

# Peri-Partum Cardiomyopathy



## Case Study

פרופ' טל בירון  
מרכז רפואי מאיר  
MEIR MEDICAL CENTER



42 yo

G4P3

39 w

C-HTN

IOL 2d

NVD

3100 gr

Apgar 9/10

PD#1

Dizziness

Resp. Insuf.

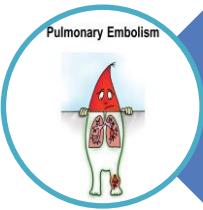
Edema



Bleeding, Anemia



Preeclampsia – severe  
features (Pulmonary edema)

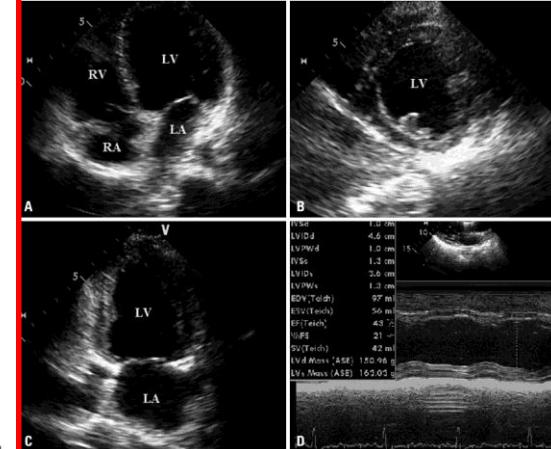
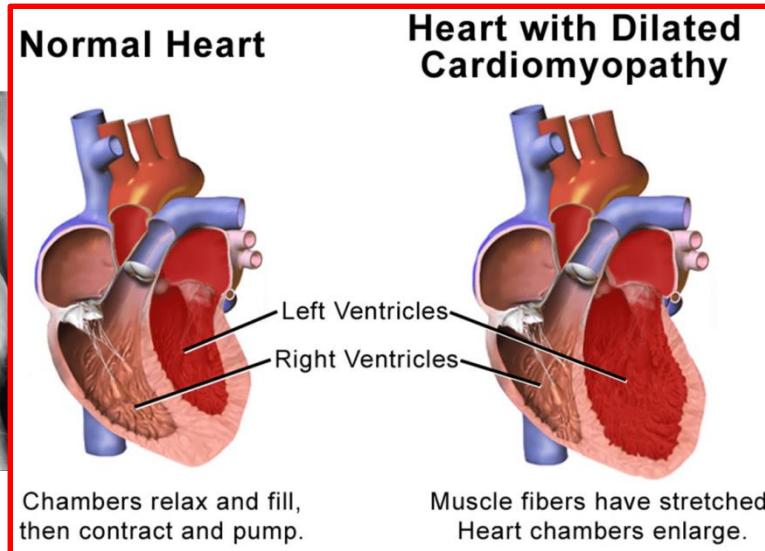
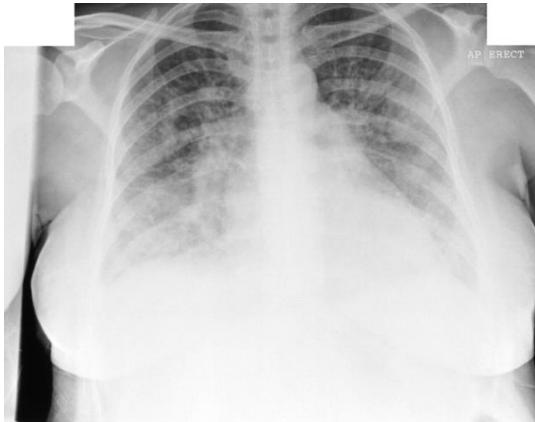


