

# Successful implantation of a left ventricular assist device in a patient with arrhythmogenic right ventricular cardiomyopathy : A potential candidate for left ventricular assist device



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## Introduction

Left ventricular assist devices (LVADs) are generally applied to advanced heart failure patients with spared right ventricular (RV) function. Therefore, patients with arrhythmogenic right ventricular cardiomyopathy (ARVC) who primarily present RV dysfunction are generally deemed ineligible for LVAD. We here report on a patient with advanced heart failure due to ARVC who successfully underwent LVAD implantation as bridge to transplantation.

## Case: A 42-year-old, male

### History of present illness

- A 42-year-old male with advanced heart failure due to ARVC was referred to our institution in 2009.
- Since he had New York Heart Association functional (NYHA) class III symptoms with symptomatic ventricular tachycardia, he received a cardiac resynchronization therapy with defibrillator.
- Because of severely reduced functional capacity with symptomatic ventricular arrhythmia, he had been listed as a candidate for heart transplantation (HTx) with status-2 medical urgency (Ambulatory status).
- After the next 8 years, he had gradually become frequent flyer for both arrhythmia and heart failure (NYHA class IV, INTERMACS profile 5) and was considered to be listed as status-1 (In-hospital, inotropic, and/or MCS dependent status).
- A series of examinations including echocardiogram, and right heart catheterization (RHC) of this patient suggests high possibility for the patient to be supported by only LVAD.
- The patient successfully underwent HeartMate II implantation with tricuspid annuloplasty in 2017. Post-operative course was uneventful, and he was discharged on postoperative day 98.
- The patient's functional status improved to NYHA class II. He has been waiting for heart transplantation for 1 year without any significant adverse events.

### Laboratory data

WBC	6900 / $\mu$ L	BUN	19 mg/dl
RBC	$481 \times 10^4$ / $\mu$ L	Cr	1.07 mg/dl
Hb	16.2 g/dl	eGFR	61.6 ml/min
Plt	$16.6 \times 10^4$ / $\mu$ L	Na	136 mEq/l
TP	7.4 g/dl	K	5.1 mEq/l
Alb	4.2 g/dl	Cl	99 mEq/l
T-Bil	0.8 mg/dl	T-cho	119 mg/dl
AST	39 U/l	HDL-C	42 mg/dl
ALT	34 U/l	LDL-C	58 mg/dl
LDH	200 U/l	PT-INR	2.08
CK	57 U/l	HbA1c	6.0 %
CRP	0.08 mg/dl	BNP	231.1 pg/ml

WBC = white blood cell; RBC = red blood cell; Plt = platelet; TP = total protein; T-Bil = total bilirubin; AST = aspartate aminotransferase; ALT = alanine aminotransferase; LDH = lactate dehydrogenase; CPK = creatine phosphorus kinase; CRP = C-reactive protein; BUN = blood urea nitrogen; eGFR = estimate glomerular filtration rate; T-cho = total cholesterol; HDL-C = high density lipoprotein cholesterol; LDL-C = low density lipoprotein cholesterol; HbA1c = Hemoglobin A1c; BNP = brain natriuretic peptide

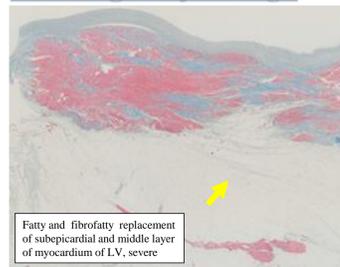
### Echocardiogram



IVS	8 mm
LVPW	8 mm
LVdD	68 mm
LVDs	62 mm
%FS	9 %
LVEF (m-Simpson)	16 %
LAD	32 mm
LAV(m-Simpson)	83 ml
LAVI	49 ml/m <sup>2</sup>
RVdD	56 mm

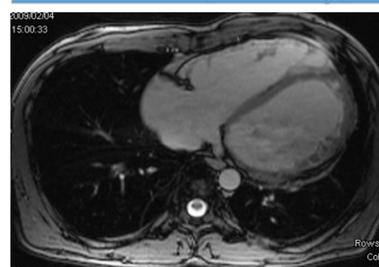
IVS = interventricular septum; LVPW = left ventricular posterior wall; LVdD = left ventricular internal dimension in diastole; LVdS = left ventricular internal dimension in systole; FS = fractional shortening; LVEF = left ventricular ejection fraction; LAD = left atrial dimension; LAV = left atrium volume; LAVI = left atrium volume index; RVdD = right ventricular internal dimension in diastole

### Histological findings



Fatty and fibrofatty replacement of subepicardial and middle layer of myocardium of LV, severe  
Histological findings of the left ventricular apical tissues obtained during surgery for the left ventricular assist device

