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Successful Second Attempt at Fontan Completion Following Fontan Takedown and Its Long-Term Outcome

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1. Abstract

3-year-old girl with single ventricle physiology underwent a second attempt at a fenestrated Fontan completion following a Fontan takedown. The preoperative diagnosis was asplenia, with single right ventricle, post bidirectional Glenn(BDG) procedure. The cardiac catheterization data showed an SVC pressure of 12mmHg, RA pressure of 6mmHg, PA index of 141mm2/m2, a pulmonary vascular resistance (Rp) of 2.7unit·m2. The SpO2 was 85-90%. The elective staged Fontan completion was performed using extracardiac total cavopulmonary connection(TCPC) procedure with an expanded polytetrafluoriethylene(ePTFE) graft, 16mm without fenestration. Although the postoperative central venous pressure(CVP) was 15-16mmHg, the pleural drainage volume was high. Therefore, the addition of fenestration (3.5mm), between the RA and the ePTFE graft was performed on postoperative day 10. Finally, the patient was taken down to the BDG circulation on POD13 because of the difficulty of maintaining stable hemodynamics. Afterwards, because of her family's strong preference, she underwent a second attempt at a Fontan completion with fenestration(4mm), after meticulous coil embolization of the collaterals and cleaning of the left subclavian artery. She torelated the procedure and final CVP was 13mmHg. Seven years later, she is oxygen free and the SpO2 is being maintained at 90%.

2. Introduction

Little is known regarding the subsequent outcome of a Fontan takedown and the potential candidacy for a second attempt at a Fontan completion In2007, Almond et.al. from Children's Hospital Boston, demonstrated the outcome of 53 cases after Fontan failure and takedown to an intermediate palliative circulation and concluded that the failure of Fontan completion does not necessarily mean that a patient cannot tolerate the Fontan circulation [1]. While Fontan patients may show a relatively high central venous pressure(CVP) > 15mmHg in the long-term period, a Fontan circulation may be considered difficult to maintain in the acute period. Herein, we present a successful redo-Fontan completion following a Fontan takedown and its long-term outcomes.This case was presented at the "Nightmare case" session at the EACTS Annual Meeting, in Vienna, in 2017 The patient's parents provided informed consent for the presentation as well as publication.

3. Case

3-year-old girl underwent a second attempt at fenestrated Fontan completion following a Fontan takedown. Using, fetal echo cardiography, she was diagnosed as Asplenia, {A(s),D,L} single right ventricle (SRV), double outlet right ventricle (DORV), pulmonary stenosis (PS), bilateral superior vena cava(SVC), common atrioventricular valve (CAVV),by fetal echocardiography and was

born via normal vaginal delivery(38weeks and 4days). The body weight at birth was 2524 g. She had undergone a right modified Blalock-Taussig shunt (RMBT): (operation1), and bilateral bidirectional Glenn (BDG) anastomosis with division of main pulmonary artery trunk while taking down RMBT(operation2) at the age of one month and two years, respectively. The following cardiac catheterization data showed an SVC pressure of 12mmHg, right atrium (RA) pressure of 6mmHg, PA index of 141mm2/m2, pulmonary vascular resistance (Rp) of 2.7unit・m2, and right ventricular end-diastolic volume (RVEDV) of 224% of the normal value with a right ventricular ejection fraction (RVEF) of 75%. The SpO2 was 85-90%. The chestradiography revealed a cardiothoracic ratio of 56.0%. The angiogram revealed many collaterals from the left internal thoracic artery (LITA) in the lung. Therefore, elective staged Fontan completion (operation3, Video1) was planned after coil embolization for LITA. A Fontan completion was performed with extracardiac total cavopulmonary connection (TCPC) procedure using expanded polytetrafluoriethylene(ePTFE) graft, 16mm without fenestrstion. Although the postoperative central venous pressure (CVP) was 15-16mmHg, pleural drainage volume was so high. Therefore, coil embolization for the systemic –pulmonary collaterals was performed on the postoperative day (POD) eight, and CVP decreased from 23mmHg to 16mmHg. However the low cardiac output condition continued, and the addition of fenestration(operation4), 3.5mm between the RA and the ePTFE graft was performed on POD10. Finally, the patient was taken down to BDG circulation(operation5) on POD13 because of difficulty in maintaining stable hemodynamics.