Pulmonary Emboli



PPCM

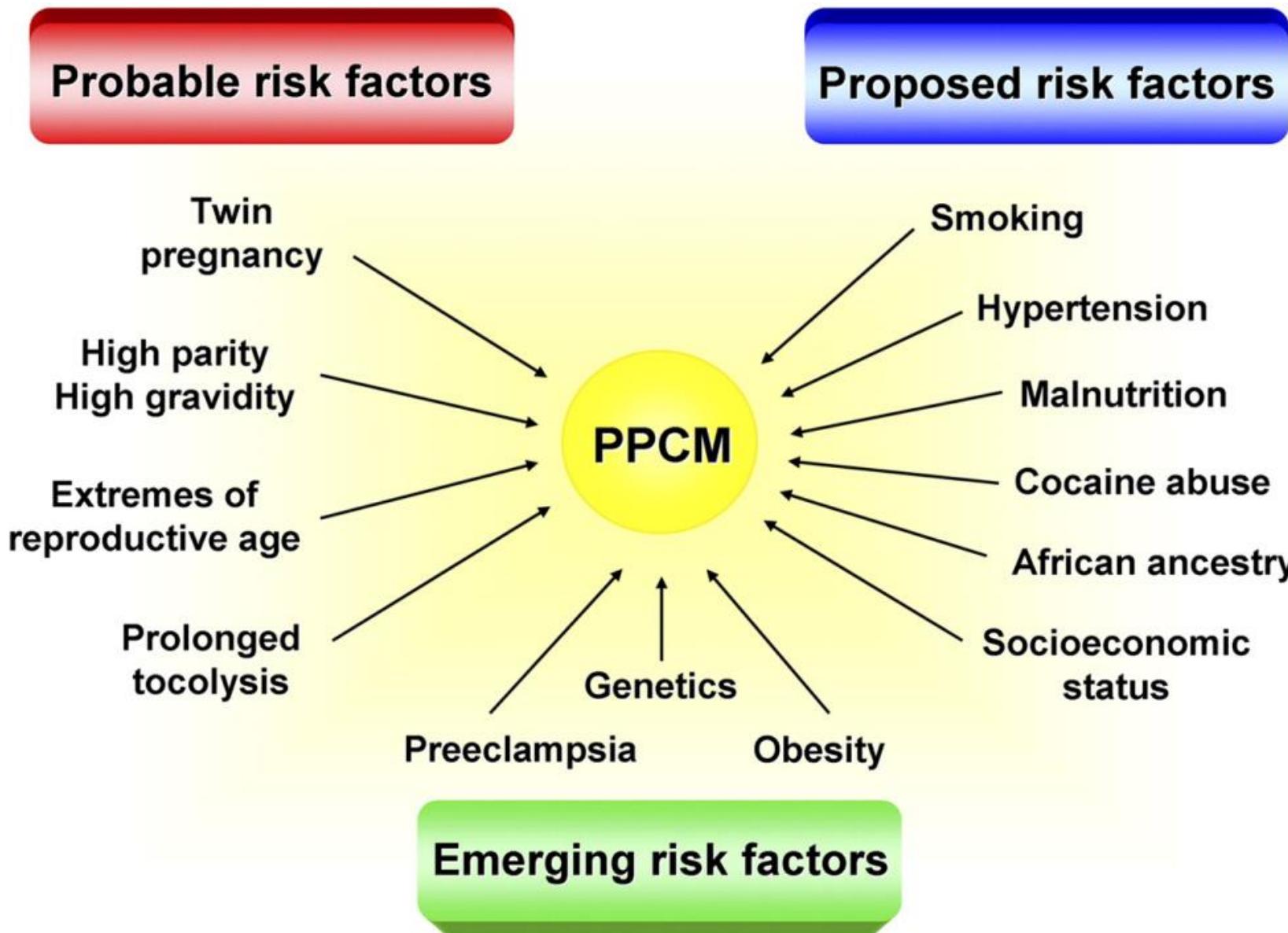
# PERIPARTUM CARDIOMYOPATHY



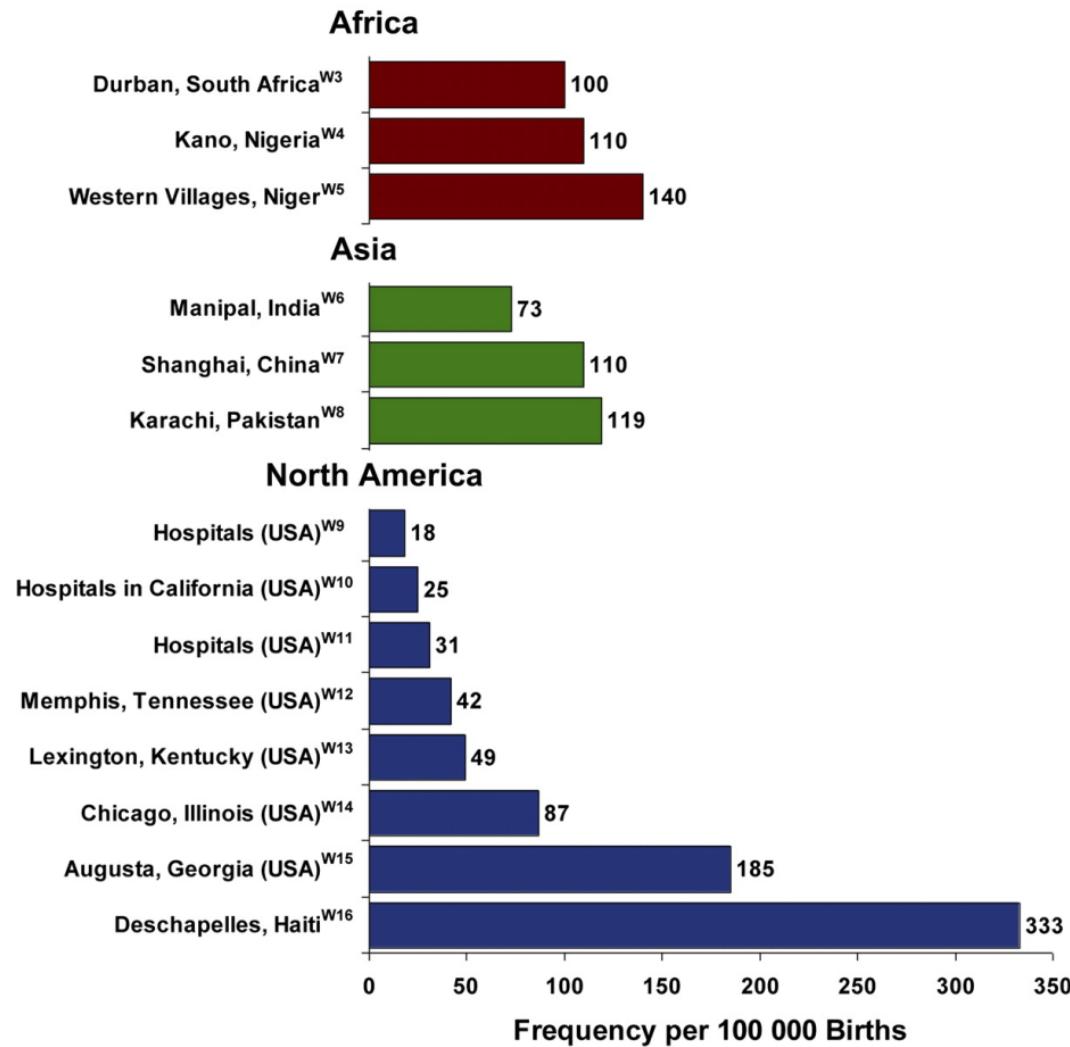
## Clinical criteria for the diagnosis of peripartum cardiomyopathy