### Cardiac MRI (February, 2009)

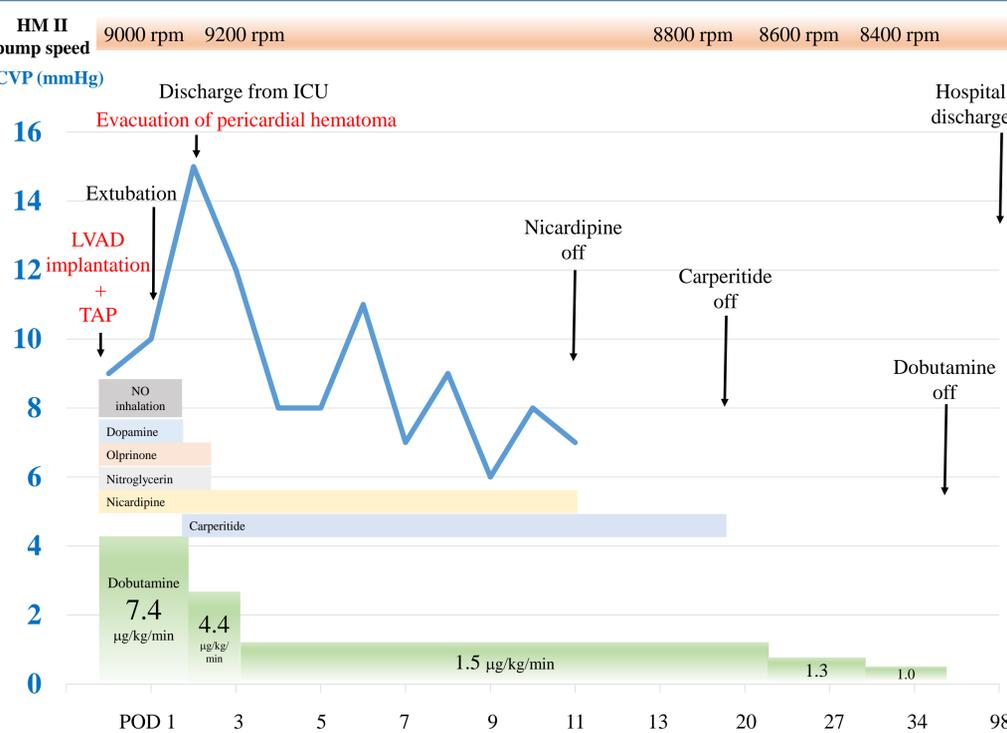


EDV = end-diastolic volume; ESV = end-systolic volume; SV = stroke volume; CO = cardiac output

	LV	RV
EDV (l), ml(ml/m <sup>2</sup> )	309 (182)	510(310)
ESV (l), ml(ml/m <sup>2</sup> )	246 (144)	410 (241)
SV (l), ml(ml/m <sup>2</sup> )	64 (37)	100 (59)
EF, %	21	20
CO (l), ml/min(ml/min/m <sup>2</sup> )	3.7 (2.2)	5.9 (3.5)

- Dilated right ventricular with severely reduced systolic function
- Normal left ventricular volume with severely reduced systolic function

## Clinical Course



### Hemodynamic parameters

	Pre LVAD operation	Post LVAD implantation HM II 8400rpm (POD 39)
PCWP, mmHg	9	5
Systolic PAP, mmHg	20	19
Diastolic PAP, mmHg	7	7
Mean PAP, mmHg	13	12
RVP, mmHg	22 / edp 10	21 / edp 5
RAP, mmHg	6	6
CO (by c-Fick), L/min	3.22	3.81
CI (by c-Fick), L/min/m <sup>2</sup>	1.89	2.38
SV (by c-Fick), ml	46	42
SVI (by c-Fick), ml/m <sup>2</sup>	27	26
PVR, wood units	1.24	1.84
PAPi	2.17	2.00
CVP/PCWP	0.66	1.20
RVSWI	189	156

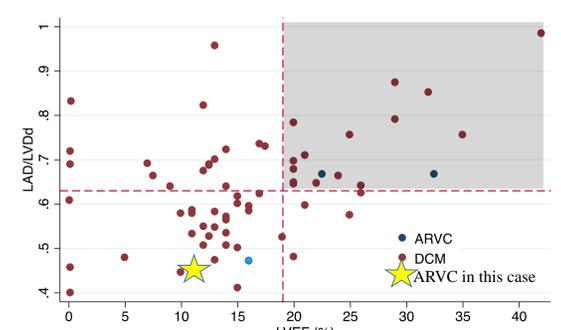
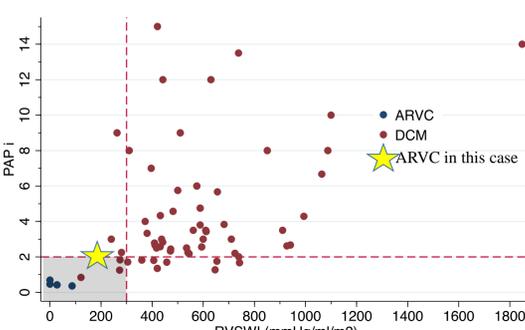
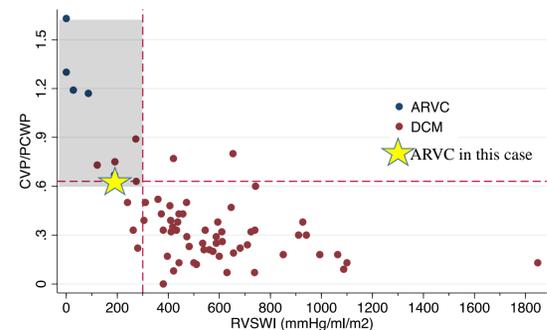
PCWP = pulmonary capillary wedged pressure; PAP = pulmonary artery pressure; RVP = right ventricular pressure; RAP = right atrial pressure; CO = cardiac output; CI = cardiac output index; SV = stroke volume; SVI = stroke volume index; PVR = pulmonary vascular resistance; PAPi = pulmonary artery pulsatility index; RVSWI = right ventricular stroke work index

