Atrial switch procedure for disease transposition of the great arteries come late at e hospital

Do Anh Tien[1,2], Luong Tuan Bao[1], Tran Hoang Ha[3], Nguyen Tran Thuy[1,2*]

SUMMARY

Background: Disease transposition of the great arteries is usually screened antenatally and operated arterial switch in the period before 2 weeks of age because without surgical intervention, 90% of children with dying before 6 months of age. However, there are still a small number of patients presenting late who are not suitable for arterial switch operation. Senning or Mustard procedures are the solutions to this situation.

Methods: We report two late cases of disease transposition of the great arteries that were diagnosed and operated on at the Pediatric Cardiovascular Surgery Department, Heart Center, E Hospital. Through these two cases, we discuss methods of diagnosis, treatment, and early results and review the medical literature on this disease.

Results: Two patients with late diagnosis: type D transposition of the great arteries were operated on by Senning and Mustard methods with good early results, improved clinical status, and no cyanosis. Postoperative ultrasonography did not detect narrowing of the blood tunnel. The electrocardiogram did not detect arrhythmias.

Conclusion: Senning or Mustard procedures give good results, are the solution for patients with late detection disease transposition of the great arteries and no longer indicated for arterial switch operation.

Keywords: *transposition of the great arteries; Senning; Mustard; atrial switch procedure.*

I. BACKGROUND

Disease transposition of the great arteries is the most common cyanotic congenital heart disease with incompatibility between the ventricles and the great arteries, specifically as follows: the aorta originates from the right ventricle, the pulmonary artery originates from the left ventricle and there is a compatibility between the atrium and ventricle[1] . The disease was first described by Mathew Bailie in 1979[2] . The aorta lies anterior to the pulmonary artery and can be either right (D-transposition) or left (L-transposition) or parallel. The pulmonary artery from the left ventricle will carry oxygen-rich blood to the lungs, while the aorta comes from the right ventricle, bringing oxygen-poor blood to the body, after perfusing the tissue will return to the venous system and to the right heart. Because there are two parallel rather than serial circulatory systems, respiratory failure and cyanosis often occur acutely shortly after birth. The child's survival depends on the mixing of blood between two circulations by accompanying lesions: ductus arteriosus, foramen ovale or ventricular septal defect. The disease accounts for about 5-7% of congenital heart diseases, the male:female ratio ranges from 1.5-3.2:1[3] .

[1] *E hospital;*
[2] *University of Medicine and Pharmacy, Vietnam National university Hanoi;*
[3] *Hanoi Medicine University*
**Corresponding author : Nguyen Tran Thuy,*
Email: drtranthuyvd@gmail.com
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Without treatment, 30% of babies die in the first week, 90% die in the first 6 months after bith[3] .

The current diagnosis of the disease is mainly based on clinical symptoms of severe cyanosis, Doppler echocardiography before and after birth. Straight chest X-ray, electrocardiogram have diagnostic value.

In 1959, Senning introduced a "physiologic correction" solution: performing surgery to repair of the right atrium wall and atrial septum to return the pulmonary vein to the right atrium, and the systemic vein to the left atrium [4] . In 1963, Mustard also introduced a method of "physiological correction" by widening the atrial septum, using a pericardial patch to redirect the blood flow of the systemic and pulmonary veins [5] . By 1975, Jatene with surgery to reposition the great arteries and replant the coronary arteries made a revolutionary turning point in the treatment of disease transposition of the great arteries [3] . Over the decades, some methods have been developed and improved to treat variations of disease transposition of the great arteries, such as the surgery of Rastelli, Lecompte, Nikaidoh, Hemi-Mustard, etc.

Today, good antenatal management has enabled early detection of most arterial inversion malformations from the fetal period [6] . Children can be operated by sugery of transpositing arteries before 2 months for patients with intact ventricular septum, before 3 months for patients with ventricular septal defect, depending on heart failure status [3] . However, the indication for surgical methods for children diagnosed late is still controversial and depends on the surgeon's experience, resuscitation ability, and postoperative care of each center [6] , including:

+ Fast 2-stage surgery (left ventricular training surgery followed by sugery of transpositing arteries)

+ Surgery to switch atrium

+ Sugery of transpositing arteries

We present two clinical cases of disease transposition of the great arteries late-diagnosed and operated on switching atrium by Senning and Mustard's method at Children's Cardiac Surgery Department - Cardiovascular Center, E Hospital to discuss the diagnosis, treatment, early patient outcomes, and looking back to the medical textbooks on this form of pulmonary venous abnormality.

