VI Congresso Nazionale di
Ecocardiochirurgia
Milano, 15-17 ottobre 2012

La cardiomiopatia peripartum. La diagnosi, la valutazione funzionale, la prognosi.

Un problema raro che può mettere in difficoltà anche cardiologi esperti. Come orientarsi praticamente?

Daniela Degli Esposti

Dipartimento di Medicina Interna, dell'Invecchiamento e Malattie Nefrologiche – U.O. Medicina Interna-Borghi – Policlinico S.Orsola-Malpighi – Università di Bologna

…no single institution sees a large enough number of patients with peripartum cardiomyopathy to conduct any significant research on this entity

Veille and Zaccaro, Am J Obstet Gynecol 1999

…the practing cardiologist may see only a few cases during his or her career

Lampert and Lang, Am Heart J 1995

PERIPARTUM CARDIOMIOPATHY: DEFINITION

Table 1 Definition of peripartum cardiomyopathy

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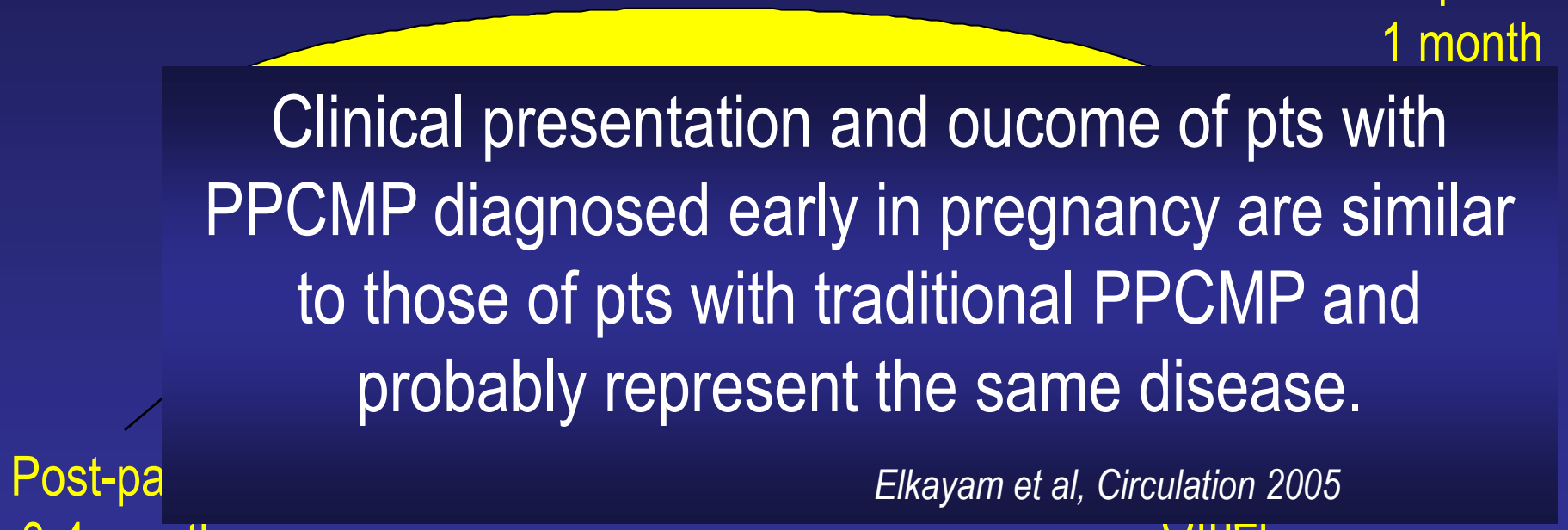
ONSET OF PPCMP IN A POOL OF 13 STUDIES BASED ON 419 CASES