## Discussion

**Table 1. Demographics and baseline echocardiographic, and hemodynamic parameters in patients with dilated cardiomyopathy (DCM, n=63), and arrhythmogenic right ventricular cardiomyopathy (ARVC, n=5).**

	ARVC (n = 5)	DCM (n = 63)	p value
Age, years	47 (43, 54)	43 (35, 53)	0.245
Male, %	100	77.8	0.303
BW, kg	56.6 (49.4, 74.4)	59.0 (52.0, 67.0)	0.823
BSA, m <sup>2</sup>	1.69 (1.54, 1.84)	1.66 (1.51, 1.79)	0.733
HR, /min	65 (58, 70)	75 (70, 88)	0.016
CI, L/min/m <sup>2</sup>	1.89 (1.38, 2.24)	1.82 (1.52, 2.12)	0.818
PCWP, mmHg	10 (9, 14)	23 (16, 30)	0.013
Systolic PAP, mmHg	20 (18, 22)	43 (33, 53)	0.001
Diastolic PAP, mmHg	9 (9, 16)	24 (17, 30)	0.008
Mean PAP, mmHg	13 (13, 16)	32 (24, 41)	0.003
RAP, mmHg	13 (10, 17)	6 (3, 9)	0.015
PVR, wood units	1.24 (0.89, 2.48)	2.69 (1.72, 4.01)	0.038
PAPi	0.46 (0.42, 0.69)	3.00 (2.18, 5.67)	0.024
RVSWI	280 (0, 140)	510 (41, 670)	<0.001
RAP/PCWP	1.19 (0.85, 1.45)	0.32 (0.19, 0.43)	<0.001
LVdD, mm	60 (42, 66)	74 (68, 83)	0.003
LVEF, %	16 (12, 28)	14 (11, 20)	0.480
LAD, mm	32 (23, 40)	47 (40, 54)	<0.001
LAD/LVdD	0.67 (0.47, 0.67)	0.63 (0.55, 0.71)	0.405

- Preoperatively, we assessed the eligibility for LVAD implantation in current patient referring to the previous reports concerning right ventricular failure (RVF) after LVAD implantation (See below for predictors of post-LVAD RVF)<sup>1-3</sup>.
- Data from ARVC cohort including current patient, and DCM cohort with successful LVAD implantation in our patient population were also assessed.
- In this case, CVP/PCWP ratio (0.66), RVSWI (189 mmHg/m<sup>2</sup>) and PAPi (2.17) were the highest in ARVC group, and LVEF (16%) was the lowest in ARVC. These hemodynamic parameters in this ARVC case are relatively similar to those with DCM group than those the other ARVC group.
- Michigan RV risk score (vasopressor use, creatinine, bilirubin and aspartate aminotransferase) for this patients was low (0 point).<sup>4</sup> These parameters suggested high possibility for the patient to be supported by the only LVAD.
- Given these result above, we decided for this ARVC patient to undergo LVAD implantation.



### Hemodynamic parameters in patients with ARVC or DCM

Vertical line in (A) divides patients in patients with RVSWI  $\leq 300$  mmHg or not; horizontal line divides patients in patients with CVP/PCWP  $\leq 0.63$  or not. Vertical line in (B) divides patients in patients with RVSWI  $\leq 300$  mmHg or not; horizontal line divides patients in patients with PAPi  $\leq 2.00$  or not. Vertical line in (C) divides patients in patients with LVEF  $\leq 19$  % or not; horizontal line divides patients in patients with LAD/LVdD  $\leq 0.63$  or not.

## Conclusions

This case report illustrated that the patient with advanced heart failure due to ARVC could be candidate for LVAD therapy. When considering LVAD therapy for ARVC, we should evaluate a combination of hemodynamic and clinical variables of patients.

## Disclosure

None

## References

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