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Successful Surgical Correction of Total Anomalous Systemic Venous Drainage: A Report of 2 Cases

Yin-qi Xu, Jian-ping Yao, Zhi-ping Wang, Xi Zhang, Pei-wu Sun, Zhong-kai Wu

Department of Cardiac Surgery, First Affiliated Hospital, Sun Yat-sen University, Guangzhou, China

ABSTRACT

Total anomalous systemic venous drainage is a rare malformation and only limited cases have been reported previously. Here we report two cases of total anomalous systemic venous drainage with successful surgical correction.

INTRODUCTION

Total anomalous systemic venous drainage (TASVD) is a rare malformation and few cases have been reported previously [Roberts 1972; Danielson 1973; Wu 2000]. Two cases of TASVD with successful surgical correction and long-term follow-up results are reported in this article. The clinical features, diagnosis, and methods of surgical correction are discussed. According to the malformation, we suggest that TASVD should be classified into two types: simple type TASVD, usually with a secondary atrial septal defect (ASD), and complex type TASVD, with other malformations.

CASE REPORTS

Case 1

A 2-year 10-month-old girl, with a bodyweight of 8 kg and with cyanosis for 2 and half years was admitted to The First Affiliated Hospital, SUN Yat-sen University on June 4, 1999. Physical examination showed moderate cyanosis at rest with marked finger and toe clubbing. There were no signs of cardiac failure. The apex beat pointed to the right side of the sternum. There was a systolic murmur grade 2 of 6 at the right sternal border along the 2nd to the 4ths intercostal spaces. The electrocardiogram showed left ventricular hypertrophy. Chest radiography revealed a slightly enlarged heart. The position of the apex directed to the right of the sternum, the stomach lay on the right side, and the liver on the left. Pulmonary vascularity was slightly less than normal. Echocardiography found a central atrial septal defect (ASD) 9 mm in diameter. The inferior vena cava (IVC) anomalously drained to the left atrium. The spleen was located on the right side and its size and shape was normal. Ultra-fast computed tomography showed double superior vena cava (SVC). The left SVC (LSVC), right SVC (RSVC), and IVC were connected with the left atrium. A central ASD was present. The diagnosis of TASVD associated with dextrocardia and ASD was diagnosed before the operation.

The patient underwent a corrective procedure on June 22, 1999. The heart was exposed through a median sternotomy incision. The heart apex was directed to the right side of the chest. The right atrium, SVC, and IVC lay on the left and the RSVC on the right side. The LSVC, RSVC, and IVC were connected with the left atrium. After cardiopulmonary bypass was instituted and aortic clamping performed, the right atrium was opened. A pericardial baffle was used to repair the ASD and directed the LSVC, RSVC, and IVC to the right atrium (Figure 1). The aortic clamping time was 84 minutes. After the operation, the hemodynamic state was stable and cyanosis disappeared. The patient was ventilated postoperatively and extubated 24 hours after the operation. The patient was discharged 12 days after surgery. The patient survived with normal growth and physical exercise during 7-year follow-up periods. She is now studying at primary school.

Case 2

A 14 year-old girl, with a bodyweight of 40 kg and with severe cyanosis after birth was hospitalized on October 11, 1999. The patient showed severe cyanosis and marked finger and toe clubbing at rest and had dyspnea on exertion frequently without signs of cardiac failure. The apex beat was pointed to the left side of the sternum. There was a systolic grade 4 of 6 murmur and a palpable thrill at the left sternal border along the 2nd to the 4th intercostal spaces. The electrocardiogram showed left deviation of the axis. Chest radiography revealed a moderate enlargement of the left ventricle. Pulmonary vascularity was slightly less than normal. Echocardiography showed tricuspid atresia (TA) associated with L-transposition of the great arteries, pulmonary stenosis,

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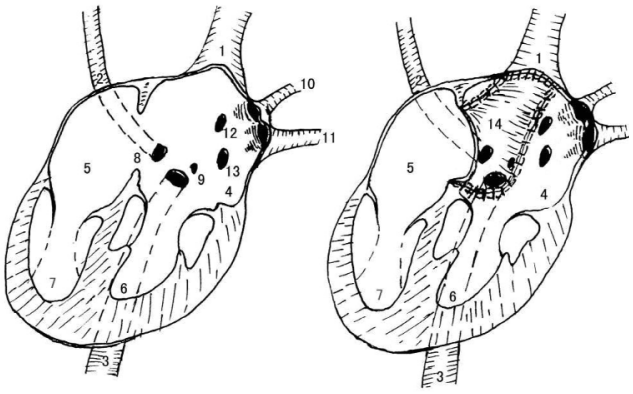
Correspondence: Dr. Zhong-kai Wu, Department of Cardiac Surgery, First Affiliated Hospital, Sun Yat-sen University, Guangzhou 510089, China; 86-20-87332200; fax: 86-20-87332308 (e-mail: wuzhongkai@hotmail.com).