From Lamper MB & Lang RM, Am Heart J 1995

CARDIOMIOPATIA PERIPARTUM : caso clinico

- *38 anni, 2° gravidanza (nessun problema alla 1° gravidanza, parto cesareo x presentazione podalica), familiarità x CI*
- *12° settimana di gestazione: …*
- *37° settimana di gestazione:*
  - *moderata astenia, riduzione tolleranza allo sforzo, NYHA II*
  - *Ecocardiogramma: DTDVsn 65 mm, DTSVsn 56 mm, VTDVsn 223.6 ml, VTSVsn 153.6 ml, AF 15%, FE 32%, aspetto globoso del Vsn, IM L/M*
- *38° settimana di gestazione: parto cesareo, M 3.920 g, senza segni di sofferenza fetale*
- *Dopo il parto: benessere soggettivo, allattamento al seno*
- *2 mesi dopo il parto:*
  - *dispnea x sforzi lievi, ortopnea, astenia marcata, notevole limitazione dell'attività fisica, classe NYHA III*
  - *FC 120b/min, T3, soffio olosistolico 3/6 L, ECG tachicardia sinusale, BBsn completo*
  - *Ecocardiogramma: DTDVsn 68 mm, AF 15%, FE 32%, IM severa, IT lieve, PVdx 35 mmHg, pattern di riempimento ventricolare restrittivo*

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  - *Ricovero ospedaliero: valutazione emodinamica/coronarografia: CMPD, coronarie indenni*
  - *Inizia tp con ACE-I, beta-bloccante, diuretico e digitale*
- *successivi controlli: progressivo miglioramento clinico/strumentale*
- *20 mesi dopo il parto: asintomatica, ECG: BBsn, Ecocardio: DTDVsn 55 mm, AF 25%, FE 48%, IM moderata, IT minima, PVdx normale, Tp: betabloccante e ARB*

CARDIOMIOPATIA PERIPARTUM : caso clinico

- *38 anni, 2° gravidanza (nessun problema alla 1° gravidanza, parto cesareo x presentazione podalica), familiarità x CI*
- *12° settimana di gestazione: riscontro occasionale di BBsn all'ECG*
- *Asintomatica, h 165 cm, peso 70 Kg, PA 120/70, lieve anemia siderocarenziale*
- *Ecocardiogramma: DTDVsn 54 mm, DTSVsn 41 mm, VTDVsn 141.3 ml, VTSVsn 74.2 ml, AF 24%, FE 48%, aspetto tendenzialmente globoso del Vsn, Asn 43 mm, IM L/M*
- *Prescritto ASA 100 mg*
- *Controlli clinici mensili, controlli ecocardiografici bimestrali*
- *Stabilità clinica e normali parametri ostetrici (indici di resistenze vascolari alla flussimetria delle arterie uterine e indici di accrescimento fetale)*
- *Progressivo peggioramento degli aspetti strutturali e funzionali del Vsn all'ecocardiogramma*

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Heart Failure Association of the European Society of Cardiology Working Group on PPCM 2010[2]	2010	PPCM is an idiopathic cardiomyopathy presenting with heart failure secondary to LV systolic dysfunction towards the end of pregnancy or in the months following delivery, where no other cause of heart failure is found. It is a diagnosis of exclusion. The left ventricle may not be dilated but the ejection fraction is nearly always reduced below 45%