- Cardiac failure in the last month of pregnancy or within a few months postpartum
- Absence of another identifiable cause
- Absence of underlying structural heart disease
- LV systolic dysfunction by echocardiographic data:
  1. EF less than 45%
  2. M-mode fractional shortening less than 30% or both
  3. LV end-diastolic dimension greater than  $2.7 \text{ cm/m}^2$

EF 30%



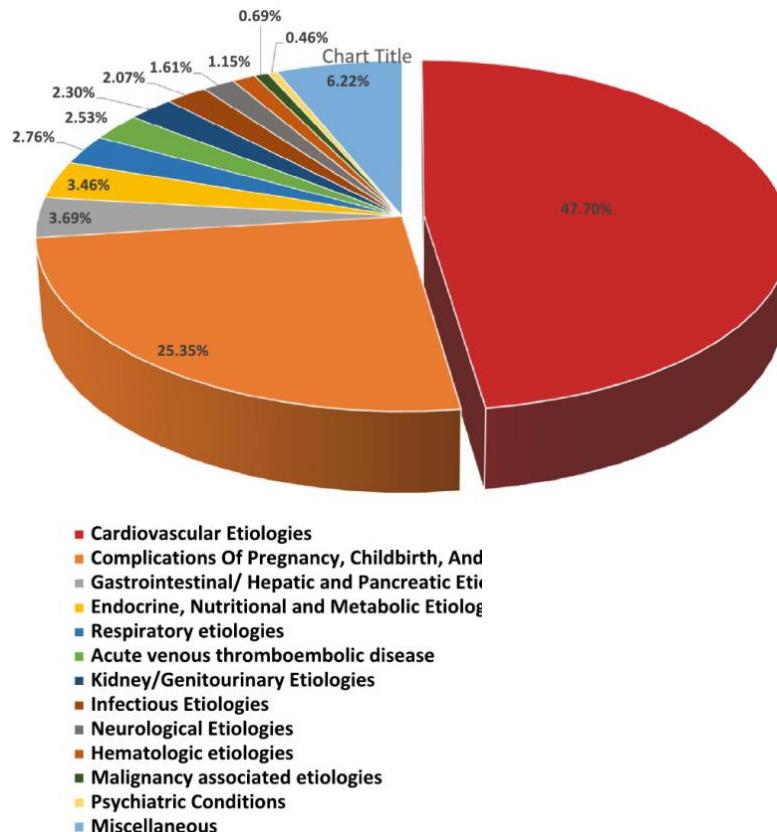
# Incidence of PPCM



# Etiologies, Predictors, and Economic Impact of 30-Day Readmissions Among Patients With Peripartum Cardiomyopathy

Mahek Shah, MD<sup>a,l</sup>, Pradhum Ram, MD<sup>b,l,\*</sup>, Kevin Bryan Lo, MD<sup>b</sup>, Soumya Patnaik, MD<sup>c</sup>, Brijesh Patel, DO<sup>a</sup>, Byomesh Tripathi, MD<sup>e</sup>, Shantanu Patil, MD<sup>d</sup>, Marvin Lu, MD<sup>b</sup>, Ulrich P. Jorde, MD<sup>f</sup>, and Vincent M. Figueiredo, MD<sup>g</sup>

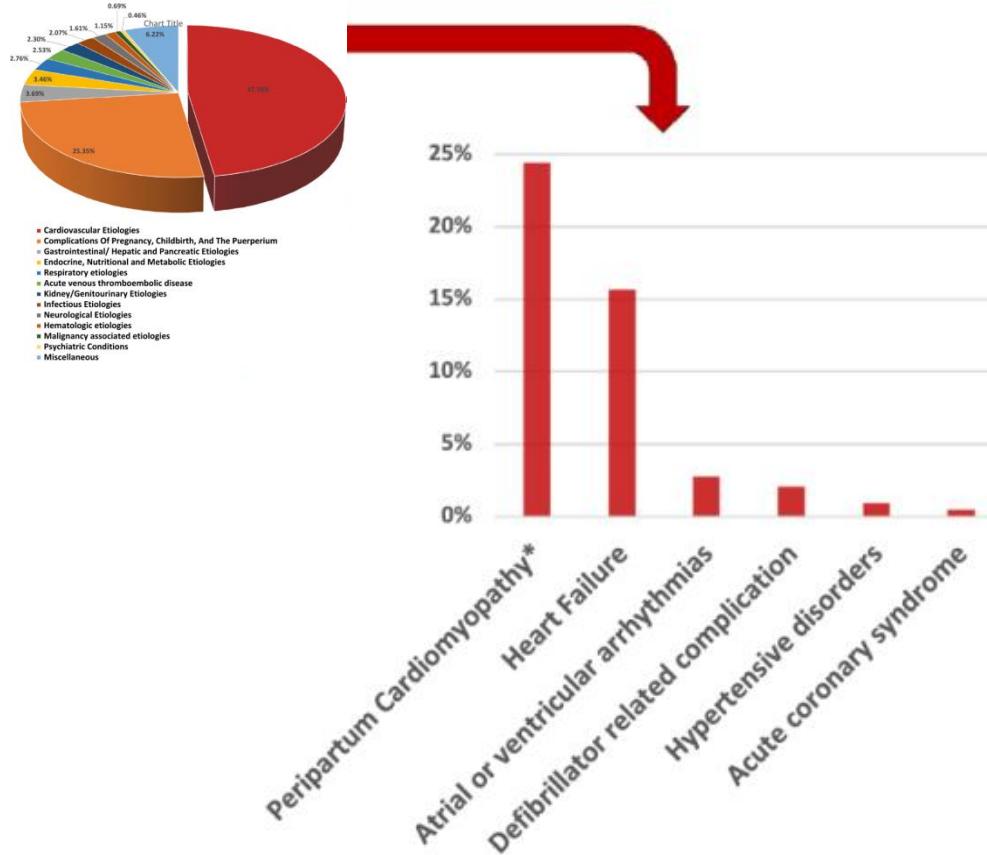
6,977 admissions with PPCM (30d PP national database)



