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=Abstract=

Treatment of Protein-losing Enteropathy After Fontan Procedure by Conversion to the Total Cavopulmonary Connection with Fenestration

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As the operative mortality has diminished and the number of survivors has increased after Fontan procedure, morbidities related to the unnatural physiology of cavopulmonary flow have developed. One of the complications by the hemodynamic derangement after Fontan procedure is a protein-losing enteropathy. This is a rare but life-threatening complication after the Fontan operation. Treatment strategies are highly variable. But, reports on successful management are limited. We experienced three cases of protein-losing enteropathy after the Fontan operation. We report that the conversion to the total extracardiac or intracardiac cavopulmonary connection with fenestration is a satisfactory treatment modality for protein-losing enteropathy after the Fontan operation.

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Key words : 1. Fontan Operation
2. Enteropathy

1 . 4 3 4 mm

5 7

가 4 2 4 mm

13 15 mmHg, 11

mmHg, 7 8 mmHg

(70 mmHg) 22

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2 , 가 가 18
 ACE(angiotensin converting enzyme) 234 mg/dL, -1-antitrypsin 15 , -1-antitrypsin
 8 . 100 g, -1-antitrypsin 19.6 mg/dL, 24
 1 1 50 ×50 (reference range; <13 ml/24hours) 27
 mm 0.85, 1 3.0 g/dL 7
 mmHg, 65%, 22/ 18(20) / 7.5/4.8 g/dL
 62%, 66%, 가 0.74 .
 58%, 140/0(8) mmHg, 2
 93%, 140/70(90) mmHg, 87%
 가 4.6/2.6 g/dL 20% 26
 75 ml 가 9
 1 8 3 Blalock-Taussig 12
 , 가 가 2 Woven Dacron . Blalock-Taussig
 / 3.7/2.0 g/dL 가 4
 2 3 , 2 3.9/2.2 g/dL /
 2.8 ×2.4 cm
 / 3.8/ 1.8 g/dL . 20% 187.0 mg/dL, -1-antitrypsin 107.6 mg/dL,
 50 ml 가 24 140 g, -1-antitrypsin 161.1
 -1-antitrypsin 403 mg/dL, .
 -1- antitrypsin 704 mg/dL, 24 34 g, 17/7(12) mmHg, 74%,
 -1-antitrypsin 59.4 ml/24hours(reference range; 15/9(12) mmHg, 73%,
 < 13 ml/24hours) . 68%, 82%,
 117/0(7) mmHg, 93%, 115/60(80)
 mmHg, 93%
 1 2 4 - 14 4
 internal baffle 5 mm . Goretex 5 mm Gortex 18 mm
 26
 14 17 mmHg, 3 5 mmHg . 13
 , ACE inhibitor, 35
 2 . 3.0 g/dL 44

antitrypsin 40 -1-
10.98 ml/24hours(reference range;<27 ml/24
hours) 1970 , , ,
3 , Kreutzer²⁾
4 6 1
, , ,
, , , 3)
, 1 5 가 -
, , , DeLeval⁴⁾ 가 .
, 1 5 - 가 .
, , , (turbulence) 가
2 5 4 mm 가
mm , 4 -
, -
2 21 (transcoronary perfusion gradient)
가 가⁵⁾ McElhinney⁶⁾
9 ,
, / 3.1/1.6 g/dL 가 3
-1-antitrypsin 303 mg/dL,
-1-antitrypsin 622 mg/dL, 24 190 g, 가
-1-antitrypsin 390.0 ml/24hours(reference
range; < 27 ml/24hours) . 1 2 가
20% 100 ml 4 , .
8 . 2 1 가
/ 3.2/1.6 g/dL , 20 % . , ,
, , ,
2 1 .
Gortex 18 mm Gortex 5 mm .
. 10 .
. 18 가 .
2.8-3.0 g/dL 25 .
. 101 S C 가 .
가 19 g/dL 가 .