INCIDENCE OF PPCMP *Blauwet LA & Cooper LT, Heart 2011*

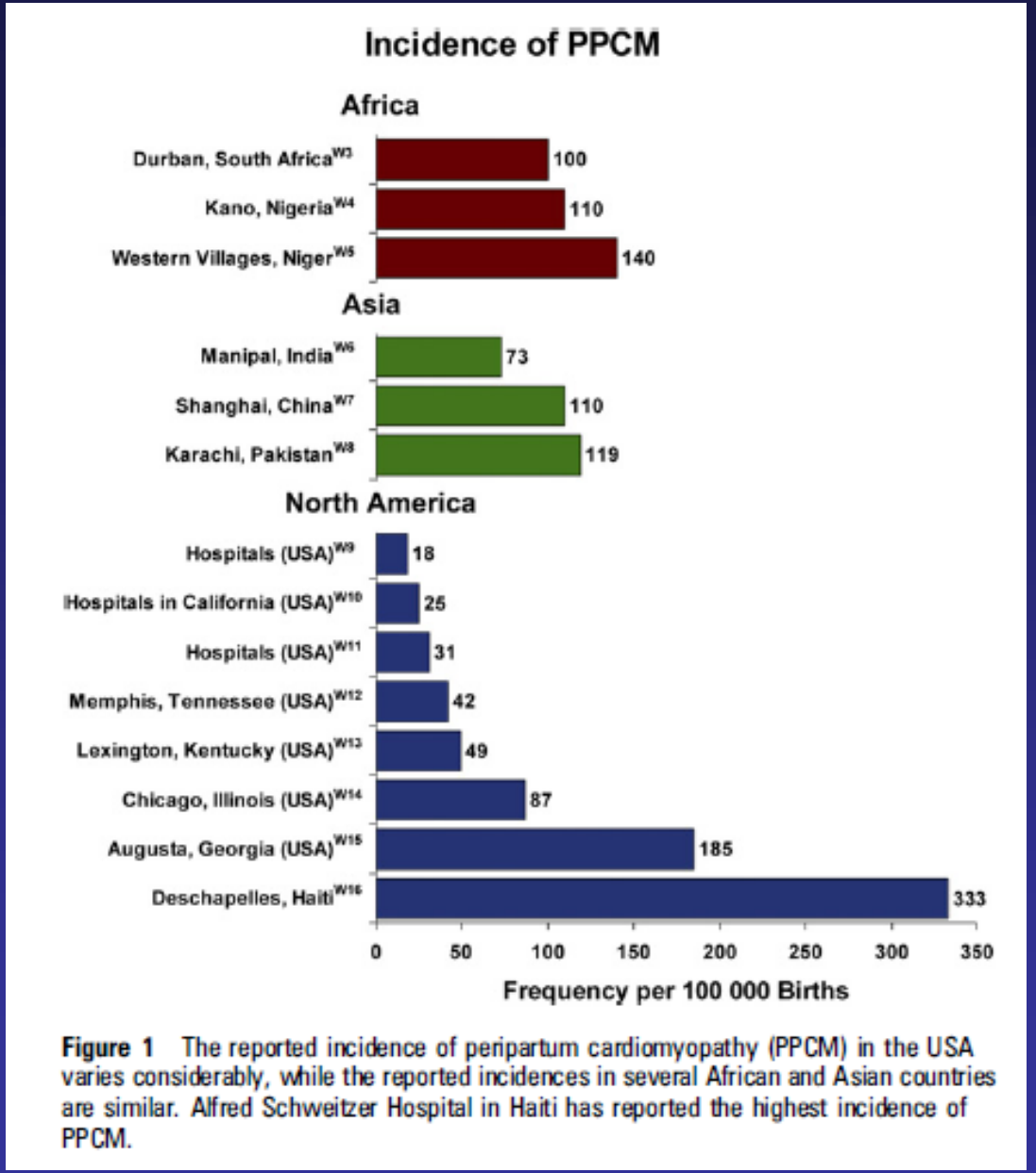

Figure 1 The reported incidence of peripartum cardiomyopathy (PPCM) in the USA varies considerably, while the reported incidences in several African and Asian countries are similar. Alfred Schweitzer Hospital in Haiti has reported the highest incidence of PPCM.

PERIPARTUM CARDIOMYOPATHY: EPIDEMIOLOGY

Incidence (marked geographical and ethnic variation in the incidence of PPCMP)

- It is not known (no population-based estimates)
- Presumably: 1/3000 - 1/4000 live births (1/1485-1/15.000)
- Black women > White women (15:1)
- **Greater risk:** Black african population (1/100 in the Nigerian Hausa tribe - ingestion of kanwa)

National Heart, Lung, Blood Institute and Office of Rare Diseases Workshop on PPCMP, JAMA 2000

PERIPARTUM CARDIOMIOPATHY

…rimane un gruppo eterogeneo con aspetti molto vari dal punto di vista epidemiologico nonostante i tentativi di restringere i criteri diagnostici…

