# Clinical analysis of intracardiac diploe lateral-tunnel total cavopulmonary connection in the treatment of complex congenital heart diseases.

# Huang Zhongyao<sup>\*</sup>, Chen Daozhong, Qiu Zhihuang

Cardiovascular Surgery, Union Hospital Affiliated, Fujian Medical University, PR China

#### Abstract

Objective: To analyse the clinical efficacy and safety of intracardiac diploe lateral-tunnel total cavopulmonary connection in the treatment of complex congenital heart diseases.

Methods: A total of 32 children with complicated congenital heart disease in our hospital from January 2010 to January 2016 were enrolled, including 11 cases with tricuspid valve atresia, 15 cases with single functional ventricles, and 6 cases with correct transposition of great artery. Surgery was performed under moderate hypothermic cardiopulmonary bypass. The patients took supine position for thoracotomy, followed by the isolation of pulmonary artery and left and right arteries. The superior vena cava was cut off for the end-to-side anastomosis of right pulmonary artery, blocking the aortic from filling protective liquid. With the incision of right atrium and the resection of atrial septum, the defect of atrial septum was widened to 2~3 cm. The pericardium slice was used as intracardiac lateral tunnel with fenestration of 0.4~0.5 cm. The pulmonary aorta of proximal arterial was cut off, and anastomosis of distal arterial and superior vena cava was performed while proximal arterial received suture.

Results: The mean arterial pressure, oxygen saturation and urine output were significantly different after operation (P<0.05), while there was no significant difference in central venous pressure (P=0.183>0.05). Postoperative early death occurred in 2 cases (6.3%) due to low cardiac output and cardiac arrest. Postoperative complications occurred in 7 cases (21.8%), pulmonary edema in 2 cases (6.3%), pleural effusion in 3 cases (9.4%), pericardial effusion in 1 case (3.1%), and arrhythmia in 1 case (3.1%). The thirty patients were followed up for 6 to 36 months. Two patients (6.7%) had chest resuscitation and improved after treatment.

Conclusion: The intracardiac diploe lateral-tunnel total cavopulmonary connection achieved good results in the treatment of complex congenital heart diseases.

**Keywords:** Intracardiac diploe lateral tunnel, Total cavopulmonary connection, Complex congenital heart disease, Clinical analysis.

#### Introduction

Fontan procedure is an effective treatment for many high-risk complex intracardiac malformations. With the accumulation of medical experience, Fontan surgical techniques continue to improve, including Total Cavopulmonary Connection (TCPC) and right atrial to right ventricular connection [1]. Now the main operation of modified Fontan is divided into extracorporeal conduit TCPC and intracardiac lateral tunnel TCPC [2]. In early stage, such surgery on the children has stringent requirements of indications. However, with the development of surgical levels, surgical indications gradually expanded, such as age, pulmonary artery and aortic ratio and pulmonary vein ectopic drainage [3]. In the past TCPC surgery, atrial diploe mainly used man-made materials like Gore-tex without growth function and wide application. The application of self-pericardial patch as the atrial diploe material gives the tunnel the potential to grow and reduce major postoperative

complications of pleural effusion after Fontan [4,5]. At present, this operation has been widely used in the hospital at home and abroad. Here, we reported the 32 cases of such surgery for children with complex congenital heart disease in our hospital for nearly six years.

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## **Materials and Methods**

#### Clinical data

Ethical approval was given by the medical ethics committee of Union Hospital Affiliated, Fujian Medical University with the following reference number: 2009022. The observation cases were 32 patients with complicated congenital heart disease in our hospital from January 2010 to January 2016, including 18 males and 14 females, aged 1.8 to 9.5 y with mean age of (4.2  $\pm$  3.9 y), weight range of 8.5 kg~38.8 kg and average weight of

 $(15.4 \pm 4.9 \text{ kg})$ . Patients showed cyanosis of lips and the end of limbs. After exercise, palpitations, shortness of breath and chest tightness occurred. IV grade systolic murmur could be heard at Rib 2 to 4 in sternum, and oxygen saturation were <70%. Children underwent electrocardiogram, echocardiography, thoracotomy and CT before operation, among which 12 cases additionally received cardiovascular angiography. Diagnosis results indicated that there were 11 cases with tricuspid valve atresia, 15 cases with functional single ventricle, and 6 cases with corrected transposition of the great arteries. All patients had well-developed branches of pulmonary artery and pulmonary artery, but Atrial Septal Defect (ASD) and Right Ventricular Outflow Tract Stenosis (RVOTS) that causing the difficulty for the implementation of ventricular repair. The data of other signs were as follows: pulmonary artery index (277.52  $\pm$  51.54 mm<sup>2</sup>/m<sup>2</sup>), McGoon index 1.8~2.5, mean pulmonary arterial pressure<18 mmHg. pulmonary vascular resistance<4 Wood units/m<sup>2</sup>, hemoglobin 151~224 g/L.

