

# Traditional Atriopulmonary Connection Fontan with Excellent Outcome for 32 Years

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Thorac Cardiovasc Surg Rep 2020;9:e18–e20.

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## Abstract

### Keywords

- ▶ cardiovascular surgery
- ▶ congenital heart disease
- ▶ coronary heart disease
- ▶ computed tomography
- ▶ computed tomography scan

Although the traditional right atrial–pulmonary artery (RA–PA) Fontan has been replaced by the total cavopulmonary connection, some RA–PA Fontan patients still have good outcome. We report a 37-year-old male who underwent traditional Fontan 32 years ago with the diagnosis of L-transposition of great arteries, subvalvular pulmonary stenosis and cardiac dextroversion. Among the recent CTA, electroconvulsive therapy, catheterization and angiography, pulsatile PA, uniform pulmonary blood, normal PA pressure and resistance were observed in this patient. The traditional RA–PA Fontan may be an alternative surgical procedure.

## Introduction

The traditional atriopulmonary connection (APC) has been used in clinical practice for functional single ventricle (FSV) by Fontan et al since 1971.<sup>1</sup> However, de Leval et al. found that pulsation of the right atrium (RA) could generate turbulence and increase the energy loss,<sup>2</sup> total cavopulmonary connection (TCPC) has become the main procedure for FSV. Should APC be eliminated? We report a 32-year follow-up case with excellent outcome.

In 1987, a 5-year-old boy was admitted to our department with severe cyanosis and diagnosed as with tricuspid atresia with L-transposition of great arteries, subvalvular pulmonary stenosis, and cardiac dextroversion. Preoperative oxygen saturation of the femoral artery was 85.4%. Subsequently, APC was performed. Postoperatively, the cyanosis disappeared and oxygen saturation was improved (97.4%). During a follow-up of 32 years, he had paroxysmal supraventricular tachycardia and with a cardiac function of New York Heart Association. In addition, he had been married for 10 years and had a healthy son.

After a 32-year follow-up, cardiac computed tomography angiogram (CTA), catheterization, angiography, and emission computed tomography (ECT) were performed recently.

CTA (▶**Fig. 1**) shows that the pulmonary artery (PA) trunk has been anastomosed with RA, while there is no stenosis between RA and PA.

Catheterization (▶**Table 1**) shows that the PA pressure (12.5 mm Hg) and the whole pulmonary vascular resistance (2.21 wood) are normal, the pulse pressure difference is approximately 7 to 9 mm Hg (left 20/11 mm Hg and right 18/11 mmHg), while the vena cava pressure is just a little higher (superior 19/10 mm Hg and inferior 18/11 mm Hg).

Angiography (▶**Fig. 2** and ▶**Video 1**) shows that the blood flow from superior vena cava (SVC) and IVC (inferior vena cava) is mixed in the large RA thoroughly and then distributes to left and right PA uniformly, which accompanied by pulsation of bilateral PAs. Electroconvulsive therapy (▶**Fig. 3**) quantitatively shows that the blood flow distributes to left (53.18%) and right (46.82) PA uniformly (▶**Table 2**).

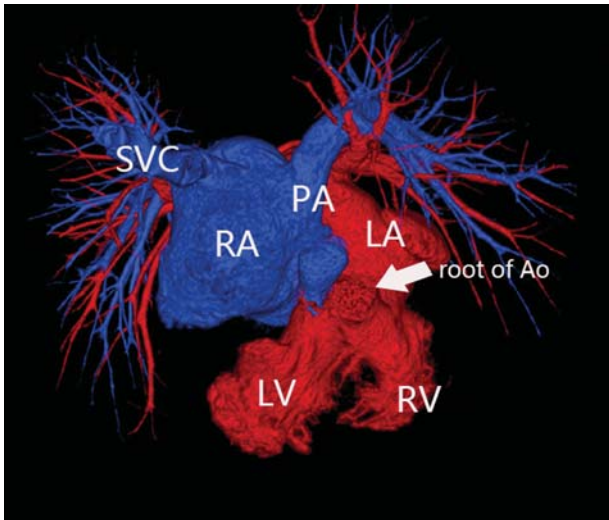
received  
June 7, 2019  
accepted  
December 6, 2019

DOI <https://doi.org/10.1055/s-0040-1708528>.  
ISSN 2194-7635.