Fattori di rischio ed etiologia della PPCMP

Blauwet LA & Cooper LT, Heart 2011

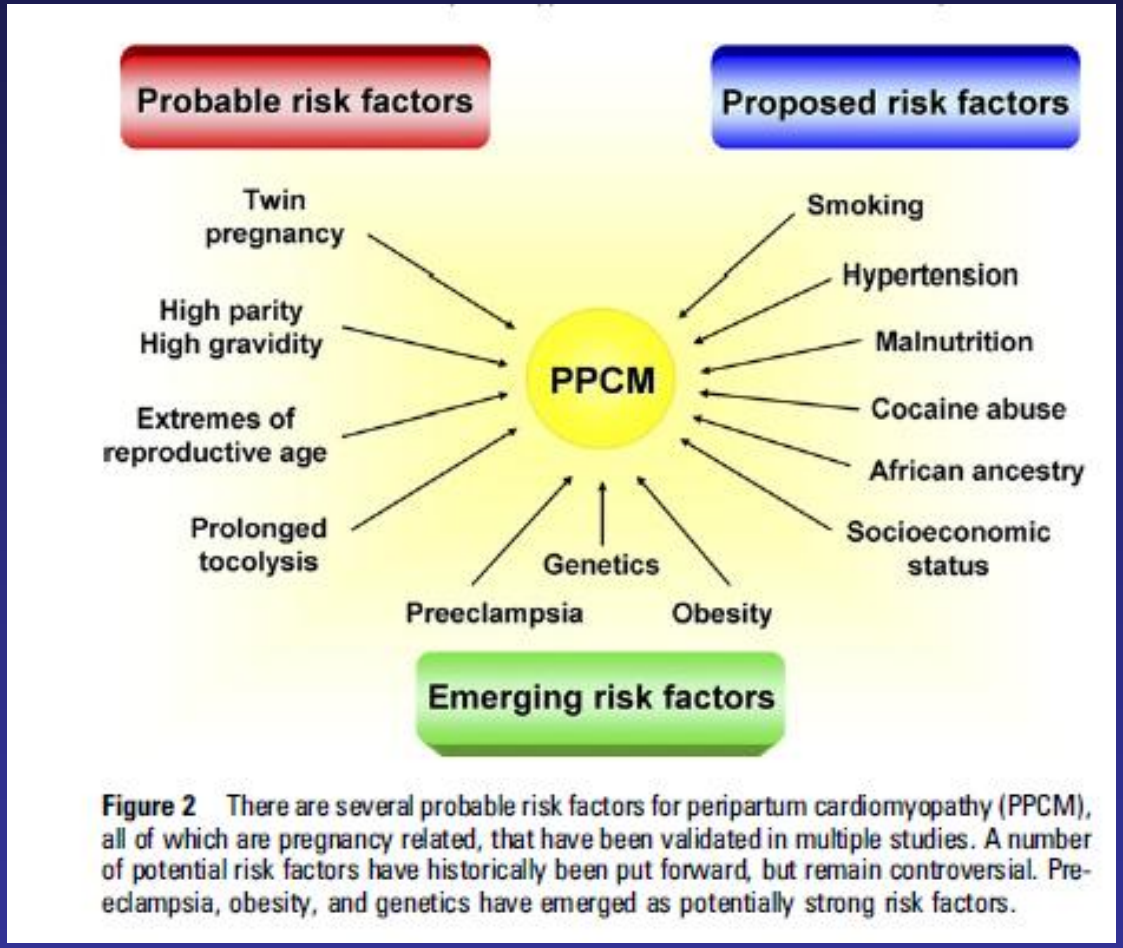

Figure 2 There are several probable risk factors for peripartum cardiomyopathy (PPCM), all of which are pregnancy related, that have been validated in multiple studies. A number of potential risk factors have historically been put forward, but remain controversial. Pre-eclampsia, obesity, and genetics have emerged as potentially strong risk factors.