# Etiologies, Predictors, and Economic Impact of 30-Day Readmissions Among Patients With Peripartum Cardiomyopathy

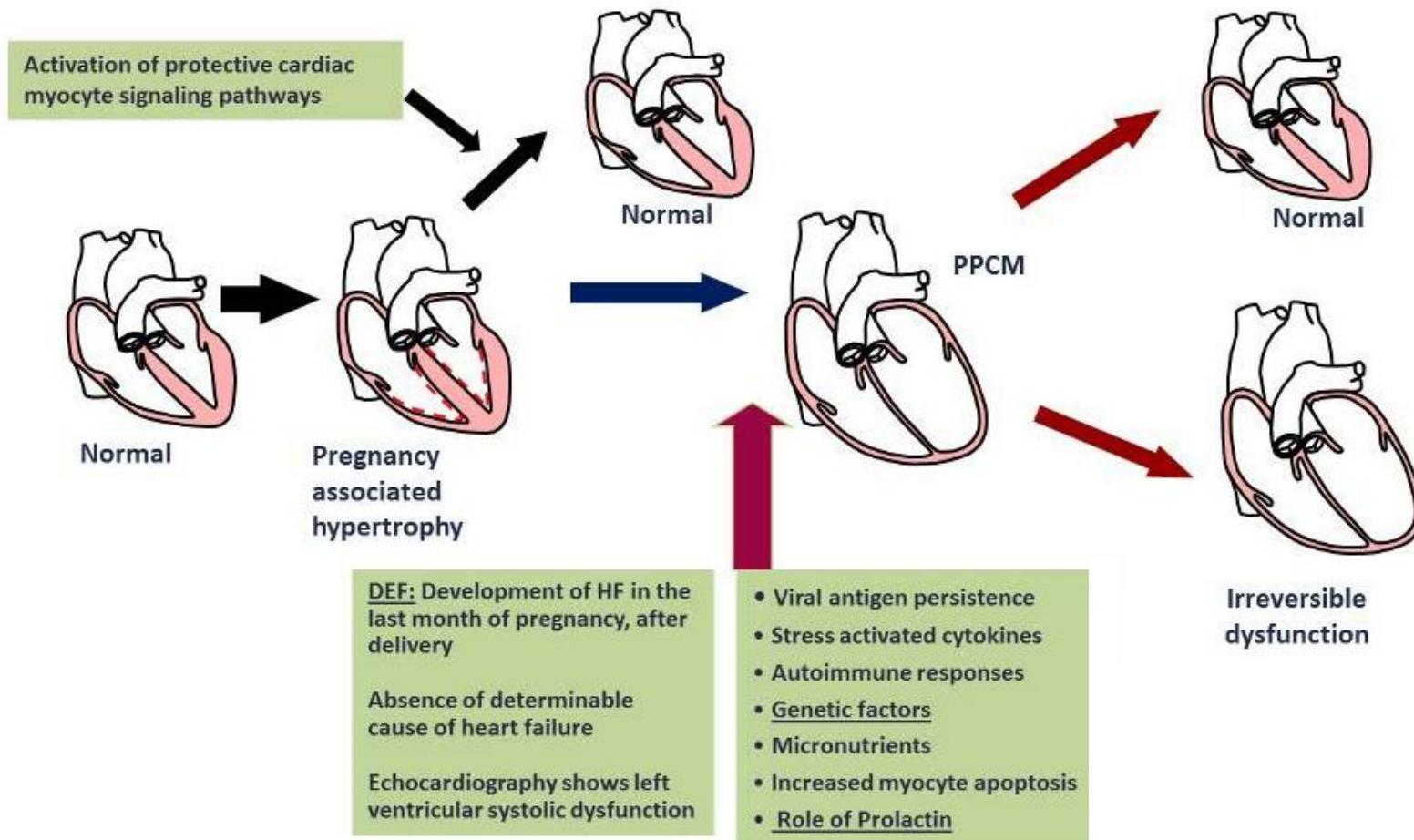
Mahek Shah, MD<sup>a,1</sup>, Pradhum Ram, MD<sup>b,1,\*</sup>, Kevin Bryan Lo, MD<sup>b</sup>, Soumya Patnaik, MD<sup>c</sup>, Brijesh Patel, DO<sup>a</sup>, Byomesh Tripathi, MD<sup>e</sup>, Shantanu Patil, MD<sup>d</sup>, Marvin Lu, MD<sup>b</sup>, Ulrich P. Jorde, MD<sup>f</sup>, and Vincent M. Figueiredo, MD<sup>g</sup>

6,977 admissions with PPCM (30d PP national database)





# Pathogenesis of PPCM



# Genetic theory

A novel human S10F-Hsp20 mutation induces lethal peripartum cardiomyopathy.