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**Fig. 1** Postoperative cardiac computed tomography angiogram. Cardiac dextroversion, Ao arises from the rudimentary RV; Ao is anterior and to the left of the pulmonary trunk (L-transposition); pulmonary trunk has been ligated above pulmonary valve. There is no stenosis in the anastomotic orifice between the RA and the pulmonary trunk. Ao, aorta; LA, left atrium; LV, left ventricle; PA, pulmonary artery; RA, right atrium; RV, right ventricle; SVC, superior vena cava.



**Fig. 2** Postoperative angiography.



**Fig. 3** Postoperative electroconvulsive therapy. Blood flow was evenly distributed in both lungs postoperative angiography.

**Table 1** Postoperative cardiac catheter data

	Oxygen saturation (%)	Pressure (mm Hg)	Resistance (Dyn.s.cm <sup>-5</sup> )
SVC	74.7	19/10 (13)	
IVC	84.7	18/11 (12)	
L-PA	82.9	20/11 (13)	
R-PA	78.6	18/11 (12)	
Residual PA-trunk		12.5	
Whole pulmonary			176.87 (2.21 wood)
Femoral artery	97.4		

Abbreviations: IVC, inferior vena cava; L-PA, left pulmonary artery; PA, pulmonary artery; R-PA, right pulmonary artery; SVC, superior vena cava.

**Table 2** Postoperative emission computed tomography

	Left lung (%)	Right lung (%)
Upper	12.95	8.96
Middle	25.01	22.76
Lower	15.22	15.11
Total	53.18	46.82

**Video 1**

The blood flow from SVC and IVC is mixed in the large RA thoroughly and then distributes to left and right PA uniformly, which accompanied by pulsation of bilateral PAs. IVC, inferior vena cava; PA, pulmonary artery; RA, right atrium; SVC, superior vena cava. Online content including video sequences viewable at: <https://www.thieme-connect.com/products/ejournals/html/10.1055/s-0040-1708528>.

**Discussion**

In 1988, de Leval et al<sup>2</sup> reported a hydrodynamic study regarding the traditional Fontan procedure (APC) and found that pulsation of the RA could generate turbulence, which hindered the forward flow and increased the energy loss, especially around corners.

However, this patient just has performed paroxysmal supraventricular tachycardia without other complications after the APC for 32 years. In addition, oxygen saturation

and left ventricle function are normal, while the vena cava pressure is slightly elevated. The patient was treated with oral metoprolol, regular reexamination of electrocardiogram. The radiofrequency ablation is planned to be performed for arrhythmia in the future.

For this patient, we find that the special anatomical structure (L-transposition and dextroversion) makes the left ventricle compress the hypertrophic RA to form a powerful pump, thereby promoting the systemic venous blood into the PA. Then rhythmic heartbeat brings pulsating pulmonary blood flow, which helps maintain normal pulmonary vascular resistance. On the other hand, the blood from IVC and SVC is fully mixed in RA; the blood of hepatic vein can distribute evenly to both pulmonary arteries, thus reducing the occurrence of pulmonary arteriovenous fistula.

The traditional APC has gradually been replaced by the TCPC.<sup>3,4</sup> However, should the APC be eliminated? Although RA could generate turbulence and increase the energy loss, the pulsation of RA also provides energy for PA flow. Therefore, the traditional RA-PA Fontan may be an alternative

surgical procedure for some special patients because of the good outcome.

#### Funding

None.

#### Conflict of Interest

No conflict of interest exists in the submission of this manuscript.

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