Fattori di rischio ed etiologia della PPCMP

Fattori di rischio

- Multiparità
- Gravidanza gemellare (13%)
- Maggiore prevalenza nella razza africana (associazione razziale o maggiore frequenza nei ceti meno abbienti?)
- PIH, PE (43%)
- Familiarità

Etiologia

- Ipotesi immunogenetica (sostenuta anche dall'esordio prevalente nell'immediato postparto)
- Deficit nutrizionali (selenio, calcio)
- Infezioni virali
- Ingestione di Kanwa ed esposizione a fango caldo-umido delle donne nigeriane (carico salino, ipertensione…)
- Risposta maladattativa allo stress emodinamico della gravidanza (aspetti fisiopatologici comuni: squilibrio angiogenico, stress ossidativo…prolattina)
- Forme fruste di predisposizione genetica alla CMPDI
- Trasmissibilità con l'embrione (un caso di PPCMP in "utero in affitto" x embrione di donna con pregressa PPCMP, un caso di HF in primigravida attempata da donazione di ovocita/embrione)

PERIPARTUM CARDIOMIOPATHY

…identità nosografica distinta

…o parte del gruppo delle CMPDI

CLINICAL AND HEMODYNAMIC COMPARISON BETWEEN PPCMP AND IDCMP

	PPCMP	IDCMP
No of patients	14	55
Duration symptoms (mo)	4.1±8*	19±18
Ejection Fraction (%	18.1±10	18.8±9
LVEDD (cm)	6.2±1.1	6.9±1
Cardiac index (L/min/m^2)	2.7±1	2.2±0.6
Complex arrhythmias	6 (60%)	42 (76%)

*P<0.001

O'Connell JB et al, JACC 1986

POSSIBLE CAUSES OF LV DYSFUNCTION OR HEART FAILURE CONFOUNDING THE DIAGNOSIS OF PPCMP IN PREGNANCY

General

- Chronic HBP heart disease
- Congenital heart disease
- Rheumatic heart disease
- Pulmonary disease
- Thromboembolism
- Sepsis

Obstetric

- Preeclampsia
- Excess administration of tocolytic agents
- Fluid overload after steroid use
- Amniotic fluid embolism

Because many of the signs and symptoms are similar to those of normal pregnancy and early postpartum period, the diagnosis of PPCMP can easily be missed

