Nikaidoh Procedure for Situs Inversus and Transposition of Great Arteries with Ventricular Septal Defect and Pulmonary Stenosis

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We present the case of a patient who underwent a Nikaidoh procedure for total correction of transposition of the great arteries (TGA), ventricular septal defect (VSD), and pulmonary stenosis (PS). A young girl, 2 years and 4 months of age, weighing 3400 g at birth, was diagnosed with TGA, VSD, patent ductus arteriosus (PDA), and left ventricular outflow tract obstruction (LVOTO), with valvular and subvalvular PS. Because of frequent cyanosis, she had received a right-sided modified Blalock-Taussig (mBT) shunt when she was 8 months old. The Nikaidoh procedure was used for total correction (aortic root translocation, LVOT enlargement, VSD patch repair, neoaorta and coronary button reimplantation, and neopulmonary artery reconstruction by monocusp valved autologous pericardial patch with the Lecompte maneuver). The patient's postoperative course was uneventful, and she was discharged from the surgical intensive care unit 10 days after the operation.

Key Words: Nikaidoh procedure • Pulmonary stenosis • Transposition of great arteries • Ventricular septal defect

INTRODUCTION

In our institution, the in-hospital survival rate after surgical correction of the transposition of great arteries (TGA) is improving.[1] However, in the field of cardiovascular surgery, surgical management of patients with TGA, ventricular septal defect (VSD), left ventricular outflow tract obstruction (LVOTO), and pulmonary stenosis (PS) continues to be a challenge. The unusual anatomical features of patients with situs inversus and dextrocardia make total correction even more difficult. Three methods have been developed for correcting TGA, VSD, and PS: the Rastelli, réparation á l'etage ventriculaire (REV), and Nikaidoh procedures. We report a case in which situs inversus and dextrocardia with TGA, VSD, and PS were corrected using the Nikaidoh procedure.

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CASE REPORT

A female patient was diagnosed with situs inversus and dextrocardia. She had atrioventricular concordance and ventriculoarterial discordance and TGA with perimembranous inlet-type VSD and valvular and subvalvular PS at birth. A modified Blalock-Taussig (mBT) shunt was created when she was 8 months old due to intermittent desaturation. She adapted well to the shunt and showed no obvious growth retardation. When she was 2 years and 4 months old and her body weight was 11 kg, she was admitted to our hospital and scheduled to undergo the next step of surgical intervention.

A grade 4/6 systolic murmur was audible at the right sternal border and oxygen saturation was approximately 75-85%. Chest radiography revealed signs typical of si-

tus inversus with dextrocardia and a right-sided gastric bubble. The electrocardiogram also showed typical findings such as inverted P and T waves on lead 1 and diminution of R wave from V1-V6. The ejection fraction calculated via follow-up echocardiography was 81.2%. The size of the VSD was 9.4 mm. The pulmonary valve was bicuspid with valvular and subvalvular PS. The size of the pulmonary annulus was 7.2 mm and that of the aorta was 17 mm. A Rastelli-type A defect with tricuspid valve straddling was also observed. From the chest computed tomography and 3-dimensional reconstruction, the anatomy of situs inversus, TGA, LVOTO, and valvular and subvalvular PS could be clearly observed. The size and morphology of the pulmonary arteries were normal. The hemodynamic data obtained from cardiac catheterization showed a high-pressure gradient (65 mmHg) across the LVOT and pulmonary valve. No significant pressure gradient was detected across the aortic valve and right ventricle. The coronary arteries of the patient followed the usual pattern and courses with the morphological left coronary artery (LCA), which supplies branches to the left anterior descending (LAD) and left circumflex artery (LCX), and the morphological right coronary artery (RCA) arising from the aortic root. The aim of the intervention was to restore normal circulation (from the morphological left ventricle (LV) to the aorta and from the morphological right ventricle (RV) to the main pulmonary artery), to repair the VSD, and to release the LVOTO. Thus, the Nikaidoh procedure was chosen as a method for total correction.

By performing a standard midline full sternotomy, a cardiopulmonary bypass was established with the aorta, and bicaval cannulation and systemic hypothermia (28 °C) were achieved. The mBT shunt was then divided. After extensive mobilization of both the pulmonary arteries, the main pulmonary artery (MPA) was transected proximally from the left ventricle. After transection of the ascending aorta, the RCA button was harvested before the aortic root was completely separated from the RV, and the Lecompte maneuver was performed to avoid excess tension and kinking (Figure 1B). The conal septum was severed into the VSD, and thus, the VSD and LVOT were enlarged (Figure 1C). The VSD was repaired using a Hemashield prosthetic patch (Hemashield® Vantage™; Maquet Cardiovascular, San Jose, CA, USA) (Figure 1D; Figure 2). The aortic root was translocated posteriorly by suturing the posterior two-thirds of the root to the pulmonary annulus and the anterior one-third to upper border of the Hemashield patch (Hemashield® Vantage™) (Figure 1D). The RCA button was reimplanted into the anterior surface of the aortic root (Figure 1E). Finally, the MPA was directly sutured to the RV opening with autologous pericardium patch augmentation and using a monocuspid valve made of Gore-Tex surgical membrane patch (GORE-TEX® Surgical Membrane, W.L. Gore and Assoc, Flagstaff, AZ, USA) (Figure 1F).