## Author(s)

Liu, GS, Gardner, G, Adly, G, Jiang, M, Cai, WF, Lam, CK, Alogaili, F, Robbins, N, Rubinstein, J, Kranias, EG  
Publication name  
Journal of cellular and molecular medicine

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Decreased expression of programmed death 1 on peripheral blood lymphocytes disrupts immune homeostasis in peripartum cardiomyopathy

Guozhi Xia <sup>a</sup>, Xin Sun <sup>b</sup>, Xiaopu Zheng <sup>a,\*</sup>, Junkui Wang <sup>c</sup>



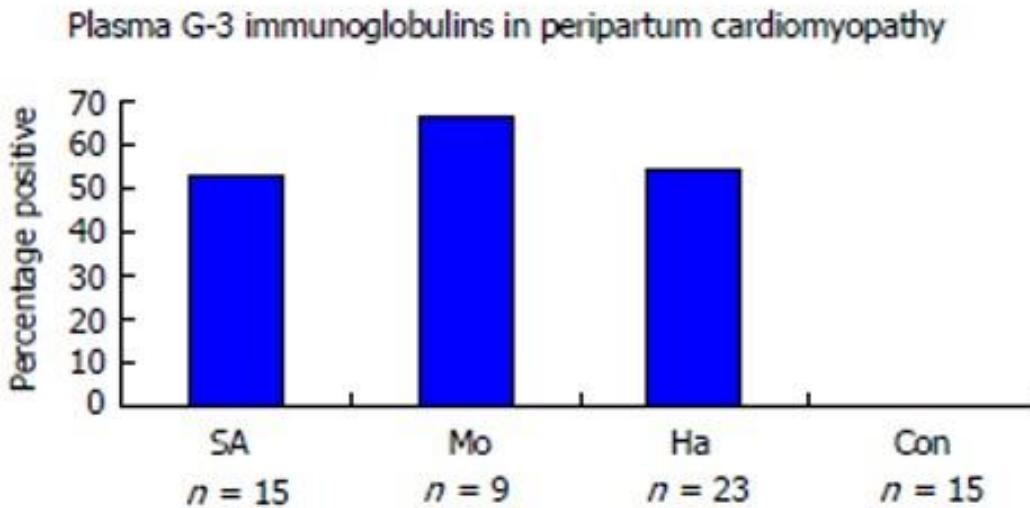
Circulation Journal  
Official Journal of the Japanese Circulation Society  
<http://www.j-circ.or.jp>

REVIEW

Peripartum Cardiomyopathy From a Genetic Perspective

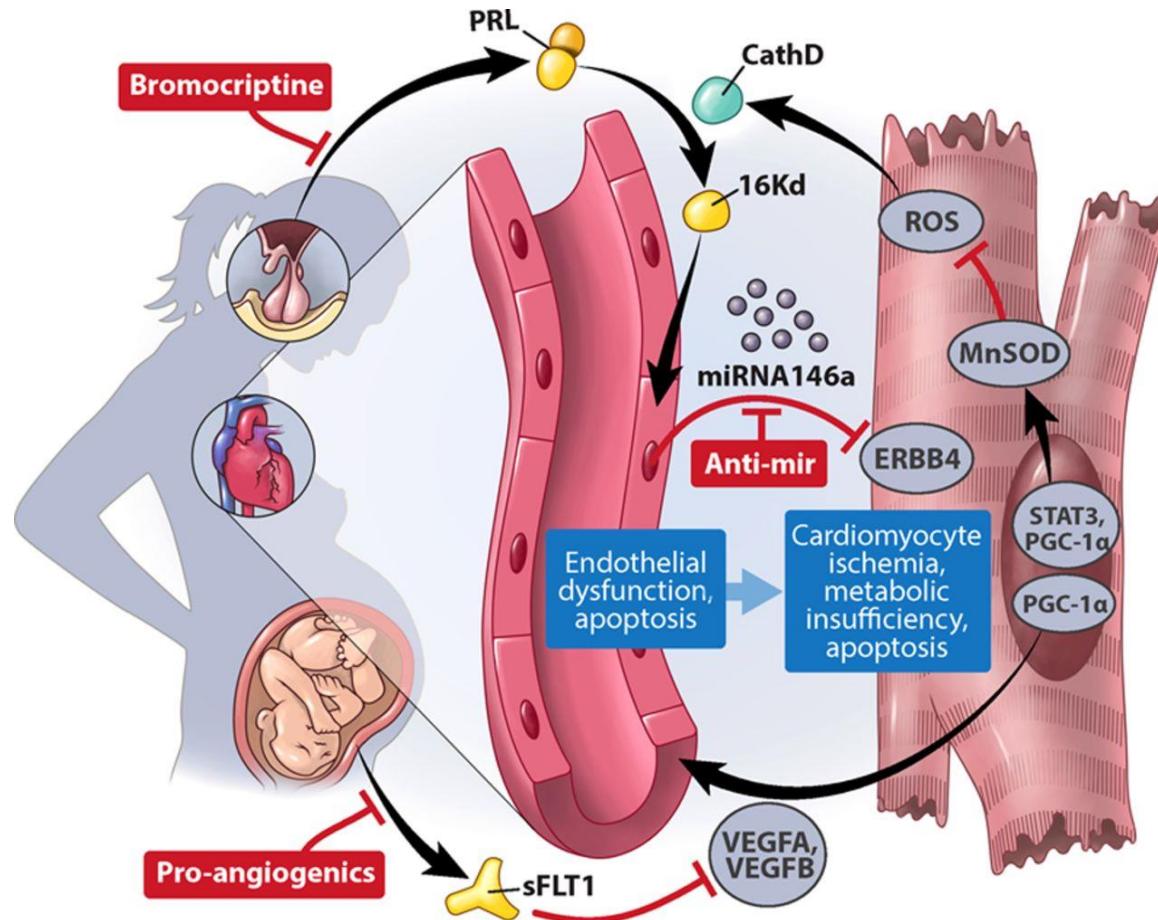
Chizuko A. Kamiya, MD, PhD; Jun Yoshimatsu, MD, PhD; Tomoaki Ikeda, MD, PhD

# Autoimmune theory



- **Multiple types of cardiac antigen antibodies**
- Cardiac myosin heavy chain antibodies in PPCM patients from two African nations.  
None were found in control normal postpartum patients.