Diagnosis remains a challenge

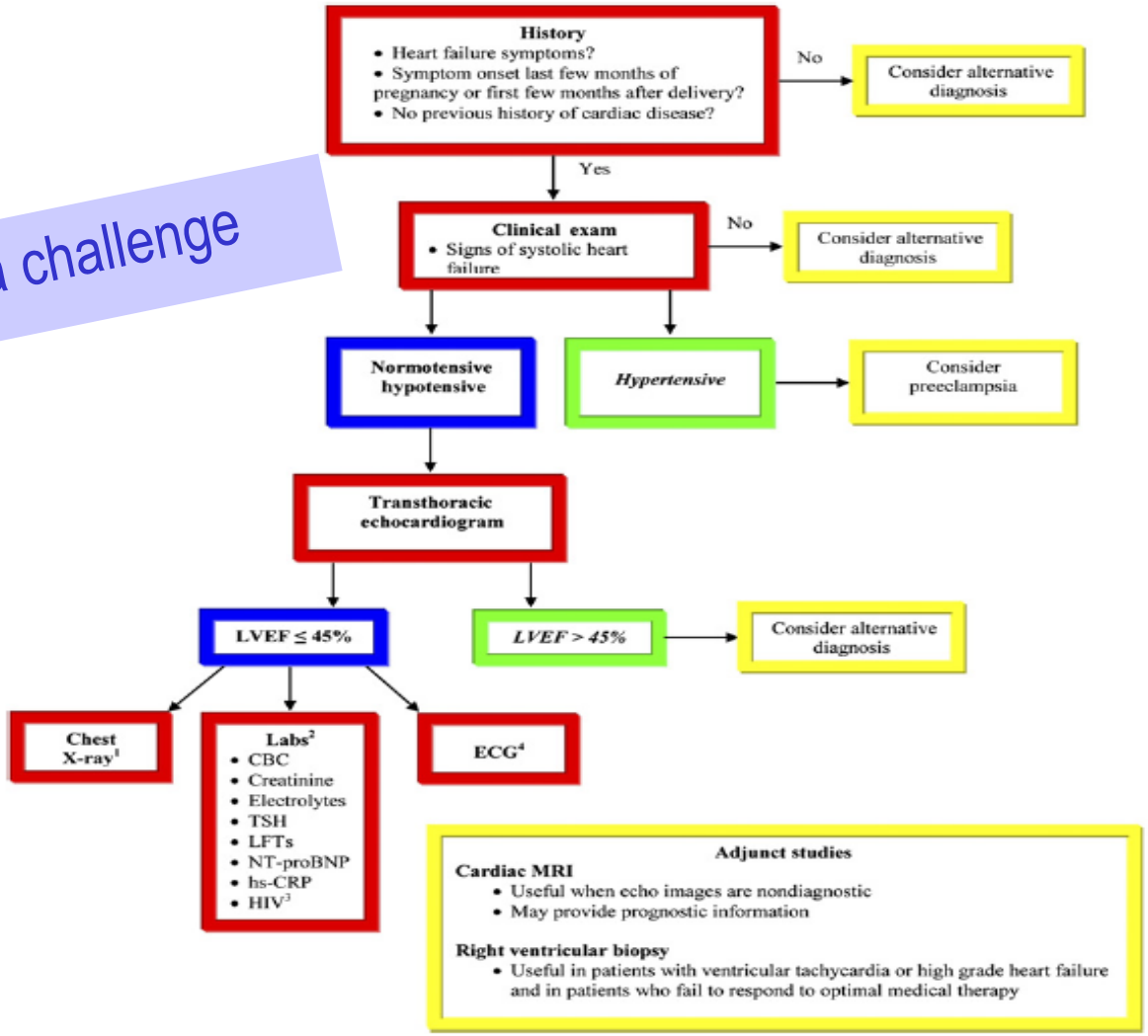

Figure 4 Diagnostic algorithm for peripartum cardiomyopathy. [1]Useful for assessing other possible aetiologies for symptoms including pneumonia and pneumothorax. [2]Useful for assessing other possible aetiologies for symptoms including severe anaemia, thyroid disease, liver disease, end stage renal disease, and infection. [3]Obtain in select cases. [4]Useful for assessing other possible aetiologies, including coronary artery dissection or thrombosis, and may provide prognostic information. CBC, complete blood count; hs-CRP, high sensitivity C reactive protein; LFTs, liver function tests; LVEF, left ventricular ejection fraction; NT-proBNP, N-terminal pro B-type natriuretic peptide; TSH, thyroid stimulating hormone.

Heart 2011;**97**:1970—1981. doi:10.1136/heartjnl-2011-300349

PERIPARTUM CARDIOMIOPATHY: DIFFERENCES WITH IDMP

- Younger age
- Better prognosis
- Higher incidence
- Mostly postpartum (IDCMP usually manifests by the 2° trimester)
- Exclusively pregnant women or peripartum period
- Varying types of hemodynamic patterns
- Unique sets of antigenes and antibodies against myocardium
- Higher incidence of myocarditis (29-78%)
- Heart size returns to normal after delivery in a greater percentage of pts
- More rapid worsening of clinical conditions

Pradipta Bhakta et al, Yonsei Med J 2007

PERIPARTUM CARDIOMIOPATHY

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PERIPARTUM CARDIOMIOPATHY: TREATMENT

- PRE-PARTUM
  - Early delivery
  - Digoxin
  - Vasodilators (Hydralazine)
  - Diuretics
  - CCB (if hypertension)
  - ß-blockers (if necessary, low fetal growth)
  - Anthycoagulants (heparin)
- POST-PARTUM
  - Digoxin
  - Diuretics
  - Vasodilators
  - **Ace-inhibitors or ARBs**
  - CCB (if hypertension)
  - ß-blockers
  - Anthycoagulants (warfarin)

Excretion of drugs and metabolytes
during breast feeding

PERIPARTUM CARDIOMIOPATHY: TREATMENT

– ADDITIONAL TREATMENTS

Experimenta therapy:

- Immunosuppressive therapy
  - Limited to myocarditis (biopsy) and not improved after 2 weeks of treatment
- Intravenous immunoglobilin
- Pentoxifylline
- Bromocriptine

Implantable cardioverter-defibrillator

Cardiac assist devices

Cardiac transplantation

- Survival (?)
  - » = Other CMP (Keogh, 1994)
  - » Reduced survival
- Early rejection

Peripartum cardiomyopathy, an autoimmune manifestation of allograft rejection?