Subsequently, because of her family's strong preference, she underwent a second attempt at a Fontan completion with fenestration, 4 mm (operation7, Video2) after a meticulous coil embolization of collaterals and cleaning of the left subclavian artery [2], surgical division of small collateral branches from subclavian artery; operation6). The final CVP was 13 mmHg and SpO2 was 80% with oxygen inhalation of 2 L/min. Two years later, CVP was kept 11-12 mmHg and the SpO2 increased to 88% under oxygen inhalation (1 L/min). Change in the hemodynamic data was shown in Table. At the presentation ("Nightmare case" session) at the EACTS annual meeting, in Vienna, in 2017(three years after redo- Fontan completion), we asked the audience, "Which is better, a BDG with many collaterals or a fenestrated TCPC with much oxygen (the current status)?" Most of them answered that the BDG with many collaterals was better. However, during the further long-term period (seven years after the redo-Fontan completion), fenestrated TCPC was shown to be better than BDG and she is currently oxygen free.

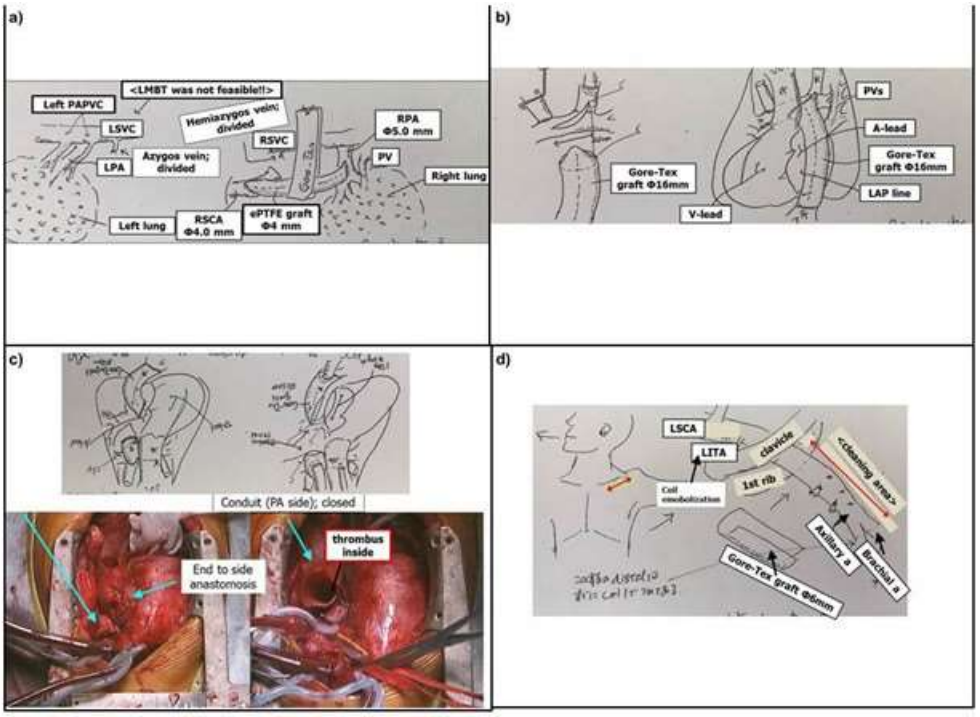

Figure: Operative findings

α) operation1. Left modified Blalock-Taussig shunt after a temporary trying of right modified Blalock-Taussig shunt(LMBT). LMBT was not feasible because of left partial anomalous pulmonary vein. Eventually bilateral thoracotomy was done

β) operation 3. First Extracardiac TCPC procedure without fenestration

χ) operation 5. The patient was taken down to the BDG circulation

δ) operation 6. Cleaning of the left subclavian artery. Collateral branches from left subclavian artery were surgically divided.

Table 1: Hemodynamic data in each phase. TCPC: total cavo pulmonary connection, BDG: bidirectional Glenn, SVC(IVC)p: superior vena cava (inferior vena cava) pressure, Rp: pulmonary vascular resistance, PA index: NaKATA pulmonary srtery index, spO2: percutaneous oxygen saturation, RVEDV: right ventriculae end-diastolic volume, RVEF right ventricular ejection fraction, CAVVR: common atrioventricular valveregurgitation, CTR: cardiothoracic ratio, BNP: brainnatrium peptide.