# Vascular imbalance theory



**Fig. 1.** Vasculo-hormonal hypothesis of the pathophysiology of PPCM. anti-mir, antibody to miRNA146a; CathD, cathepsinD; ERBB4, avian erythroblastic leukemia viral oncogene homolog 4; miRNA, microRNA; PRL, prolactin; ROS, reactive oxygen species; STAT3, signal transducer and activator of transcription 3. (From Arany Z, Elkayam U. Peripartum cardiomyopathy. Circulation 2016;133(14):1404; with permission.)



## Bromocriptine

2.5 mg daily for 7 days  
higher / longer treatment in  
severe cases

## Oral HF Drugs

Beta-blocker and ACE-inhibitor  
after stabilisation,  
MRA after ablation



## Diuretics

Furosemide iv

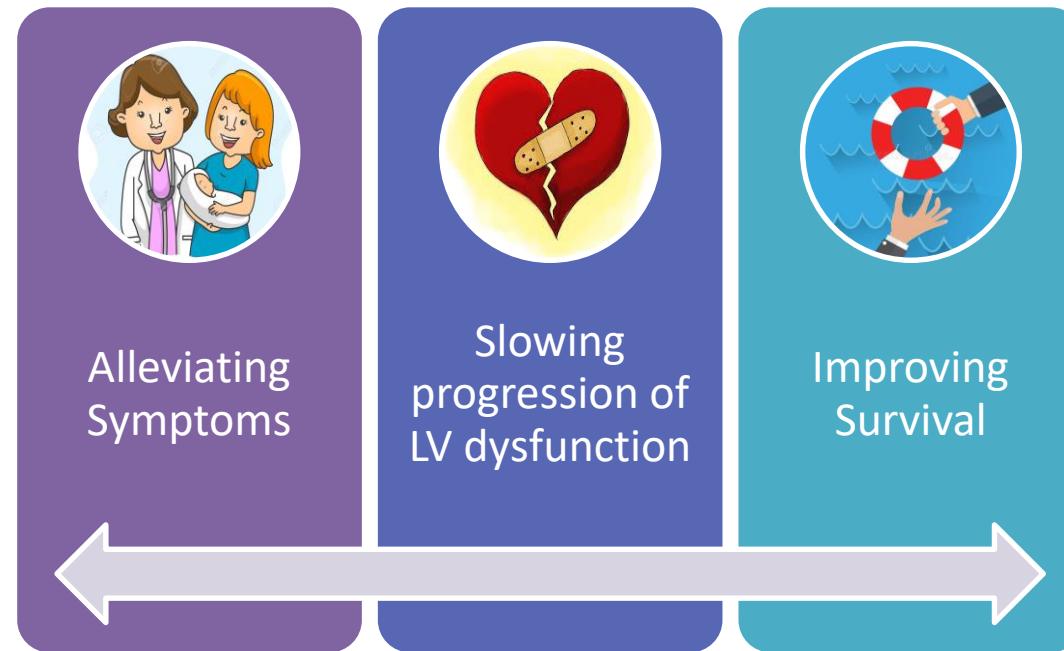
## Relaxants

Vasodilators iv if SBP > 110 mmHg

## Anticoagulation

Heparin immediately  
in at least prophylactic dose

# Management



## Goals

- Treat hypertension
- Fluid restriction
- Dietary salt restriction
- Routine exercise postpartum if stable

## Drugs for routine use

- Diuretics
- $\beta$ -Blockers
- Vasodilators

## Therapies in selected patients

- Aldosterone antagonists
- Digoxin
- Anticoagulation
- Implantable defibrillators
- Biventricular pacing
- Inotropes
- LVAD/cardiac transplantation

**Table 2**  
**Self-assessment tool in identifying women at risk for potential relapse**

Symptom/Sign	0 Points	1 Point	2 Points
Orthopnea	None	Need to elevate head only	Need to elevate body $>45^\circ$
Dyspnea	None	When climbing $\geq 8$ stairs	Walking level
Unexplained cough	None	Night time	Day and night
Pitting edema	None	Below knee	Above and below knee
Weight gain (9th mo)	$\leq 907$ g/wk	907–1814 g/wk	$>1814$ lbs/wk
Palpitations	None	When lying down	Any position day and night

Scoring and action: 0 to 2 low risk, observe. 3 to 4 mild risk, consider BNP/Hs-CRP → ECHO if abnormal.  $\geq 5$  high risk, BNP, Hs-CRP, ECHO.

From Fett JD. Personal commentary: monitoring subsequent pregnancy in recovered peripartum cardiomyopathy mothers. Crit Pathw Cardiol 2009;8(4):174; with permission.