Norbert Gleicher [a,b,*], Uri Elkayam [c]

[a] From the Center for Human Reproduction and the Foundation for Reproductive Medicine, New York, NY, United States
[b] Department of Obstetrics, Gynecology and Reproductive Sciences, Yale University School of Medicine, New Haven, CT, United States
[c] Department of Medicine, Division of Cardiology Heart Failure Program and the Department of Obstetrics and Gynecology, University of Southern California, Keck School of Medicine, United States

Take-home messages

- There is convincing circumstantial evidence in support of abnormal autoimmune function as an underlying pathophysiologic mechanism in the development of PPCM.
- It is, however, tempting to speculate further that these observed autoimmune responses are not representative of an autoimmune condition, but reflect the typically ob… autoimmune components of malfu… this case pregnancy-) tol…
- Building on this … which have been … complications, may … times life-threateni…

An improved understanding of the pathophysiological pathway leading to PPCM may result in the development of novel disease-modifying therapies for this condition

Autoimmunity Reviews 8 (2009) 384–387

Peripartum Cardiomyopathy: Recent Insights in its Pathophysiology

Denise Hilfiker-Kleiner*, Karen Sliwa, and Helmut Drexler

Peripartum/Postpartum cardiomyopathy (PPCM) is a serious, potentially life-threatening heart disease of uncertain etiology in previously healthy women. Previous clinical and experimental data have identified inflammation, autoimmune processes, apoptosis, and impaired cardiac (systemic) microvasculature as typical features in the pathophysiology of PPCM. However, recent data have shown that unbalanced peri/postpartum oxidative stress is linked to proteolytic cleavage of the nursing hormone prolactin into a potent antiangiogenic, proapoptotic, and pro-inflammatory factor. These observations strongly suggest that prolactin cleavage can operate as a specific pathomechanism for the development of PPCM. Consistent with these findings, inhibition of prolactin secretion by bromocriptine, a dopamine D2 receptor agonist, prevented the development of PPCM in an animal model of PPCM, and first clinical experience are promising in this respect. Thus, inhibition of prolactin release may represent a novel specific therapeutic approach to either prevent or treat patients with acute PPCM. In this review, we highlighting the current knowledge on risk factors, chanisms, and treatment options for

