

Original Article



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Echocardiography

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Pulsatility Index in Different Modifications of Fontan Palliation: An Echocardiographic Assessment

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ABSTRACT


BACKGROUND: Adding pulsation to the Fontan circulation might change the fate of patients palliated by this procedure. Our aim was to compare the pulsatility index (PI) of the pulmonary artery (PA) between the various modifications of Fontan palliation.

METHODS: Doppler-derived PI was measured in PA branches of a cohort of 28 patients palliated by 6 modifications of Fontan procedure. A group of normal individuals was included for comparison.

RESULTS: Atriopulmonary connection (APC) group had the highest PA branches PI and statistically was close to the PI of the normal individuals (right pulmonary artery [RPA] PI of 1.58 vs. 1.63; $p = 0.99$ and left pulmonary artery [LPA] PI of 1.54 vs. 1.68; $p = 0.46$, respectively). The lowest PA branches PI was seen in the group of extracardiac total cavopulmonary connection (RPA PI of 0.62 and LPA PI of 0.65). Other 4 modifications including the extracardiac conduit with oversewn pulmonary valve, extracardiac conduit with preserved adjusted antegrade flow, extracardiac conduit from inferior vena cava onto the rudimentary right ventricle and lateral tunnel had a mean "RPA and LPA" PI of "1.19 and 1.17", "1.16 and 1.11", "1.13 and 1.11", "0.82 and 0.84", respectively. The modified Dunnett's *post hoc* test has shown a significant statistical decline in PI of all modifications compared to the normal individuals except for the APC group.

CONCLUSIONS: Fontan palliated patients in different groups of surgical modification showed a spectrum of Doppler-derived PI with the highest amounts belong to the groups of pulsatile Fontan.

Keywords: Echocardiography; Flow pattern; Pulmonary artery; Univentricular heart

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The authors have no financial conflicts of interest.

Author Contributions

Conceptualization: Shabanian R, Navabi MA; Data curation: Sedaghat A, Esmaeeli Z, Ahani M; Formal analysis: Rahimzadeh M; Investigation: Shabanian R, Nikdoost A, Ahani M; Methodology: Dadkhah M, Rahimzadeh M; Supervision: Navabi MA; Validation: Shabanian R, Dehestani A; Writing - original draft: Shabanian R, Akbari Asbagh P, Sedaghat A, Dadkhah M; Writing - review & editing: Shabanian R, Dehestani A.

INTRODUCTION

It has been claimed that nonpulsatile flow in Fontan circulation is a drawback for this kind of palliation.^{1,2)} Lack of pulsatility may lead to a decrease in the diameter of the pulmonary arteries, which increases pulmonary vascular resistance (PVR).³⁻⁵⁾ It is a well-known fact that the low PVR has a vital role in Fontan physiology.⁶⁾ Hence, adding pulse to Fontan circulation was an important concern since the introduction of this type of surgery. Moreover, the evolved modifications of Fontan palliation might have a different index of pulsatility. Here, we compared the echocardiographic index of pulsatility among our cohort of patients palliated by different modifications of Fontan circulation.

METHODS

In our cohort of Fontan subjects, we found 6 different modifications of Fontan palliation including the 3 well known modifications of atriopulmonary connection (APC), lateral tunnel (LT) and extracardiac total cavopulmonary connection (ECTCPC). Three more additional modifications of extracardiac conduit with preserved adjusted antegrade flow (ECPAF), extracardiac conduit with oversewn pulmonary valve (ECOPV) and extracardiac conduit from inferior vena cava onto the rudimentary right ventricle (ECIRC) were also enrolled into the study. The schematic illustrations of these modifications are shown in **Figure 1**. The last 3 modifications are composed of a limited group of our patients with ECTCPC that was tried to add pulsation to their circulation. In 4 patients in the group of ECTCPC with adjusted