Table 1. Patient Characteristics

Patient	Diagnosis	Prior Surgery	Age at Fontan	Interval to PLE	Age at TCPC	Type of TCPC	Fenestration Size(mm)
case #1	DORV remote VSD PS	RA to PA anastomosis with fenestration	3yr 3mo	2yr 3mo	7yr 7mo	Intracardiac	5
case #2	Tricuspid atresia VSD PS ASD, PDA	RA to PA anastomosis without fenestration	12 yr	14yr	26yr	Extracardiac	5
case #3	DORV with rudimentary LV Mitral atresia PS	RA to PA anastomosis with internal baffling and fenestration	2yr 5mo	9mo	4yr 6mo	Extracardiac	5

Table 1. Patient Characteristics

Patient	Preoperative			Postoperative			Other Complications	Result of PLE
	Saturation(%)	CVP (mmHg)	-1-antitrypsin clearance in stool (ml/24hr)	Saturation(%)	CVP (mmHg)	-1-antitrypsin clearance in stool (ml/24hr)		
case #1	87	20	59.4	93	14	8.4	Giant RA	Resolved
case #2	93	12	161.1	94	12	10.98	Giant RA RA thrombi Atrial tachycardia Ascites	Resolved
case #3	80	14	390.0	92	11	349.1	Enlarged internal baffling	Recurred & Resolving

ASD, atrial septal defect; CVP, central venous pressure; DORV, double outlet of right ventricle; LV, left ventricle; PA, pulmonary artery; PDA, patent ductus arteriosus; PLE, protein-losing enteropathy; PS, pulmonary stenosis; RA, right atrium; TCPC, total cavopulmonary connection; VSD, ventricular septal defect

1980 가 가
가 가
(15 20 mmHg) ACE 가
1 2

medium-chain triglycerides(MCT)가

가

가

(fenestration)

9, 2, 3, 14

-1-antitrypsin
internal baffle

(Table

1).

11 14 mmHg

2

가 1

가

2

가 2.0 g/dL

, 3

가 2.0 g/dL

Goretex 5 mm

, internal baffle

external baffle

가

가

(Fig. 1).

가

1)

, 2)

, 3)

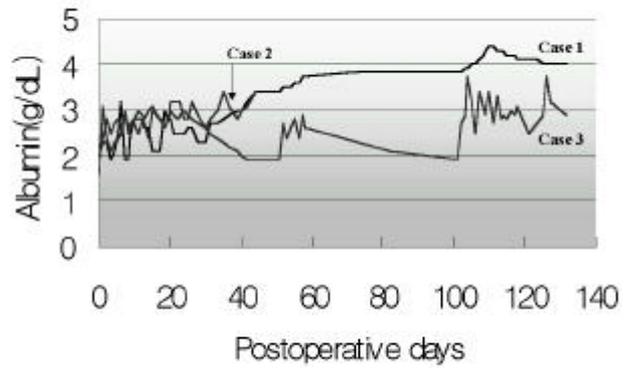
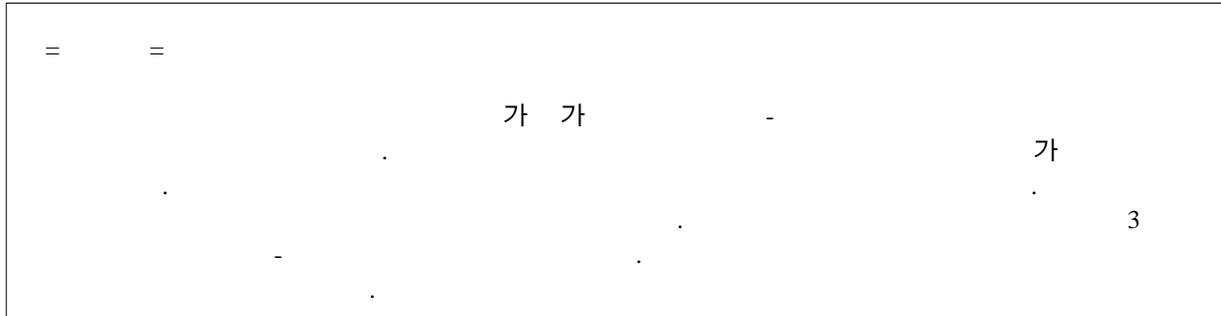


Fig. 1. Postoperative changes of albumin

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