II. CLINICAL CASES

Clinical case 1. Female child, 42 months, history: first child, normal delivery, full term, birth weight 3000g, no asphyxia after birth, no obvious cyanosis. After 1 month, the infant's cyanosis gradually increased, cyanosis increased when suckling, crying, poor feeding, poor weight gain. The family brought the child to be examined and then detected congenital heart disease: transposition of the great arteries. The child had a surgery to widen the atrial septum at 38 days old at Hue Central Hospital. After surgery, the child's condition improved. The child had been managed and monitored periodically, psychomotor development was normal. This time, children with increased cyanosis, fatigue, poor appetite was transferred to Heart Center – E Hospital. Examination: Child awake, A/AVPU, dyspnea Ross II-III, subcutaneous hemorrhage of various morphologies, multiple ages, purple lips and extremities, heartbeat 120 times/min regular, rapid breathing 50 times/min, lung no rales, SpO2 75%, abdomen soft, liver drooping ribs, fingers and toes were clubbed. After confirming that

there was a state of hemoconcentration, subcutaneous hemorrhage due to thrombocytopenia, the child was treated in stable medical treatment. The 1st and 2nd echocardiograms gave similar results: simple transposition of the great arteries type D, the atrial septum was widened and ventilated to 14mm in size, 2-way shunt, left ventricular posterior wall thickness was 7mm, ventricular size left was in the normal limit. However, because the patient's age was not suitable for artery transpositing surgery, we decided to perform Murstard surgery. The patient underwent thoracotomy along the mid-sternal line, de-adhesion, and an extracorporeal circulation machine with a catheter placed into the ascending aorta, superior vena cava, and inferior vena cava. Custodiol solution was used to paralyze the heart through the aortic root. The new septum was made of Edwards pericardial patch. Machine running time was 117 minutes, aortic pairing time was 86 minutes, supporting time was 18 minutes. Total surgery time was 280 minutes. Due to unstable hemodynamic status, the patient was left open sternum and closed after 1 day. After surgery, the patient was mechanically ventilated, actively resuscitated and extubated after 92 hours. The patient continued medical treatment and was discharged 27 days after surgery. Postoperative ultrasound and at the time of re-examination after 30 months gave good results: the left vena cava connection and the right pulmonary-ventricular vein were open. Electrocardiogram sinus rhythm, regular, right axis, no arrhythmia detected. The clinical condition after 30 months improved significantly compared to before surgery, the children no longer showed shortness of breath (Ross I) or cyanosis (SpO2 98%), participating in physical activities equivalent to children of the same age.

Clinical case 2. Male child, 4 months, history: 3rd child, normal delivery, full term, birth weight 3500g, no asphyxia after birth, no obvious cyanosis. Poor feeding, slow weight gain, gradually increasing cyanosis, rapid breathing, being examined to detect congenital heart disease, transferred to Heart Center – E Hospital Examination: Child awake, A/AVPU, dyspnea Ross II, mucosal skin much purple conjunctiva, rapid breathing, no rales, SpO2 55%, heartbeat 155 times/min, abdomen soft, liver drooping ribs. Ultrasound resulted: simple transposition of the arteries, type D, thickened right ventricle, pushed left interventricular septum, banana-shaped left ventricle, left ventricular posterior wall thickness was 3.9mm, left ventricular mass index 42g /m2, normal coronary artery origin, PFO hole size 5mm, 2-way shunt, ductus arteriosus closed mostly, low shunt flow. The patient was diagnosed with a late transposition of the ateries, and we decided to choose Senning surgery. The patient underwent thoracotomy along the mid-sternal line, and was run on an extracorporeal circulation machine with a cannula placed into the ascending aorta, superior vena cava, and inferior vena cava. Custodiol solution was used to paralyze the heart through the aortic root. The new septum is created by the atrial septum and the pericardium itself. Machine running time was 100 minutes, aortic pairing time was 81 minutes, supporting time was 10 minutes. Total surgery time was 180 minutes. Due to unstable hemodynamic status, the patient was left open sternum and closed after