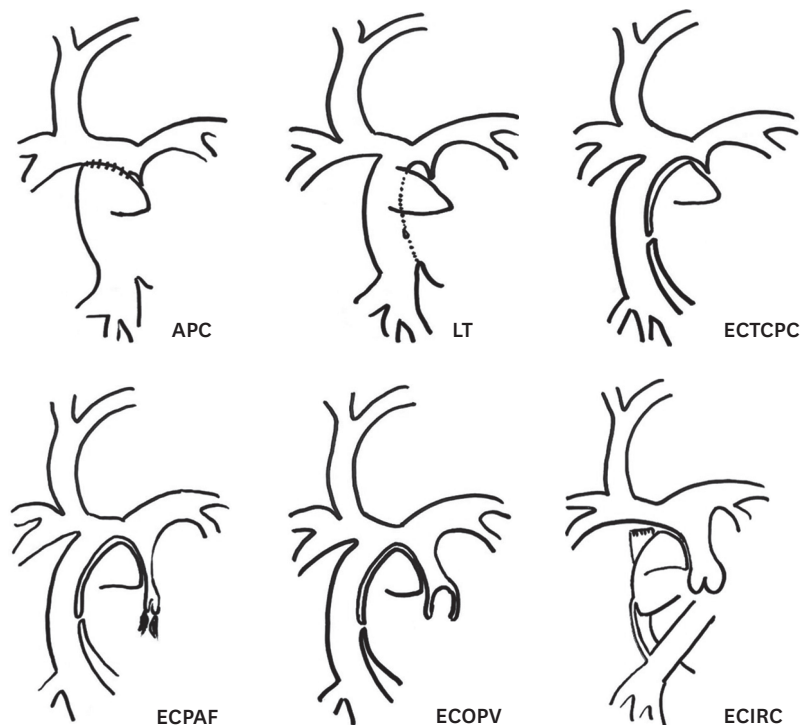


Figure 1. Schematic surgical illustration of 6 types of Fontan palliation in our cohort including APC, LT, ECTCPC, ECPAF, ECOPV and ECIRC.

APC: atriopulmonary connection, LT: lateral tunnel, ECTCPC: extracardiac total cavopulmonary connection, ECPAF: extracardiac conduit with preserved adjusted antegrade flow, ECOPV: extracardiac conduit with oversewn pulmonary valve, ECIRC: extracardiac conduit from inferior vena cava to right ventricular outflow tract.

antegrade flow, the restrictive antegrade flow through the main pulmonary artery (PA) has been further adjusted or preserved. The well-developed pulmonary valve was oversewn in 8 patients in the group of ECOPV to act as a membrane with forward and backward movements (**Supplementary Video 1**). The forward movement of the oversewn valve will propel the line of blood into the distal PA branches without adding a volume load to the pathway. In the group of ECIRC, 2 patients with tricuspid atresia type Ia who had a well-developed right ventricular outflow tract (RVOT) and pulmonary valve, the Fontan conduit was connected directly onto the right ventricle (RV) body. The contraction of the rudimentary RV will add pulsatility to the pulmonary circulation (**Supplementary Video 2**).

Patients included should have normal sinus rhythm and be in the class I New York Heart Association (NYHA) functional classification. Exclusion criteria were systolic dysfunction as measured by fractional area change (FAC) of less than 40%, PA branch stenosis and suboptimal Doppler flow tracing for measurements. Pulsed Doppler tracing of the vessels during quiet respiration with the patient in the supine position was digitized with envelope tracing to measure the pulsatility index (PI). The technique was described in detail in our previous study published in 2015.⁷⁾ By assuming that the vessels diameter is constant throughout the cardiac cycle, PI was calculated as maximum velocity minus minimum velocity divided by mean velocity.²⁾⁸⁾ All echocardiographic measurements were obtained by only one experienced pediatric cardiologist in Fontan field.

We used SPSS software version 26 (SPSS Inc., Chicago, IL, USA) for statistical analysis. Categorical data were expressed as count or percentage. The normally distributed data were reported as mean \pm standard deviation. The Robust Brown-Forsythe test (a modified one-way analysis of variance [ANOVA] test) was used for comparison of means between the groups because homogeneity of variances was not met. When the significance of modified ANOVA test was less than 0.05, the Dunnett's *post hoc* test was used to compare the treatment groups with the control group. The research protocol was approved by our center institutional ethics committee. Informed consent was taken from the patients or their parents.

RESULTS

Twenty-eight patients (10 males and 18 females aged 4 to 33 years) who underwent different modifications of Fontan palliation were enrolled into the study and compared with 6 normal individuals (2 males and 4 females aged 5 to 30 years). Patients' demographic characteristics and a few preoperative parameters such as PVR, mean of pulmonary artery pressure (PAP), atrioventricular valve regurgitation and ventricular function that could affect the PI are summarized in **Table 1**. It also shows in detail the anatomical basis of congenital heart disease in 6 groups of Fontan modification. Using Dunnett's *post hoc* test, the mean and standard deviation of the weight, calculated Nakata index, PAP, PVR and FAC showed no significant difference among the 6 groups (**Table 1**). The groups could not be matched in all baseline characteristics because of the patient paucity in a few groups of the study. For example, we could find only 2 patients palliated by APC modification in our Fontan cohort because of the obsolescence and conversion of the APC procedure to the ECTCPC type. All patients were in class I NYHA functional classification and had normal sinus rhythm with favorable systolic function. Moreover, the mean of PAP and PVR were less than 15 mmHg and 3 Wood units.m² respectively in any patients who had catheterization after the Fontan completion (not all the patients).

