Senning procedure, Nakaidoh procedure, and Réparation à l'Etage ventriculaire.[7–10] Compared with other congenital heart diseases, transposition of the great arteries has the characteristics of a critical condition, complex operation process, long treatment cycle, and high operation cost, which has a great impact on the family and society, so it has great clinical significance.

Presentation

An abandoned 7-year-old boy from a remote village in southern China, who experienced severe cyanosis and tachypnoea, was referred to our institution for cardiac defect repair. Physical examination revealed a thin body, poor nutritional development, hexadactyly on the right hand, a decreased arterial oxygen saturation level (50% without oxygen at room temperature), and severe cyanosis with clubbed fingers and toes. There are no significant indicators of liver and kidney function. The electrocardiogram showed no obvious abnormality. Meanwhile, acute heart failure with cyanosis, such as patent ductus arteriosus and tetralogy of Fallot, still requires differential diagnosis and consideration in this age group. Chest radiography showed enlarged heart shadow, apical protuberance, increased pulmonary vascularity, and an "egg on a string"

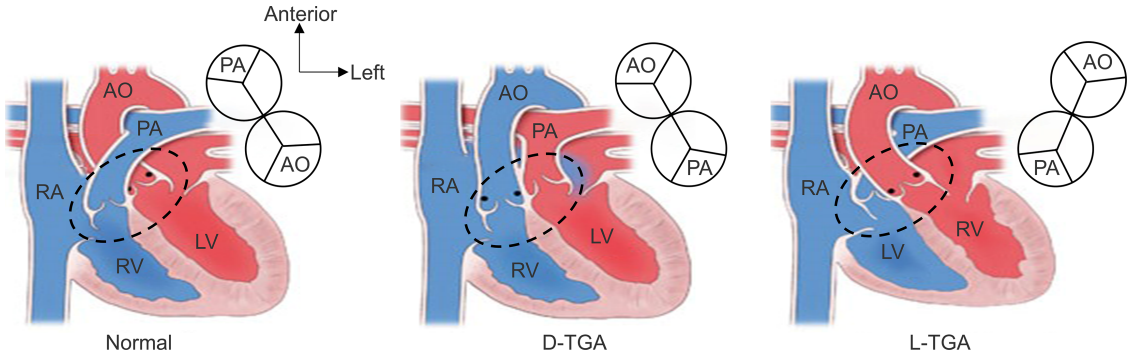

Figure 1. Schematic diagram shows the spatial relationships between the great arteries in patients with TGA. TGA = transposition of the great arteries; D = dextro; L = levo; AO = aorta; PA = pulmonary artery.

appearance (Figure 2A). Echocardiography revealed transposition of the great arteries, atrial septal defect, patent ductus arteriosus, pulmonary hypertension (Figure 2B), and the ventricular septum protruded to the left. Cardiac CT was performed and confirmed transposition of the great arteries, with the right ventricle on the right side of the left ventricle and the aorta on the front and right side of the pulmonary artery (Figure 3A); patent ductus arteriosus (Figure 3C), with the aorta connected to the right ventricle and pulmonary artery connected to the left ventricle, reversing the anatomical relationship of large arteries (Figure 3B–F); and normal origins of the coronary arteries (Figure 3B and D). Diagnostic cardiac catheterisation further confirmed pulmonary hypertension and left ventricular hypotension (data not shown).

There has been no relevant report at home and abroad, which has brought us great trouble in the reference to experience and the choice of surgical scheme. Under these circumstances that the patient presents low left ventricular pressure, a staged repair may be performed for ventricular training to induce left ventricular hypertrophy, which would provide adequate postoperative systemic circulatory support. Following communication with the patient's family, the aortopulmonary shunt +pulmonary artery banding+patent ductus arteriosus ligation then were performed to exercise the function of the left ventricle and ensure adequate oxygenation. Repeat postoperative evaluation was then performed before the arterial switch to measure and assess left ventricular wall thickness, ventricular volumes, ejection fraction, left ventricular muscle mass, etc. After more than two months of left ventricular training, the patient revealed normal left ventricular wall thickness and muscle mass for age. Then the Switch operation, defect repair, and patent ductus arteriosus ligation were successfully performed. Cardiac CT revealed the pulmonary artery on the front and right side of the aorta (Figure 4A), with the pulmonary artery and aorta anatomically correct (Figure 4B–F). The patient was discharged on the 15th day after the operation, and the arterial oxygen saturation returned to normal level (99%). There was no recurrence or other serious complications in the follow-up, but long-term follow-up is necessary.