#### Surgical methods

All patients underwent surgery under mild or moderate hypothermic cardiopulmonary bypass. With nasal or oral tracheal intubation, intravenous access was established for general anesthesia. Patients with supine position received median sternotomy incision and resection of both thymuses. After fully exposure and isolation of pulmonary artery, left and right pulmonary arteries were isolated to hilus pulmonis, superior vena cava to the left and right innominate vein connection and inferior vena cava to the diaphragm. Arterial duct or ligament was ligated, followed by the ligation of collateral vessels. Aortic perfusion tube was placed into conventional ascending aorta, and the superior vena cava was traversed at 1cm away from the right atrium. Parallel incision 1.0~2.5 cm was performed at the right upper pulmonary artery and arterial. Then the distal end of superior vena cava and right pulmonary artery incision received end-to-side anastomosis with 5-0 Prolene absorbable suture. Besides, continuous suture of posterior wall and interrupted suture of anterior wall were performed with caution to distortion. After blocking the aorta, cold myocardium protection liquid was infused. With the right atrial incision and resection of the atrial septum, selfpericardial patch was taken for lateral tunnel of intracardiac diploe, followed by the link of upper and inferior vena cava. Then the suture from the middle of atrial septum periphery and the suture from lateral incision of coronary sinus were performed, bypassing the inferior vena cava incision to the beginning point. Conventional fenestration of intracardiac diploe window was 0.4~0.5 cm and pulmonary artery was cut off at the beginning of proximal arterial valve. Distal arterial and proximal ends of superior vena cava were sutured with 5-0 Prolene absorbable line anastomosis with caution to distortions, using pericardial patch to extend anastomosis if necessary. Proximal end used 5-0 Prolene absorbable line for the suture of terminal. Postoperative patients took position of upper body 45°, lower body 30°, with moderate fluid infusion. Meanwhile, patients received intravenous dopamine 2~10 µg/

(min•kg), sodium nitroprusside  $0.5\sim1.5 \ \mu g/(min•kg)$ , epinephrine  $0.1\sim0.30 \ \mu g/(min•kg)$ . Dosage was reduced according to the condition to control the central venous pressure at  $12\sim18$  mmHg. With assisted mechanical ventilator, arterial blood CO<sub>2</sub> partial pressure was maintained at  $30\sim35$ mmHg. Besides, blood pressure, ECG, oxygen saturation and chest drainage were monitored. When the condition was stable, ventilator was stopped and the tracheal intubation was removed.

# Indicators and efficacy evaluation

The clinical indicators before and after surgery including hemodynamics, oxygen saturation, breathing and other indicators were recorded. According to the changes in indicators, the efficacy evaluation criteria were established.

**Marked:** The clinical symptoms disappeared; postoperative CT and ultrasound examination had no vascular atresia or stenosis; breathing and hemodynamic indicators tended to normal; no complications occurred. Effective: postoperative patients had remission of clinical symptoms; postoperative CT and ultrasound examination had no vascular atresia or stenosis; breathing and hemodynamic indicators improved; fewer complications occurred. Invalid: the clinical symptoms of patients after surgery did not ease; postoperative CT and ultrasound examination had vascular abnormalities; breathing and hemodynamic indicators did not improve; serious complications occurred.

#### Statistical methods

The data were analysed by SPSS 18.0. Measurement data was recorded as mean  $\pm$  standard deviation, and t-test was applied with P<0.05 as significant statistical difference.

## Results

#### Operation comparison of the two groups

There were significant differences in the mean arterial pressure, oxygen saturation and urine output between the two groups (P<0.05), but no significant difference showed in central venous pressure (P>0.05, Table 1).

Table 1. Comparison between the stud	<i>ly group and the control group.</i>
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Groups	Mean arterial pressure (mmHg)	Oxygen saturation (%)	Central venous pressure (mmHg)	Urine output (ml/kg•h)
Before	50.4 ± 4.3	75.3 ± 2.5	15.8 ± 1.1	1.1 ± 0.3
After	75.7 ± 5.2	90.2 ± 2.3	15.4 ± 1.2	3.7 ± 1.0
t	-20.537	-24.024	1.346	-13.640

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Р	<0.001	<0.001	0.183	<0.001

## Follow-up of patients

After surgery of 32 patients, there were 14 marked cases, 16 effective cases and 2 invalid cases. The total effective rate was 93.8%. For the postoperative early death in 2 cases (6.3%), one was caused by low cardiac output and another by cardiac arrest. Postoperative complications occurred in 7 cases (21.8%), including pulmonary edema in 2 cases (6.3%), pleural effusion in 3 cases (9.4%), pericardial effusion in 1 case (3.1%) and arrhythmia in 1 case (3.1%). The above symptoms were improved after symptomatic treatment. During 6 to 39 months of follow-up for 30 patients, no late death occurred. Two patients (6.7%) had chest resuscitation and improved after treatment.