Table 1. Demographic characteristics of patient with different modifications of Fontan

Modification	Congenital heart disease	Age (years)	p*	Weight (kg)	p*	Time since Fontan (years)	p*	Nakata index (mm ² /m ²)	p*	Mean of PAP (mmHg)	p*	PVR index (WU/m ²)	p*	FAC (%)	p*	AVVR
APC (n = 2)	Tricuspid atresia (n = 2)	32.0 ± 1.4	0.003	55.0 ± 7.1	0.860	21.0 ± 7.1	0.004	307.0 ± 25.9	1.000	10.0 ± 0.7	0.870	1.10 ± 0.14	0.530	48.0 ± 7.0	0.840	Trivial
ECOPV (n = 8)	Tricuspid atresia (n = 3) DORV, non-committed VSD, double discordance (n = 3) DILV, double discordance (n = 2)	10.0 ± 3.3	0.760	34.0 ± 10.8	0.890	5.0 ± 2.4	0.590	308.0 ± 69.1	1.000	12.0 ± 1.8	1.000	1.50 ± 0.44	0.960	44.0 ± 7.9	0.990	Trivial to mild
ECPAF (n = 4)	Tricuspid atresia (n = 3) DILV, double discordance (n = 1)	13.0 ± 8.7	1.000	50.0 ± 25.6	0.940	9.0 ± 3.8	1.000	267.0 ± 102.7	0.920	13.0 ± 2.4	0.990	1.70 ± 0.48	1.000	43.0 ± 12.7	1.000	Trivial to mild
ECIRC (n = 2)	Tricuspid atresia (n = 2)	7.0 ± 0.7	0.550	24.0 ± 3.6	0.710	5.0 ± 0.7	0.810	246.0 ± 97.0	0.880	11.0 ± 1.4	0.870	1.20 ± 0.21	0.770	40.0 ± 1.1	0.990	Trivial
LT (n = 2)	Tricuspid atresia (n = 2)	24.0 ± 0.7	0.110	51.0 ± 4.2	0.960	19.0 ± 2.8	0.020	235.0 ± 111.2	0.810	11.0 ± 2.1	0.990	1.30 ± 0.35	0.770	43.0 ± 2.8	1.000	Trivial to mild
ECTCPC (n = 10)	Tricuspid atresia (n = 2) DORV, non-committed VSD, double discordance (n = 2) DILV, double discordance (n = 3) CAVSD unbalanced (n = 2) Pulmonary atresia, intact ventricular septum (n = 1)	13.0 ± 7.5		41.0 ± 23.1		8.0 ± 5.6		314.0 ± 115.9		12.0 ± 2.3		1.70 ± 0.66		42.0 ± 4.5		Trivial to mild

Data are reported as mean ± standard deviation. Values in bold indicate significant p-value.

APC: atrioventricular connection, ECOPV: extracardiac conduit with oversewn pulmonary valve, ECPAF: extracardiac conduit with preserved adjusted antegrade flow, ECIRC: extracardiac conduit from inferior vena cava to right ventricular outflow tract, LT: lateral tunnel, ECTCPC: extracardiac total cavopulmonary connection, DORV: double outlet right ventricle, VSD: ventricular septal defect, DILV: double inlet left ventricle, AVSD: atrioventricular septal defect, CAVSD: complete atrioventricular septal defect, PAP: pulmonary artery pressure, PVR: pulmonary vascular resistance, FAC: fractional area change, AVVR: atrioventricular valve regurgitation.

*Comparison of variables between different Fontan groups and ECTCPC group using Dunnett's post hoc test.