Two days after the surgery, the sternum was approximated and the wound closed. The patient was extubated 1 week later and discharged from the intensive care unit 2 weeks after the operation. Postoperative echocardiography showed moderate pulmonary regurgitation but no neoaortic stenosis.

DISCUSSION

For patients with TGA, VSD, and PS, surgical correction can be performed using 3 methods: Rastelli, REV, and Nikaidoh procedures.

Rastelli and his colleagues proposed the Rastelli procedure in 1969.[2] In patients with VSD, an intracardiac baffle is placed to direct LV blood to the aorta, and an artificial extracardiac valved conduit is constructed to connect the RV and the pulmonary artery. This was the first procedure that restored the role of the morphological LV to support systemic circulation. However, this procedure has some limitations. Unfavorable intracardiac anatomy precluded the adoption of this technique. Fixed size, sternal compression, and obstruction of the extracardiac conduit and recurring LVOTO increase the rate of reoperation. A low freedom from conduit reoperation rate after Rastelli procedure was reported by Kreutzer et al.: 56%, 25%, and 21% after 5, 10, and 15 years, respectively.[3] Another drawback of this procedure was poor long-term survival. Dearani et al.[4] followed up 160 hospital survivors for at least 10 years, and reported an actuarial survival rate of 74% at 10 years and 59% at 20 years after the correctional surgery.

In 1982, Lecompte described the REV procedure.[5]

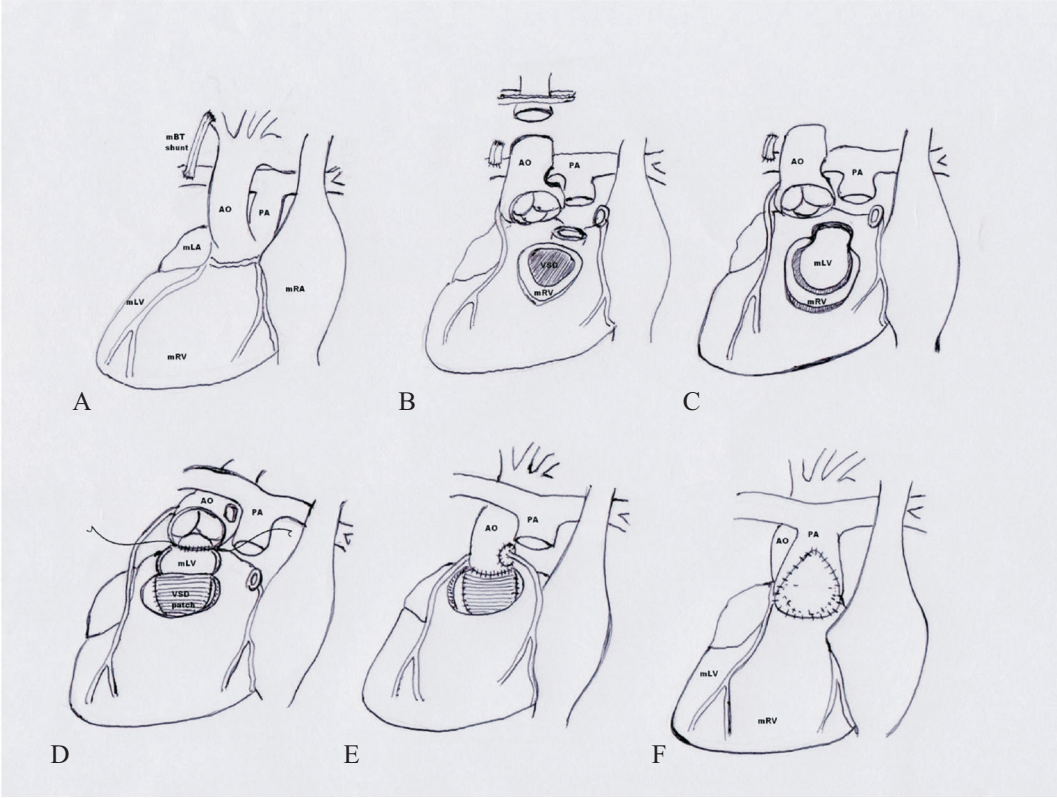

Figure 1. *(A) Situs inversus heart. (B) Harvest the right coronary artery (RCA) button, separate the aortic root from mRV, and transect the main pulmonary artery (MPA). (C) Bisect the conal septum into the ventricular septal defect (VSD) to enlarge the VSD and left ventricular outflow tract (LVOT). (D) VSD patch repair with Hemashield patch. Using the Lecompte maneuver, the aortic root is translocated posteriorly to the LVOT (continuous suture is placed with posterior two-thirds to the pulmonary annulus and anterior one-third to the upper border of the VSD patch). (E) Reimplant the RCA button to the aortic root. (F) Suture the MPA to the right ventricular outflow tract (RVOT) with autologous pericardium patch augmentation and monocuspid patch made of Gore-Tex membrane. AO, aorta; PA, pulmonary artery; VSD, ventricular septal defect; mRV, morphological right ventricle; mRA, morphological right atrium; mLV, morphological left ventricle; mLA, morphological left atrium.*