## Discussion

Functional single ventricle heart disease is a kind of complicated congenital heart disease due to unilateral ventricle unable to conduct physiological function and biventricular repair difficult to implement [6]. In the 1970s, Fontan surgery was applied to the treatment of heart disease with the principle to isolate the lung and body circulation from the parallel state into a series state. The separation of the venous blood and pulmonary venous blood reduces pulmonary vascular resistance and improves the venous pressure. Venous blood flows into the pulmonary circulation, eliminating unilateral ventricular overload status and improving hypoxia condition [7]. Fontan surgery as a physiological means of treatment is the best way to maintain patients with congenital heart disease keeping normal blood circulation. In early stage, such surgery on the children has stringent requirements of indications. However, with the development of surgical levels, surgical indication requirements gradually expanded, such as age, pulmonary artery and aortic ratio and pulmonary vein ectopic drainage. But pulmonary artery development, pulmonary vascular resistance and pulmonary artery pressure still need to meet requirements. The in-depth study of hemodynamics found that in the linear flow of blood of blood vessels, right atrial and pulmonary artery anastomotic corners have great corner resistance and energy consumption. Therefore, the original surgery was modified to become TCPC [8]. As the most successful modified Fontan, TCPC can provide better blood flow. The suture of right atrium away from the atrioventricular node avoids the conduction of the block, reduces heart rate disorders, and eliminates unilateral ventricular overload. Meanwhile, it can increase pulmonary artery forward blood flow, improve hemodynamic parameters, and ease the cyanosis and other symptoms caused by chronic hypoxia [9,10].

Main clinical procedures of TCPC were extracardiac vein TCPC and intracardiac lateral tunnel TCPC. With own characteristics, clinical choice of the two procedures depends mainly on the experience of doctors or hospitals. Studies have suggested that compared with extracardiac tunnel, intracardiac lateral tunnel TCPC can be applied to younger patients, as it can reduce the loss of blood flow energy and promote the inferior vena cava into the lung cycle [11]. In the past TCPC operation, the intracardiac diploe is mainly composed of Goretex and other man-made materials which are easy to form thrombosis and need long-term anticoagulation. All kinds of problems caused by anticoagulation therapy seriously threaten patients' life. Meanwhile, artificial materials easily lead to infection and endocarditis. Without growth function, the application was limited in the younger patients. In this study, we used self-pericardium patch as a diploe with growth potential for young patients. And the use of autologous material had low risk for thrombosis without long-term anticoagulant therapy. The study pointed out that atrial diploe fenestration can reduce the mortality of Fontan patients. By reducing the venous pressure to reduce interstitial fluid and lymphatic exudation, Fontan reduced the pleural effusion and shortened the hospital stay [12-14]. Therefore, in this study, we performed atrial diploe fenestration with diameter of 0.4~0.5 cm. In this study, mean arterial pressure, blood oxygen saturation and urine output before and after surgery were significantly improved, indicating that this surgery could reduce the burden on the heart and improve hypoxia. The total effective rate after surgery was 93.8% with an ideal effect. Postoperative complications occurred in 7 cases (21.8%), including pulmonary edema in 2 cases (6.3%), pleural effusion in 3 cases (9.4%), pericardial effusion in 1 case (3.1%) and arrhythmia in 1 case (3.1%). Application of vasoactive drugs, supplements, regulating heart rhythm drugs, cardiac drugs, diuretic, chest drainage and other methods of symptomatic treatment cured or improved these symptoms. Due to cardiopulmonary bypass replacing the blocked aorta, TCPC surgery may damage the myocardial cells and capillary endothelium. Increased postoperative venous pressure exacerbates this injury, easily leading to pleural effusion [15-17]. Although conventional atrial diploe fenestration expenses part of the oxygen saturation, it can increase the amount of cardiac output to maintain early postoperative hemodynamic stability and reduce venous pressure. From the results of this study, it seems useful. From the results of follow-up, the fenestration aperture gradually becomes smaller as time, while blood oxygen saturation will rise. Two years after surgery, half of apertures can be closed naturally. If 2 y follow-up found that oxygen saturation continued low, patients need to block heart catheter. In this study, autologous pericardium patch was used to construct intracardiac diploe lateral tunnel without long-term anticoagulation after operation. No thrombosis occurred during follow-up. The pericardial growth was not determined in this study due to shorter follow-up times.

In summary, intracardiac diploe lateral-tunnel TCPC has good effect in the treatment of complex congenital heart disease. And the diploe fenestration can reduce the occurrence of pleural effusion and shorten the hospital stay. Pericardium patch prevents thrombosis as diploe tunnel material, and it is an ideal repair material of TCPC surgery.

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#### \*Correspondence to

Huang Zhongyao

Cardiovascular Surgery

Union Hospital Affiliated

Fujian Medical University

PR China