Doppler flow velocities of maximum, minimum and mean in addition to PI of PA branches are shown in **Table 2**. Compared to the normal group, all 6 groups of Fontan modifications had lower peak velocities and greater minimum velocities of Doppler flow. **Figure 2** shows the pattern of pulsed Doppler flow in the 6 studied modifications. Compared to the normal group, the PI of both PA branches were significantly lower in all modifications of Fontan except for the APC group ($p < 0.001$): APC RPA PI = 1.58 vs. normal right pulmonary artery (RPA) PI = 1.63, $p = 0.99$ and APC LPA PI = 1.54 vs. normal LPA PI = 1.68, $p = 0.46$ (**Table 2**, **Figures 3 and 4**). Furthermore, the ECTCPC group had the lowest PI in both PA branches

Table 2. Comparison of velocities and PIs of PA branches flows between 6 modifications of Fontan and normal groups

Pathway	Fontan modifications	Number	V max (cm/s)	p*	V min (cm/s)	p*	V mean (cm/s)	p*	PI	p*
RPA	APC	2	63.00 ± 5.66	0.018	13.00 ± 2.83	0.012	31.50 ± 0.70	0.005	1.58 ± 0.05	0.990
	ECOPV	8	63.32 ± 9.49	< 0.001	17.75 ± 1.47	< 0.001	38.31 ± 7.42	0.001	1.19 ± 0.10	< 0.001
	ECPAF	4	72.75 ± 9.03	0.026	20.00 ± 4.08	< 0.001	45.62 ± 6.88	0.064	1.16 ± 0.04	< 0.001
	ECIRC	2	50.00 ± 12.73	0.001	13.50 ± 4.95	0.009	32.65 ± 9.54	0.007	1.13 ± 0.09	< 0.001
	LT	2	47.50 ± 3.53	0.001	22.00 ± 1.41	< 0.001	30.95 ± 1.34	0.004	0.82 ± 0.03	< 0.001
	ECTCPC	10	46.30 ± 14.01	< 0.001	24.03 ± 7.45	< 0.001	35.22 ± 10.48	< 0.001	0.62 ± 0.10	< 0.001
	Normal	6	101.77 ± 24.55		00.00 ± 00.00		63.82 ± 16.83		1.63 ± 0.10	
LPA	APC	2	49.00 ± 1.41	0.001	13.25 ± 1.06	0.002	23.10 ± 0.42	< 0.001	1.54 ± 0.05	0.456
	ECOPV	8	48.45 ± 11.05	< 0.001	14.94 ± 4.31	< 0.001	28.84 ± 6.53	< 0.001	1.17 ± 0.09	< 0.001
	ECPAF	4	65.12 ± 19.72	0.001	18.90 ± 3.99	< 0.001	41.15 ± 13.32	0.004	1.11 ± 0.08	< 0.001
	ECIRC	2	47.85 ± 5.44	< 0.001	13.75 ± 2.47	0.002	30.55 ± 4.03	0.003	1.11 ± 0.05	< 0.001
	LT	2	46.90 ± 7.21	< 0.001	20.25 ± 4.31	< 0.001	31.50 ± 2.12	0.003	0.84 ± 0.03	< 0.001
	ECTCPC	10	40.18 ± 12.05	< 0.001	20.84 ± 5.17	< 0.001	24.27 ± 8.60	< 0.001	0.65 ± 0.10	< 0.001
	Normal	6	123.25 ± 38.21		00.00 ± 00.00		73.15 ± 24.87		1.68 ± 0.16	

Values in bold indicate significant p-value.

APC: atriopulmonary connection, ECIRC: extracardiac conduit from inferior vena cava to right ventricular outflow tract, ECOPV: extracardiac conduit with oversewn pulmonary valve, ECPAF: extracardiac conduit with preserved adjusted antegrade flow, ECTCPC: extracardiac total cavopulmonary connection, LT: lateral tunnel, LPA: left pulmonary artery, PI: pulsatility index, RPA: right pulmonary artery, PA: pulmonary artery, V: velocity.

*Each parameter was compared to the relevant normal group parameter with the Dunnett's *post hoc* test.