Comment

Transposition of the great arteries is a common life-threatening complex cyanotic congenital heart disease in infants and young children, and its incidence rate is about 5–7% of congenital heart disease.[11] The clinical manifestations of transposition of the great arteries depend entirely on the degree of blood mixing between systemic circulation and pulmonary circulation. Most patients have symptoms and signs in the neonatal period, such as the following typical clinical manifestations: cyanosis, tachypnoea, and murmur.[12,13] Because of the loss of the physiological principle of circulatory interaction between systemic circulation and pulmonary circulation, it is necessary to mix oxygen-enriched and hypoxia blood to maintain the life of the infant. Mixing can occur through the atrial septal or ventricular septal defect, patent ductus arteriosus, or bronchopulmonary collateral circulation.[4] Otherwise, the disease will deteriorate rapidly, resulting in acidosis, progressive deterioration, and even death. Thus, accurate diagnosis and early treatment are very important.

Echocardiography is the main non-invasive diagnostic tool in early screening, which can reveal the abnormal origin of the aorta and pulmonary trunk and any related intracardiac defects.[14] At the same time, some other auxiliary examinations are also necessary, such as electrocardiography, chest radiography, CT angiography, and even cardiac catheterisation. Recently, CT angiography has become a reference standard for the identification and qualitative analysis of macrovascular abnormalities. It can not only accurately diagnose the origin and course of arteries but also provide a three-dimensional evaluation of the relationship between vessels and adjacent structures and provide cross-sections of cardiac structures from different angles.[14,15] Therefore, it can be considered as the preferred imaging method for the non-invasive depiction of macrovascular anatomy, and it is a valuable diagnostic tool in surgical intervention and postoperative follow-up. Cardiac catheterisation angiography is an invasive method, but it is rarely used to diagnose transposition of the great arteries. However, it is the gold standard to elucidate the origin of the coronary artery,[14] and it also provides the diagnostic basis for collateral circulation, intracardiac defect, and vascular anatomy and provides a critical important guidance for balloon atrial septostomy.[16–19]

The initial treatment of patients with transposition of the great arteries is centred on ensuring adequate oxygenation. The common management strategy is to use prostaglandin E1 to keep the ductus arteriosus unobstructed and perform balloon atrial septostomy.[5,6] Once the patient's haemodynamics is stable, corrective surgery can be performed. Surgical repair of transposition of the great arteries is usually performed around the first week after birth. At present, transposition of the great arteries has two common surgical methods: arterial switch operation, which is the standard operation for patients without pulmonary artery stenosis[20] (if there are other intracardiac defects, they will also be repaired during this period), and Rastelli procedure, which is suitable for patients with transposition of the great arteries, large ventricular septal defect, and pulmonary artery stenosis.[21] Other

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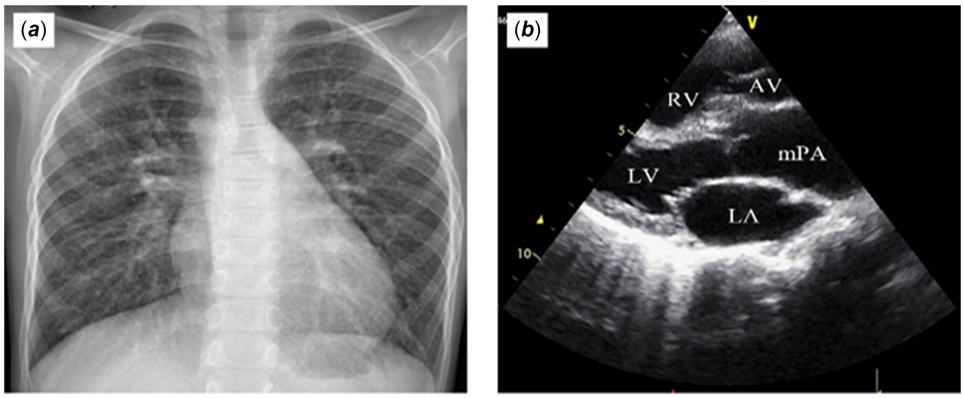

Figure 2. Chest radiography showed enlarged heart shadow, apical protuberance, and an "egg on a string" appearance (***a***). Echocardiography revealed transposition of the great arteries (***b***). LA = left atrium; AV = aortic valve; LV = left ventricle; RV = right ventricle, mPA = main pulmonary artery.