1. Left superior vena cava
2. Right superior vena cava
3. Inferior vena cava
4. Left atrium
5. Right atrium
6. Left ventricle
7. Right ventricle
8. Atrial septal defect
9. Coronary sinus
10. Left superior pulmonary vein
11. Left inferior pulmonary vein
12. Right superior pulmonary vein
13. Right inferior pulmonary vein
14. Patch

Figure 1. Case 1 of total anomalous systemic venous drainage reported, before (left) and after (right) surgical correction.

a common atrium, and a huge ventricular septal defect (VSD) 3.4 cm in diameter. The diagnosis of TA associated with a common atrium and huge VSD was diagnosed preoperatively.

The patient accepted a surgical correction of total cavopulmonary connection under conventional cardiopulmonary bypass without aortic clamping on October 25, 1999. The heart was exposed through a median sternotomy incision. The ascending aorta lay on the left side and the pulmonary artery on the right side. There was a TA associated with pulmonary stenosis, common atrium, a huge VSD, and the LSVC. The RSVC, LSVC, and IVC all connected with the left atrium. The diagnosis of TASVD associated with TA was then confirmed. After cardiopulmonary bypass was instituted on the beating heart, a procedure of total cavopulmonary connection was performed. Via the incision of the right atrium to remove the atrial septum, an artificial conduit was created with a pericardial patch to connect the IVC to the RSVC. Then both the RSVC and LSVC were incised transversally at the level of the right and left pulmonary artery. The distal opening of the RSVC and LSVC were anastomosed to the upper side of the right and left pulmonary artery, and the proximal opening of the RSVC was anastomosed to the lower side of the right pulmonary artery, and the proximal opening of the LSVC and the pulmonary arterial root were closed (Figure 2). The aorta was not clamped during the operation. The hemodynamic function was stable and cyanosis disappeared after surgery. The patient was extubated 24 hours after the operation and discharged 21 days later. Seven-year follow-up periods did not show any abnormal findings with normal growth and physical exercise.

DISCUSSION

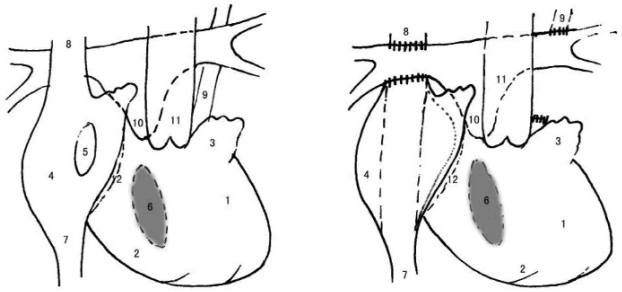
Anomalies of systemic venous drainage are not uncommon, and persistent LSVC represents the most common form of anomalous systemic venous return, occurring in 2% to 4% of all congenital heart disease. However, TASVD is an extremely unusual congenital heart disease. Reports of TASVD are so rare in medical literature—no more then 10 cases have been reported in medical journals—that the characteristics of the disease were not clearly depicted until the 2000s [Roberts 1972; Danielson 1973; Wu 2000]. It is difficult to distinguish TASVD from other congenital cyanotic heart diseases before surgery. Even echocardiography and cineangiocardiography could not provide a correct diagnosis [Roberts 1972; Danielson 1973; Wu 2000; Tacy 2001].

Usually TASVD is combined with complex congenital heart disease. After reviewing the reported 5 cases of TASVD from the literature as well as our 2 cases, some common findings might be helpful for the diagnosis of TASVD. These include cyanosis, atrial/ventricle level communication (ASD or common atrium, VSD) and bi-SVC in all cases. TASVD is frequently associated with complex malformations, such as complete atrioventricular canal, transposition of the great arteries, TA, VSD, and dextrocardia. Electrocardiography usually shows a left deviation of axis or the left ventricular hypertrophy. Therefore, cyanosis with left heart strain in electrocardiography may indicate the diagnosis of TASVD. Ultra-fast computed tomography might be a better way to establish the diagnosis, as showed in our first case.

In general, anomalous systemic venous connections are classified as follows [Tacy 2001]:

- Anomalies of RSVC
- Persistent LSVC
- Anomalies right IVC
- Persistent left IVC
- Anomalous connection of hepatic veins
- Anomalies involving the coronary sinus

Based on other reports of TASVD, combined with our limited experience, we suggest that TASVD should be



1. Left ventricle
2. Right ventricle
3. Left atrium
4. Right atrium
5. Atrial septal defect
6. Ventricular septal defect
7. Inferior vena cava
8. Right superior vena cava
9. Left superior vena cava
10. Pulmonary atresia
11. Aorta
12. Tricuspid atresia

Figure 2. Case 2 of total anomalous systemic venous drainage reported, before (left) and after (right) surgical correction.

classified into two types: simple type TASVD, usually with a secondary ASD, and complex type TASVD with other malformations, no matter whether the secondary ASD exists or not. Black reported a case of TASVD to coronary sinus in association with HLHS in 1997. In our second case, TASVD associated with TA would be complex type TASVD. Surgical correction of simple type TASVD could be successfully achieved to repair ASD and direct the LSVC, RSVC, and IVC to return to the right atrium by a pericardial baffle. The narrowing of the orifice of the vena cava and the pulmonary veins should be avoided. For the complex type, the result of the surgical management of TASVD was also satisfied with different surgical procedures in our second case. Total cavopulmonary connection may be a good option in such a rare type of cyanotic congenital heart diseases.

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