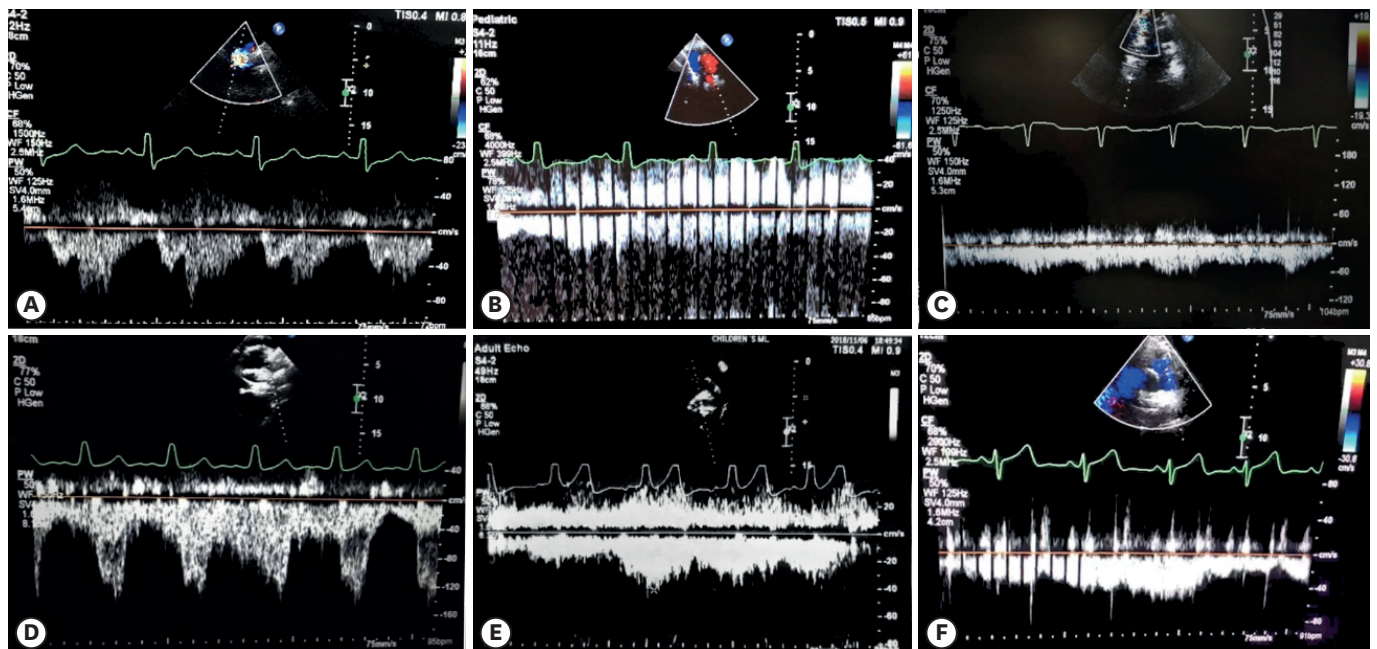


Figure 2. Examples of Doppler flow patterns in PA branches of 6 modifications of Fontan. (A) APC, (B) LT, (C) ECTCPC, (D) ECPAF, (E) ECOPV, (F) ECIRC. APC: atriopulmonary connection, LT: lateral tunnel, ECTCPC: extracardiac total cavopulmonary connection, ECPAF: extracardiac conduit with preserved adjusted antegrade flow, ECOPV: extracardiac conduit with oversewn pulmonary valve, ECIRC: extracardiac conduit from inferior vena cava to right ventricular outflow tract, PA: pulmonary artery.

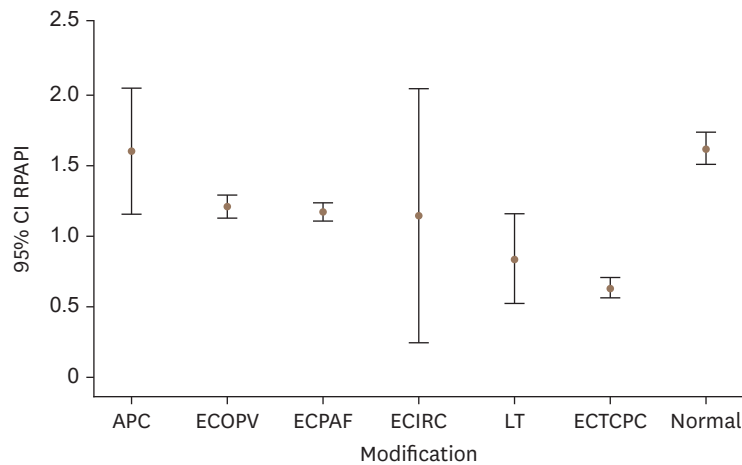


Figure 3. Comparison of mean RPA PI in 6 Fontan modifications. Error bar represents 95% CI. RPA: right pulmonary artery, PI: pulsatility index, CI: confidence interval, APC: atriopulmonary connection, ECOPV: extracardiac conduit with oversewn pulmonary valve, ECPAF: extracardiac conduit with preserved adjusted antegrade flow, ECIRC: extracardiac conduit from inferior vena cava to right ventricular outflow tract, LT: lateral tunnel, ECTCPC: extracardiac total cavopulmonary connection.