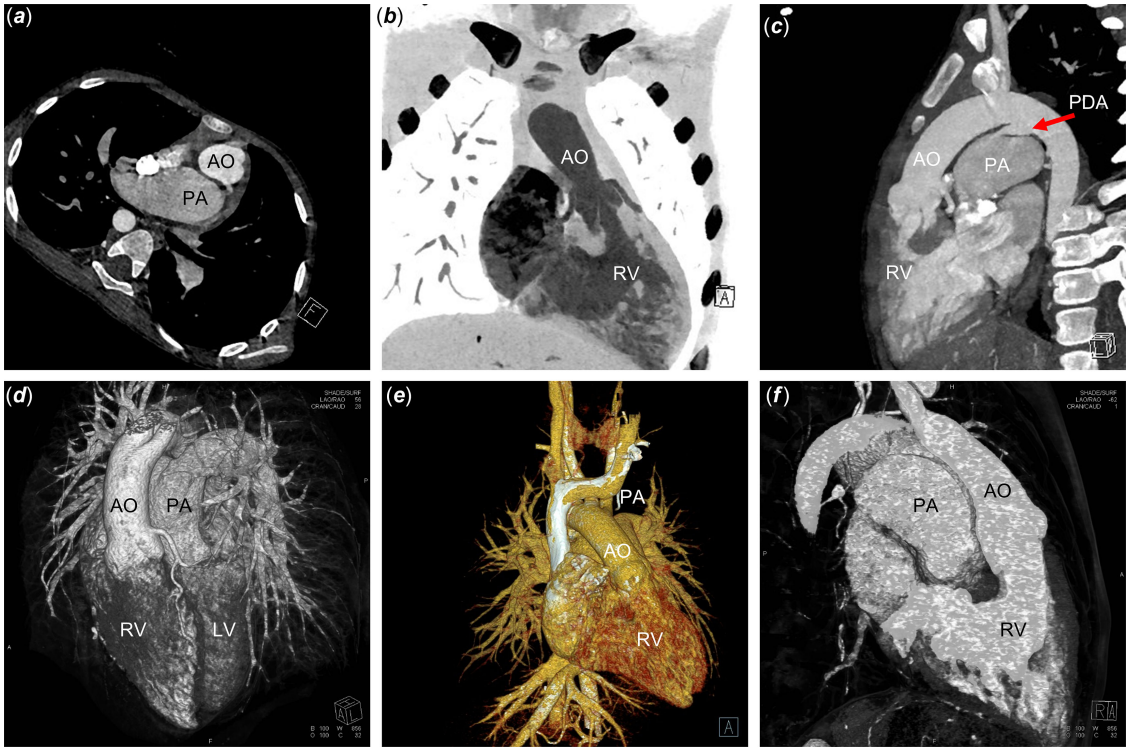

Figure 3. Preoperative cardiac CT confirmed transposition of the great arteries (***a***), with the anatomical relationship of large arteries reversed (***b***–***f***), and PDA (***c***). AO = aorta; PA = pulmonary artery; LV = left ventricle; RV = right ventricle; PDA = patent ductus arteriosus.

corrective procedures include the Mustard and Senning procedure, Nakaidoh procedure, and Réparation à l'Etage ventriculaire, but these are less commonly performed.[7–10]

Although the prognosis of transposition of the great arteries patients after surgical correction is good and the survival rate is more than 90%,[1,10] there may be some complications. These include aortic root dilatation, pulmonary artery stenosis, arrhythmia, obstruction or leakage of the baffle (Rastelli procedure), and aortic regurgitation.[22] Recent studies have shown that adolescents who need special education services after experiencing arterial switch operation may suffer from attention deficit hyperactivity disorder.[23] Therefore, it is very important to give children more care and education, even special education services, and regular postoperative reviews for the improvement of children's quality of life.