There were 2 major differences between the REV and Rastelli procedures. First, the muscular outlet septum was partially resected, providing a better pathway for LVOT. Second, the pulmonary trunk was brought anteriorly with the Lecompte maneuver and was re-implanted to the RV. The REV procedure has several advantages over the conventional Rastelli procedure: the REV operation allows complete repair during patient infancy, is feasible in patients with anatomic contraindications to the Rastelli procedure, and reduces the need for reoperation because of conduit obstruction and residual right ventricular outflow tract obstruction (RVOTO) and LVOTO.[6,7] However, despite having better results than the Rastelli procedure, the REV procedure has been reported to result in RV-PA obstruction in 26% of patients.[8]

The Nikaidoh procedure, introduced by Nikaidoh in 1984,[9] aimed to restore the normal anatomical orga-

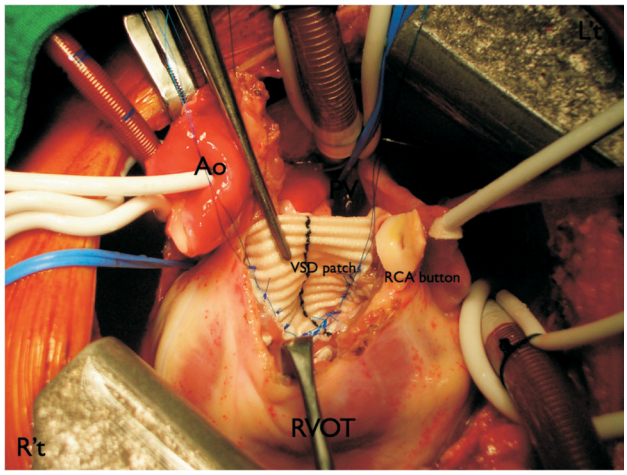

Figure 2. *The conal septum was bisected into the ventricular septal defect (VSD), and a Hemashield patch was applied for VSD repair. After VSD patch repair, the left ventricular outflow tract (LVOT) was enlarged, and thus pulmonary stenosis (PS) was relieved. Ao, aorta; RCA, right coronary artery; RVOT, right ventricular outflow tract.*

nization by which the LVOT and RVOT can be better aligned. It involves harvesting the aortic root from the RV, bisecting the muscular outlet septum, and patch repairing the VSD, thus relieving LVOTO, bringing forward the pulmonary trunk using the Lecompte maneuver, and reimplanting the aortic root to the LVOT and the pulmonary trunk to the RV with or without MPA plasty. In cases of TGA and LVOTO with inlet or small VSD and small RV or straddling atrioventricular valve, which were not amenable to the Rastelli and REV procedures, the Nikaidoh procedure was the only way to achieve biventricular repair.[10] The limitation of the Nikaidoh procedure becomes apparent in the presence of anomalous coronary anatomy. The improper course of a major coronary artery would make harvesting the aortic root dangerous if not impossible. Compared to the conventional Rastelli procedure for TGA, VSD, and LVOTO, the technically challenging Nikaidoh procedure was superior in terms of reintervention rate for RVOT compression, LVOTO, aortic insufficiency, and midterm survival.[11]

For our patient who had sinus inversus, TGA, VSD, and LVOTO, any of the 3 procedures could be chosen for definite correction. However, the first and difficult part in choosing the procedure to be followed was the unusual anatomy of the situs inversus that was totally opposite to what the surgeon was used to managing. Therefore, complete preoperative imaging studies for comprehensive understanding of the anatomy were crucial. The second difficulty was in the intracardiac rerouting of LVOT to the aorta because of the small VSD and Rastelli type A defect with straddling of the tricuspid valve, as shown by echocardiography. Thus, the Rastelli and REV procedures were precluded. The patterns of coronary arteries of the patient were normal, but there was a communication branch of the LCA and RCA crossing the RVOT. Considering the anatomy, reintervention rate, and long-term survival potential, we chose the Nikaidoh procedure as the designated procedure for total correction. We paid special attention while the aortic root was being harvested to avoid injuring the coronary arteries.

We used the Nikaidoh procedure for total surgical correction in a patient with dextrocardia, situs inversus, TGA, VSD, and PS. Compared to the Rastelli and REV procedures, the Nikaidoh procedure can provide improved mid-term survival benefits[11] and lower reintervention rate. To our knowledge, this is the first report of this condition in Taiwan.

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