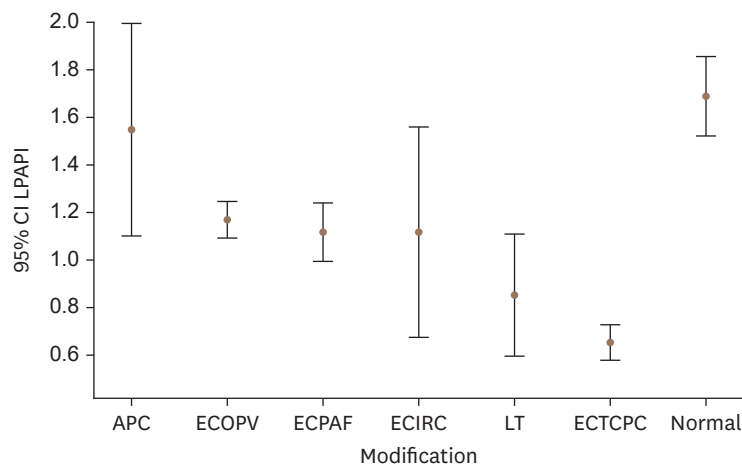


Figure 4. Comparison of mean LPA PI in 6 Fontan modifications. Error bar represents 95% CI. LPA: left pulmonary artery, PI: pulsatility index, CI: confidence interval, APC: atriopulmonary connection, ECOPV: extracardiac conduit with oversewn pulmonary valve, ECPAF: extracardiac conduit with preserved adjusted antegrade flow, ECIRC: extracardiac conduit from inferior vena cava to right ventricular outflow tract, LT: lateral tunnel, ECTCPC: extracardiac total cavopulmonary connection.

among all the Fontan modifications: RPA PI = 0.62 and LPA PI = 0.65. The PI of PA branches was lower in the order of ECTCPC, LT, ECIRC, ECPAF, ECOPV and APC (**Table 2**). The Doppler velocities and PI in Glenn and Fontan conduit or pathway are shown in **Table 3**. We also used a new index of global PI calculated as the sum of PA branches, Glenn and Fontan conduit or pathway PI divided by 4 in the Fontan palliated patients. In normal individuals, the average of PA branches PI was calculated as the global pulsatility (**Table 4**). The APC and the ECTCPC groups had the highest and lowest global PI of 1.46 vs. 0.63, respectively. No significant statistical difference was found between the mean of global PI in the groups of APC and normal control (1.46 vs. 1.66, $p = 0.11$).

Table 3. Comparison of velocities and PIs of Glenn and Fontan conduit flows between ECTCPC and 5 other modifications of Fontan

Pathway	Fontan modifications	Number	V max (cm/s)	p*	V min (cm/s)	p*	V mean (cm/s)	p*	PI	p*
Glenn	APC	2	54.00 ± 16.97	0.19	13.80 ± 1.13	0.19	30.00 ± 11.31	0.98	1.33 ± 0.02	< 0.001
	ECOPV	8	41.06 ± 15.63	0.86	12.08 ± 3.38	0.001	24.14 ± 6.08	0.67	1.04 ± 0.19	< 0.001
	ECPAF	4	37.25 ± 5.62	1.00	11.17 ± 1.76	0.004	23.82 ± 2.39	0.77	1.09 ± 0.13	< 0.001
	ECIRC	2	43.25 ± 5.73	0.91	4.60 ± 6.50	< 0.001	24.10 ± 0.56	0.93	1.59 ± 0.47	< 0.001
	LT	2	32.00 ± 2.83	0.99	17.25 ± 1.06	0.87	22.30 ± 0.42	0.72	0.66 ± 0.05	0.985
	ECTCPC	10	36.27 ± 7.07		19.93 ± 4.49		27.53 ± 5.90		0.59 ± 0.09	
Fontan Conduit	APC	2	64.50 ± 13.43	0.60	17.00 ± 2.83	0.70	34.00 ± 7.07	1.00	1.39 ± 0.02	< 0.001
	ECOPV	8	62.56 ± 11.45	0.25	18.96 ± 3.28	0.50	39.57 ± 6.23	0.79	1.11 ± 0.20	< 0.001
	ECPAF	4	73.70 ± 21.09	0.06	19.25 ± 7.49	0.74	50.90 ± 15.53	0.002	1.07 ± 0.09	0.002
	ECIRC	2	42.85 ± 20.86	0.99	5.60 ± 7.92	0.04	22.90 ± 14.00	0.63	1.78 ± 0.53	< 0.001
	LT	2	43.70 ± 6.08	1.00	22.60 ± 2.83	0.99	31.00 ± 1.41	0.99	0.68 ± 0.07	0.999
	ECTCPC	10	46.89 ± 19.08		25.17 ± 12.23		34.00 ± 12.42		0.64 ± 0.09	

Values in bold indicate significant p-value.