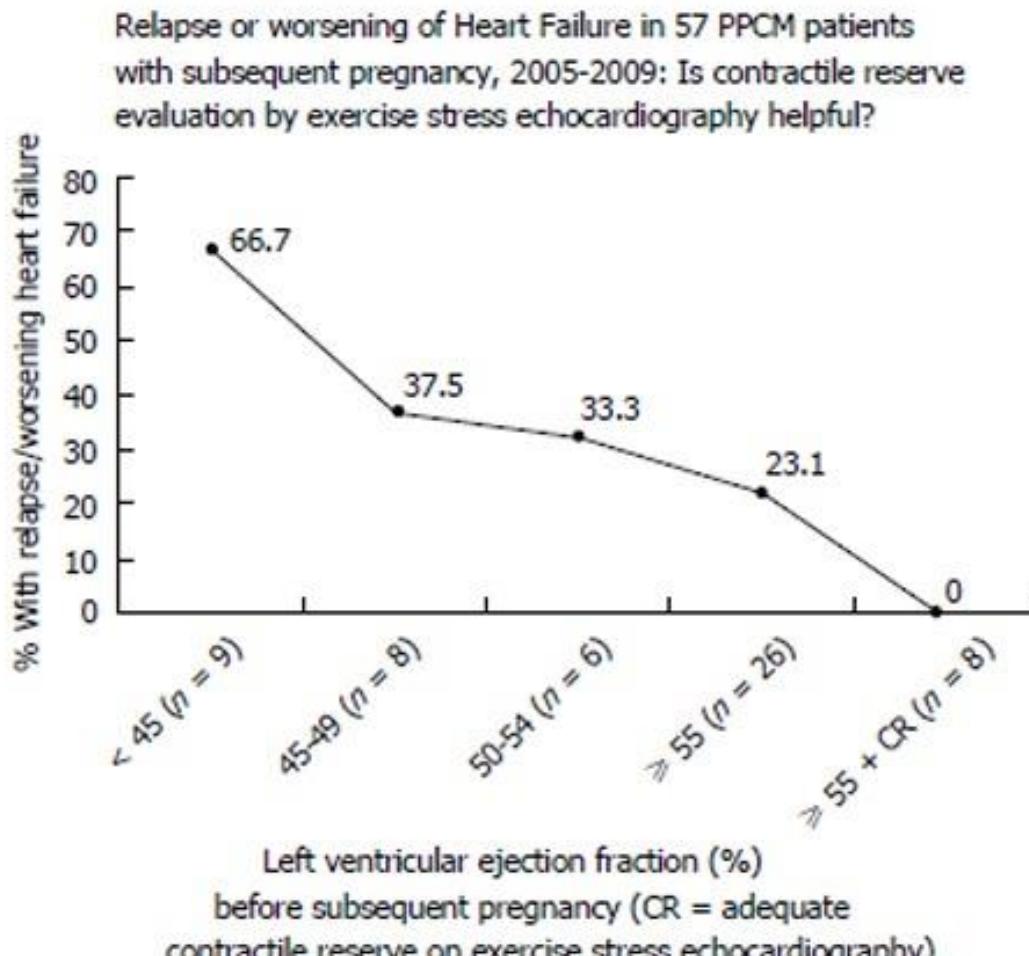
43 yo...  
Anti-hypertensives  
B Blockers  
EF=35%

Pre-gestational  
consult

“Pregnancy is  
**NOT**  
recommended”



# Risk for relapse of heart failure in a post PPCM pregnancy



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## MATERNAL AND FETAL OUTCOMES OF SUBSEQUENT PREGNANCIES IN WOMEN WITH PERIPARTUM CARDIOMYOPATHY

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ILYAS S. KARAALP, M.D., OMAR R. WANIS, M.D., AFSHAN HAMEED, M.D., ISRAEL GVAZDA, B.S.,  
AND ARAHAB SHOTAN, M.D.

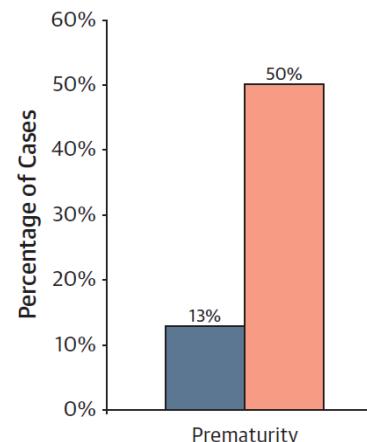
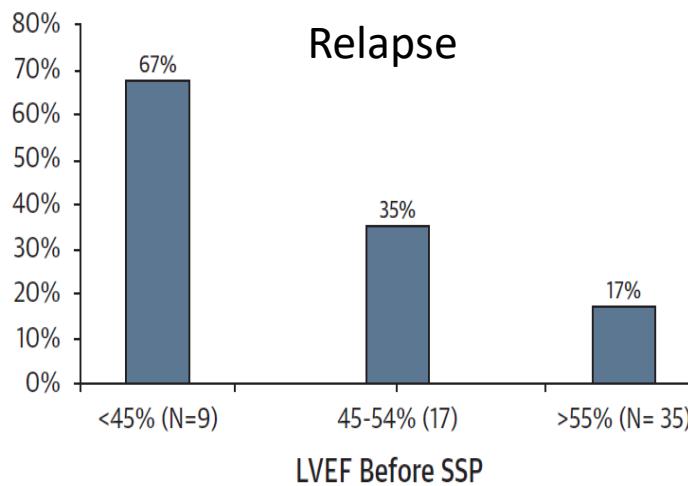
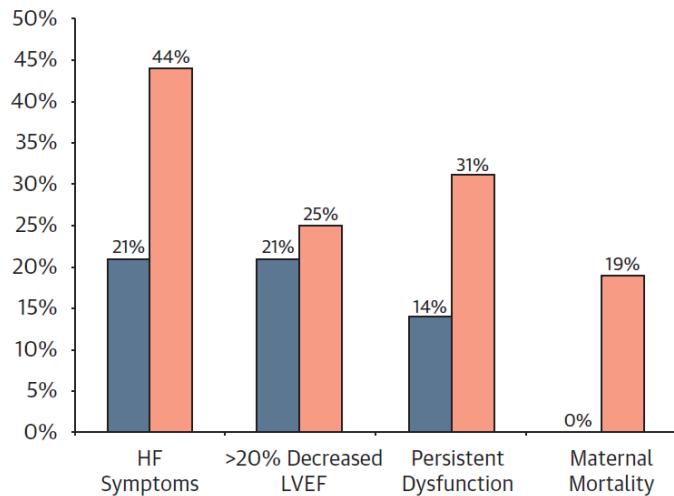
**TABLE 1.** INCIDENCE OF MATERNAL COMPLICATIONS DURING  
THE FIRST SUBSEQUENT PREGNANCY IN WOMEN WHO HAD  
HAD PERIPARTUM CARDIOMYOPATHY.\*