In this case, the other complex heart defects that are associated with cyanosis and that make survival possible in the first place are much more interesting in terms of differential diagnosis: tricuspid atresia or tetralogy of Fallot. This child only survived because it had atrial septal defect and patent ductus arteriosus. We successfully reported for the first time the efficacy and safety of staged repair of an older transposition of the great arteries with an intact ventricular septum. Considering the large population of China, the number of transposition of the great arteries cases reported is far fewer than it should be. We believe that the following main factors contribute to this phenomenon. First, there is a lack of a stable and advanced medical system, especially in rural areas of China. Because of the lack of an excellent medical system, the opportunity for surgical repair for transposition of the great arteries is limited. Second, financial constraints hinder the referral

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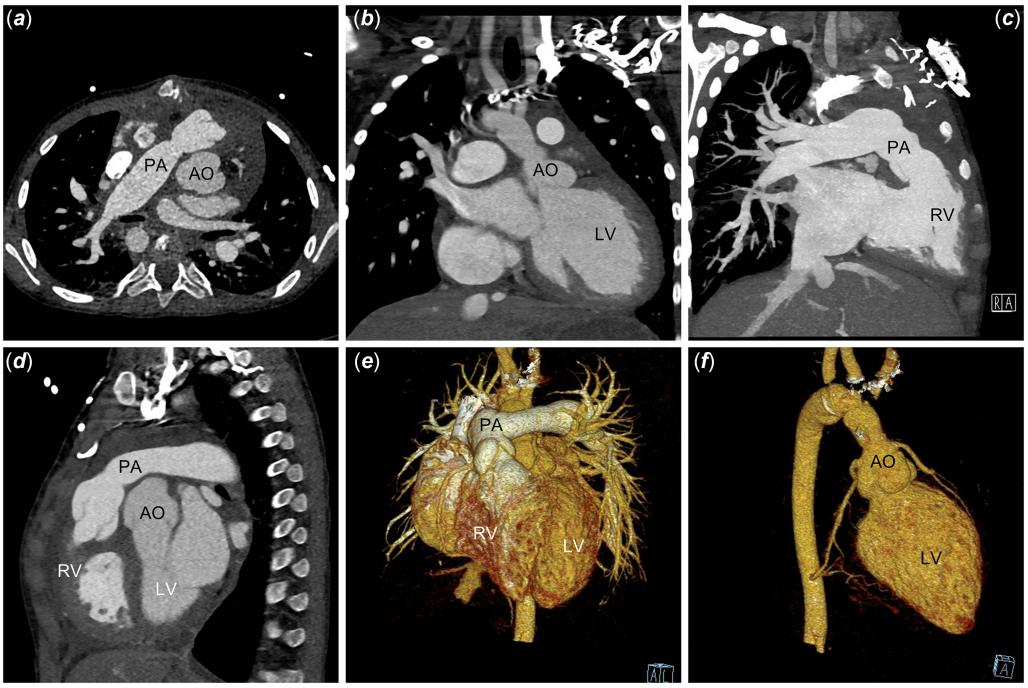

Figure 4. Postoperative cardiac CT revealed the pulmonary artery on the front and right side of the aorta (***a***), with the pulmonary artery and aorta anatomically correct (***b–f***). AO = aorta; PA = pulmonary artery; LV = left ventricle; RV = right ventricle.

of patients with transposition of the great arteries to better hospitals. There are still many patients with transposition of the great arteries or complex congenital heart disease who are not diagnosed and treated in time in the early stage. Finally, due to the lack of understanding and publicity of transposition of the great arteries, especially in remote areas of our country, people ignore and misunderstand transposition of the great arteries and miss the best opportunity for treatment. Taken together, this illustrative report highlights the essence of raising awareness and developing accurate treatment strategy of the disease, especially in remote rural areas of eastern countries, where the level of health care and services is relatively underdeveloped.

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Competing interests. The authors declare no conflicts of interest.

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