APC: atriopulmonary connection, ECIRC: extracardiac conduit from inferior vena cava to right ventricular outflow tract, ECOPV: extracardiac conduit with oversewn pulmonary valve, ECPAF: extracardiac conduit with preserved adjusted antegrade flow, ECTCPC: extracardiac total cavopulmonary connection; LT: lateral tunnel, PI: pulsatility index, V: velocity.

*Each parameter was compared to the relevant ECTCPC group parameter with the Dunnett's *post hoc* test.

Table 4. Comparison of global PIs of PA branches flows between 6 modifications of Fontan and normal groups with the Dunnett's *post hoc* test

Fontan modifications	Number	Global PI	p*
APC	2	1.46 ± 0.03	0.11
ECOPV	8	1.13 ± 0.09	< 0.001
ECPAF	4	1.11 ± 0.07	< 0.001
ECIRC	2	1.41 ± 0.21	0.02
LT	2	0.75 ± 0.05	< 0.001
ECTCPC	10	0.63 ± 0.07	< 0.001
Normal	6	1.66 ± 0.08	

Values in bold indicate significant p-value.

APC: atriopulmonary connection, ECIRC: extracardiac conduit from inferior vena cava to right ventricular outflow tract, ECOPV: extracardiac conduit with oversewn pulmonary valve, ECPAF: extracardiac conduit with preserved adjusted antegrade flow, ECTCPC: extracardiac total cavopulmonary connection, PI: pulsatility index, LT: lateral tunnel.

DISCUSSION

Creation of pulsatile Fontan circulation is a dream in the surgical management of single ventricle pathology. It has been claimed that pulsatile flow in Fontan palliation will promote the growth of pulmonary arteries and hamper the elevation of PVR over time.⁵⁽⁶⁾⁹⁾ Experimental studies have shown that pulsatile flow has an undeniable impact on the growth of great PA branches and pulmonary capillary system.¹⁰⁽¹²⁾ Moreover, noninvasive methods including magnetic resonance²⁽¹³⁾ and echocardiographic imaging have been used to measure the PI in Fontan circulation. Although there are some limitations to achieve the optimal Doppler tracing and also obligatory assumptions for simple calculation, Doppler-derived PI seems to be a reproducible technique in Fontan palliated population.

Our findings showed that the APC modification has the closest PI to the normal cardiovascular circulation. Rebergen et al.¹⁴⁾ showed the similarities of the biphasic pulsation in the pulmonary arteries of APC modification with the v and a wave of a normal jugular vein. They concluded that inclusion of the atria in the Fontan circulation will better preserve the pulsation. However, the near normal PI in the APC modification does not bring a favorable cardiovascular function and outcome.¹⁵⁾ Although the inclusion of atrial contraction and relaxation in APC modification will add pulsation to the PA circulation, it has been shown that exaggerated flow disturbances at the level of atriopulmonary anastomosis will cause energy losses.⁸⁽¹⁴⁾ The progressive atrial

enlargement in this type of Fontan as a nidus for arrhythmia and mural thrombosis has led to the obsolescence of this kind of procedure.^{15,16)} Our patients in the group of APC were 2 female athletes aged more than 30 years who were surprisingly in class I NYHA and had a past history of successful childbearing. However, it should be noted that the fate of them cannot be attributed to all patients of APC modification.

In the ECPAF modification, preserved adjusted antegrade flow will add volume load in addition to the pulsation onto the Fontan circulation that may not always be tolerated. In some cases, we had to occlude the preserved antegrade flow because of Fontan failure. This modification also had a PI of about 1.1.

In ECOPV modification the oversewn pulmonary valve will act as a membrane with forward movement into the Fontan pathway adding pulsatility without imposing extra volume on the circulation; an advantage of this modification compared to the group with preserved adjusted antegrade flow. Although the PI in ECOPV group was more than 1.1, this type of surgery is limited to a minority of single ventricle physiology with double discordance anatomy. Moreover, the long-term outcome of this modification needs to be assessed.