GROUP	No. of WOMEN	SYMPTOMS OF HEART FAILURE	>20% DECREASE IN LVEF	DECREASED LVEF AT FOLLOW-UP	DEATH
		no. of women (%)			
All women	44				
Group 1	28	6 (21)	6 (21)	4 (14)	0
Group 2	16	7 (44)	4 (25)	5 (31)	3 (19)†
Women who did not have abortions	35				
Group 1	23	6 (26)	4 (17)	2 (9)	0
Group 2	12	6 (50)	4 (33)	5 (42)	3 (25)‡

# Risk of Subsequent Pregnancy in Women With a History of Peripartum Cardiomyopathy

All      NI LVF      HF  
       

Uri Elkayam, MD



Group	Maternal Outcome			Fetal Outcome		
	No Relapse	Relapse	Death	Live Birth	Abortions	Stillbirth
Group A	74.4%	23.3%	2.3%	93%	4.7%	2.3%
Group B	37.5%	54.2%	8.3%	83.3%	16.7%	0%

**TABLE 3** Patients With Persistent Left Ventricular Dysfunction Before Subsequent Pregnancy

First Author (Ref. #)	Year	No. of Pregnancies	Deterioration of LV Function	Symptoms of Heart Failure	Persistently Decreased LVEF at Follow-Up	Death
Elkayam (10)	2001	12	4 (33)	6 (50)	5 (42)	3 (25)
Avila (12)	2002	9	NA	4 (44)	2 (22)	1 (11)
Sliwa (13)	2004	4	4 (100)	4 (100)	2 (50)	2 (50)
Chapa (14)	2005	4	0 (0)	0 (0)	4 (100)	0 (0)
Fett (15)	2006	16	8 (50)	8 (53)	7 (44)	1 (6)
Habli (16)	2008	10	9 (53)	NA	5 (29)	1 (6)
Hilfiker-Kleiner (18)	2007	12	5 (42)	NA	6 (50)	3 (25)
Fett (19)*	2010	26	10 (46)	NA	5 (80)	1 (0.4)*
Total		93	40/84 (48)	22/45 (49)	36/93 (39)	11/67 (16)

Values are n (%). \*Most patients identified by an Internet support group of living patients with a history of peripartum cardiomyopathy; mortality rate was therefore not available.

Abbreviations as in Table 2.

**TABLE 4** Patients With Normal Left Ventricular Function Before Subsequent Pregnancy

First Author (Ref. #)	Year	No. of Pregnancies	Deterioration of LV Function	Symptoms of Heart Failure	Persistently Decreased LVEF at Follow-Up	Death
Elkayam (10)	2001	23	4 (17)	6 (20)	2 (50)	0 (0)
Avila (12)	2002	6	NA	0 (0)	NA	0 (0)
Sliwa (13)	2004	2	2 (10)	2 (100)	2 (100)	0 (0)
Chappa (14)	2005	4	4 (100)	4 (100)	3 (75)	0 (0)
Habli (16)	2008	21	NA	6 (28)	NA	0 (0)
Chee (21)	2009	2	0 (0)	0 (0)	0 (0)	0 (0)
Fett (19)*	2010	35	8 (17)	NA	1 (12)	-
Mandal (20)	2010	5	NA	2 (40)	1 (50)	0 (0)
Total		98	18/66 (27)	20/63 (32)	9/71 (13)	0/63 (0)

Values are n (%). \*Most patients in this study were identified by an Internet support group of living patients with history of peripartum cardiomyopathy; mortality rate was therefore not available.

Abbreviations as in Table 2.

Peripartum Cardiomyopathy (PPCM)  
(Idiopathic form of left ventricular [LV] systolic dysfunction develops during pregnancy or post-partum)

### PERSISTENT LV DYSFUNCTION

Higher risk of relapse with subsequent pregnancies (SSP)

~50% show further deterioration in LV dysfunction

Increased morbidity and mortality with SSP

Premature delivery and abortion more common

### COMPLETE RECOVERY

Better prognosis with SSP compared to persistent LV dysfunction

~20% have a relapse

Rate of recovery is higher, morbidity and mortality is lower

Likely to have a normal pregnancy

Careful and close monitoring recommended

### CENTRAL ILLUSTRATION Risks in Subsequent Pregnancies in PPCM

Varied pathways for patients who develop an idiopathic form of left ventricular (LV) systolic dysfunction during pregnancy or post-partum. PPCM = peripartum cardiomyopathy; SSP = subsequent pregnancy.



43 yo...  
Anti-hypertensives  
B Blockers  
EF=35%

Pre-  
gestational  
consult

"Pregnancy is  
**NOT**  
recommended"



Pregnancy...



High-Risk follow-up



Repeated echo



32w – Deterioration: EF, LV function



Steroids for FLM



Cesarean Delivery



Healthy baby, Maternal Partial Recovery



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