	Pre1st TCPC (=BDG)	Post failed TCPC	Pre redo-f-TCPC	Post f-TCPC1	Post f- TCPC2	At present
Age	1y7mo	3y1mo	3y7mo	4y10mo	5y9mo	10y6mo
SVCp (mmHg)	13-14	14	12-13	14	12	
IVCp (mmHg)	6	6	6	14	11	
Rp (unit·m^2)		2.1				
Qp (L/min/m^2)	3.9	3.1		4.6	3.7	
PA index (mm^2/m^2)	141	181	167			
RVEDV (%of normal value)	224	184				
RVEF (%)	61	63				
SpO2 (%)	85-90		82-84	85-90	88-90	90
Oxygen (L/min)	none		none	0.5	1	none
CAVVR	mild		Trivial-mild	mild	trivial	trivial
CTR (%)	50			45	51	50
BNP (pg/ml)	74.7			30	35.7	30

4. Discussion

The ultimate goal of the staged Fontan procedure which is performed in patients with single ventricle physiology is the achievement of optimal systemic oxygen delivery (i.e.cardiac output) withas low CVP as possible. To accompolish this goal, it is important to prepare the more flexible pulmonary compliance before Fontan completion and to perform the more supportable surgical technique at Fontan completion. However, there are some critical but important surgical technical points and disturbing conditions to be removed. “The disturbing conditions” that can affect the Fontan circulation during both the acute and late period. are, for example, systemic atrioventricular valve regurgitation, poor systemic ventricular function, high pulmonary resistance, and many systemic-pulmonary collaterals. With regard the aortopulmonary collaterals, Grosse-Wortmann et al. from the Toronto Sick Children’s Hospital performed a multimodality study and concluded that the hemodynamic consequences of the aortopulmonary collateral flow translate into early adverse outcomes after total cavopulmonary connection (TCPC) completion [3]. In the current case,

the repeated coil embolization for aortopulmonary collaterals and SCA cleaning [2] were helpful for the maintenance of the Fontan circulation in the acute period. The patient was able to tolerate the Fontan circulation in the long-term period. Next, an important surgical technique that was considered, was a fenestration. Although some groups continue to suggest that a fenestration is not helpful for the majority of patients undergoing a Fontan procedure, the majority of recent reports that have examined this issue have suggest that a fenestration is helpful in reducing morbidity, particularly pleural effusions, shortening hospitalization, and possibly reducing mortality [4-6]. In particular, Lemler et al performed a prospective randomized trial in which 25 patients were randomized to a fenestrated Fontan and 24 to a non-fenestrated Fontan procedure. They concluded that the fenestration performed at the time of Fontan surgery improves short-term outcome in standard-risk patients by decreasing pleural drainage, hospital length of stay, and need for additional postoperative procedures [5]. Similarly. Although we reported our experience with a two-staged extracardiac TCPC without fenestration in 2009[7], we have changed the Fontan strategy to the routine use of fenestration even in the patient who is considered as the good candidate, Fenestration is a safe valve for a Fontan completion because the Fontan circulation is not yet well understood. We believe that fenestration is helpful in Fontan completion in both standard –risk patients and high-risk patients, The second factor that contributes to a successful Fontan completion is anastomosis method. Although it is famaous that TCPC is superior to atriopulmonary connection or Bjork procedur], it is still controversial whether extracardiac TCPC is superior to lateral tunnel. Recently, an anterior anastomosis of TCPC was reported in the literature [8], although we had already performed the similar technique since 2013(not officially reported, just presented at the domestic meeting [9]. Finally, according to Almond et.al. The failure of a Fontan completion does not necessarily mean that a patient cannot tolerate a Fontan circulation [1]. From this surgical case, we learned the meaning of the definitive surgery. Glenn anastomosis is an example of palliative surgery, and Fontan is a definitive surgery even if with fenestration. After the completion of fenestrated Fontan, collaterals were diminished. That is why systemic venous return can easily flow into the pulmonary vascular beds. This was consistent with the disappearance of the desaturation. The drainage volume of pleural effusion was proportional to the cardiac output with Qp/Qs closer to 1.0. In conclusion, we succeeded the second attempt at the Fontan completion following the Fontan takedown. The patient may have had treatable lesions that affected Fontan circulation adversely. After determining whether the conditions were favorable, she was identified as a potential candidate to ubdergo redo Fontan completion after setting favorable conditions. As a result, the fenestration contributed to the less pleural drainage. Probably the fenestration decreased the CVP and systemic congestion, and increased cardiac output. Furthermore, we have the another experience with Fontan completion for a tricuspid atresia patient with left ventricular non-compaction [10]. Appropriate preload contributed to adequate cardiac output. However, the current report does not include the exercise test in the long-term period. Therefore, the excersize capacity of this patient is not known.

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