We had 2 patients in the ECIRC group that Fontan conduit was anastomosed directly onto the rudimentary RV with a well-developed RVOT and pulmonary valve (2 cases of tricuspid atresia type Ib). This type of operation makes the Fontan circulation pulsatile. However, the profound backflow into the hepatic veins may cause liver congestion that forced us to convert it to ECTCPC in one patient (**Figure 5, Supplementary Video 3**). The ECIRC group had a PI of about 1.1 in both PA branches.

The 2 modifications of LT and ECTCPC had the least PI in PA branches, 0.8 and 0.6, respectively. Two mechanisms were suggested for the pulsatility observed in the flow pattern of the patients with LT modification. The first is the change of left atrial pressure that is transmitted directly to the LT via the patch movement. The contraction of the remaining atrial muscular wall in the pathway of LT is the second mechanism that may give some pulsatility to the circulation.⁸⁾

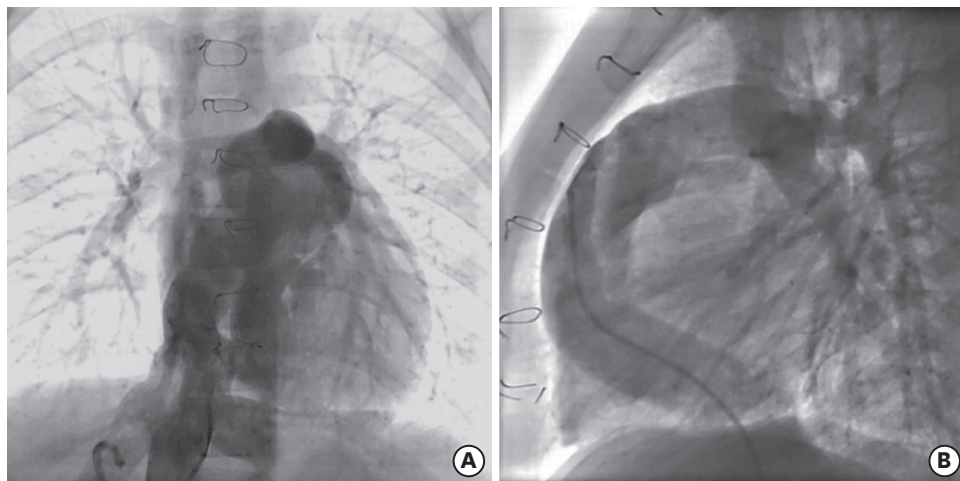


Figure 5. Conduit injection in (A) AP and (B) lateral views in a case of ECIRC showing prominent hepatic vein engorgement due to profound reversal flow concomitant with systolic contraction of rudimentary RV. AP: atriopulmonary, ECIRC: extracardiac conduit from inferior vena cava to right ventricular outflow tract, RV: right ventricle.

We had limited number of samples in a few groups of patients due to either obsolescence or novelty of the operation. Moreover, difficulty in obtaining the optimal echo views in patients with multiple sternotomies, difficulty in alignment of the sample volumes in PA branches by transducer angulation and using the assumption of fixed vessel diameter during respiration were other limitations in our study.

In conclusion, we found that ECTCPC and APC modifications had the lowest and the highest PI in PA branches respectively. Furthermore, we could add pulsation to the Fontan circulation by preserving an oversewn well-developed pulmonary valve in cases of double discordance or preserving a tiny adjusted antegrade flow in cases having pulmonary stenosis and by connection of Fontan conduit directly onto a rudimentary RV in cases of tricuspid atresia. However, it is important to notify that these surgical modifications might lead to complications in some patients that Fontan conversion to ECTCPC type should be considered as the last resort.

SUPPLEMENTARY MATERIALS

Supplementary Video 1

Parasternal echo view in a double discordant patient showing to and fro movement of oversewn well-developed pulmonary valve without passage of color across it.

[Click here to view](#)

Supplementary Video 2

Short axis view in a case of extracardiac conduit from inferior vena cava to right ventricular outflow tract (ECIRC) showing smooth red color passage across the right ventricular outflow tract into a well-developed pulmonary valve in addition to contractile motion of rudimentary right ventricle (RV).

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Supplementary Video 3

Anteroposterior and lateral conduit injection in a case of extracardiac conduit from inferior vena cava to right ventricular outflow tract (ECIRC) showing its connection onto the rudimentary right ventricle (RV) with profound reversal flow causing hepatic vein engorgement.

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