2 days. After surgery, the patient was mechanically ventilated, actively resuscitated and extubated after 74 hours. The patient continued medical treatment and was discharged 18 days after surgery. Postoperative ultrasound and re-examination after 18 months showed good results: the left vena cava connection and the right pulmonary-ventricular vein were open. Right ventricle dilated, tricuspid valve moderately open. Electrocardiogram sinus rhythm, regular, right axis, no arrhythmia detected. Clinically, the patient improved well, did not have shortness of breath (Ross I) or cyanosis (SpO2 97%), fed well, gained weight.

III. DISCUSSION

From the early 1960s to the mid-1980s, Mustard and Senning operations were the treatment of choice for the disease transposition of the great arteries (d-TGA) [7] . Complications may occur after surgery such as: right ventricular failure, arrhythmia, tricuspid valve dysfunction, problems related to the septum and sudden death. According to P Moons and coworkes through a multicenter study [7] , patients in the Senning surgery group had a higher cumulative survival rate than patients in the Mustard surgery group; septal obstruction occurs more often after Mustard surgery; patients in the Senning group had better cardiac function and tended to participate in more sports activities. However, the operation in Senning surgery involves many complicated steps, causing more difficulties for the surgeon than in Mustard [5 surgery] .

Our decision to choose atrial conversion surgery depends on: (1) cardiac Doppler echocardiography, (2) age of the patient, (3) arterial lungs hypertension status due to associated ventricular septal defect, (4) complexity of coronary artery anatomy. On cardiac Doppler echocardiography, left ventricular posterior wall thickness less than 4mm, left ventricular septum deviation, left ventricular banana shape, left ventricular dysfunction, left ventricular mass index <35g/m2... are considered inappropriate. for arteriovenous surgery [6,8] . At that time, the author suggested that two-stage rapid surgery could be performed (left ventricular training 1-2 weeks then arterial transfer). However, after assessing the benefits and risks of rapid two-stage surgery and to suit the situation in our facility, we performed atrial conversion. The authors also indicated that the age for left ventricular training to be effective is before 3-6 months postpartum [6] .

In the 1st clinical case, the patient had typical presentation of late-onset cyanotic congenital heart disease with microcytic polycythemia vera, very high hematocrit, thrombocytopenic purpura, and survival due to mixed blood in the blood. tympanic layer. The child was diagnosed with simple arterial inversion and was no longer indicated for artery bypass surgery. Without early surgery, the clinical condition will become more and more severe and leave many consequences for the child's development, even death. We performed Mustard surgery after stable medical treatment. In the 2nd clinical case, the patient showed severe cyanosis due to the mixing of blood through the foramen PFO and the ductus arteriosus not being enough to meet the growing needs of the child, the ductus arteriosus tended to close. On echocardiography, the indicators showed no longer suitable for artery bypass surgery. We

performed Senning surgery. Leaving the sternum open after surgery is necessary in many cases. In young children, the heart, lungs, and soft tissues tend to be edematous after a prolonged period of cardiopulmonary bypass. The opening of the sternum in high-risk cases helps to stabilize the patient's hemodynamics before closing the sternum[8].

Close follow-up after surgery is essential. Because the right ventricle now plays the role of the systemic ventricle, the risk of right ventricular failure, tricuspid regurgitation due to annulus dilatation is a major obstacle in the future. Arrhythmias due to interventions in the atria and atrial septum, narrowing of the blood tunnel due to surgical techniques should be detected and treated promptly. In our cases, the patient did not experience any complications after surgery.

IV. CONCLUSION

Delayed disease of transposition of the great arteries is a rare congenital heart disease that is difficult to treat and carries a high risk of severe complications. Therefore, it is necessary to make a good prenatal diagnosis and closely monitor the child after surgery. Early Senning and Mustard procedures give good results.

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