2008;18:173–179) © 2008

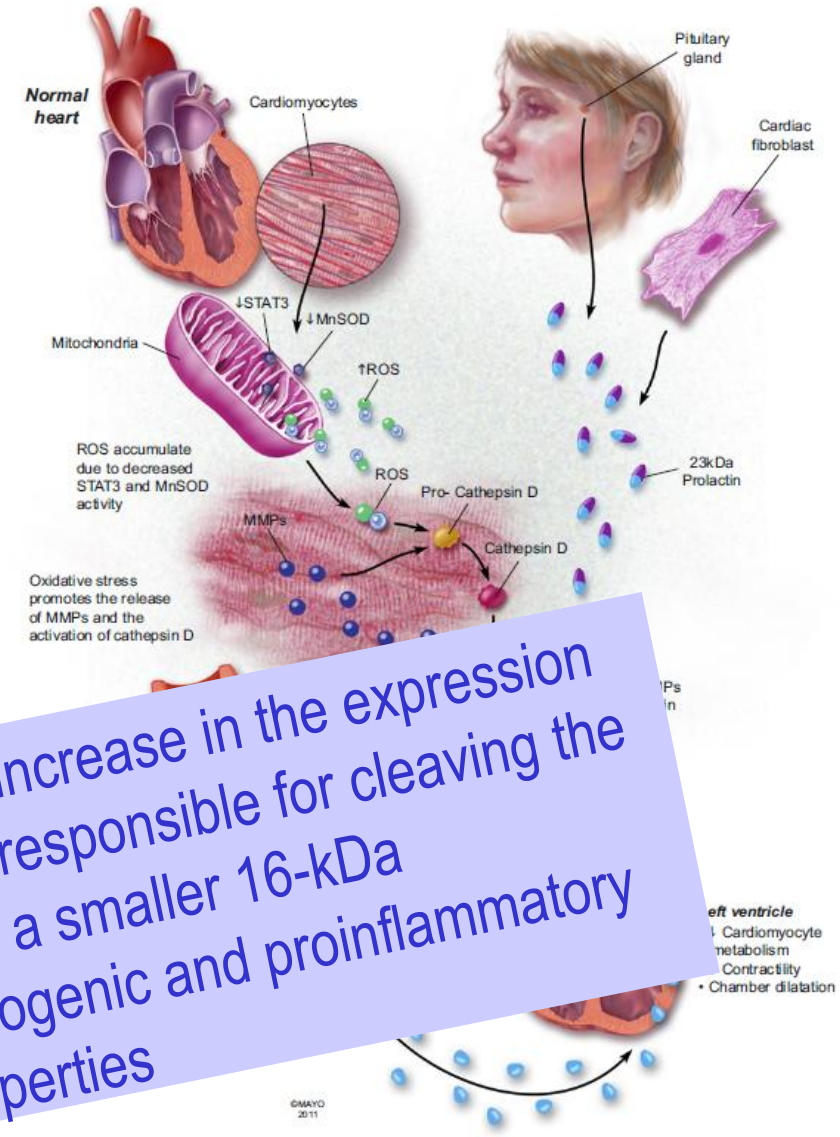

Figure 3 Reduced concentrations of STAT-3 in the cardiomyocyte leads to attenuated expression of the oxygen radical scavenger MnSOD. This leads to accumulation of ROS which promote the release of MMPs and activation of cathepsin D, and prolactin (23 kDa) is produced by the pituitary gland and cardiac fibroblasts. Cathepsin D and MMPs cleave 23 kDa prolactin into its 16 kDa form. Expression of 16 kDa prolactin decreases cardiomyocyte metabolism and destroys endothelial cells, resulting in vasoconstriction, apoptosis, inflammation, and dissociation of capillary structures. MMPs, matrix metalloproteinases; MnSOD, manganese sodium dismutase; ROS, reactive oxygen species; STAT-3, signal transducer and activator of transcription 3. Adapted from Hilfiker-Kleiner *et al*,[11] with permission.

…detrimental effect of oxidative stress…increase in the expression and activity of cathepsine-D, which is responsible for cleaving the 32-kDa form of prolactin to a smaller 16-kDa fragment…vasoconstrictor, antiangiogenic and proinflammatory apoptotic properties

Evaluation of Bromocriptine in the Treatment of Acute Severe Peripartum Cardiomyopathy

A Proof-of-Concept Pilot Study

Karen Sliwa, MD, PhD; Lori Blauwet, MD; Kemi Tibazarwa, MD; Elena Libhaber, PhD; Jan-Peter Smedema, MD, MMed(Int); Anthony Becker, MD; John McMurray, MD, FESC; Hatice Yamac, MD; Saida Labidi, MSc; Ingrid Struman, PhD; Denise Hilfiker-Kleiner, PhD

Background—Peripartum cardiomyopathy (PPCM) is a potentially life-threatening heart disease that occurs in previously healthy women. We identified prolactin, mainly its 16-kDa angiostatic and proapoptotic form, as a key factor in the pathophysiology. Previous reports suggest that bromocriptine may have beneficial effects in women with acute PPCM.

Methods and Results—A prospective, single-center, randomized, open-label, proof-of-concept pilot study of women with newly diagnosed PPCM receiving standard care (PPCM-Std; n=10) versus standard care plus bromocriptine for 2 weeks (PPCM-Br, n=10) was conducted. Because mothers receiving bromocriptine could not breast-feed, the cardiac outcome of their children (n=21) was studied as a secondary end point. Blinded clinical, hemodynamic, and echocardiographic assessments were performed at baseline and 6 months after diagnosis. Cardiac magnetic resonance imaging was performed 4 to 6 weeks after diagnosis in PPCM-Br patients. There were no significant differences in baseline characteristics, including serum 16-kDa prolactin levels and cathepsin D activity, between the 2 study groups. PPCM-Br patients displayed greater recovery of left ventricular ejection fraction (27% to 58%; $P=0.012$) compared with PPCM-Std patients (27% to 36%) at 6 months. One patient in the PPCM-Br group died compared with 4 patients in the PPCM-Std group. Significantly fewer PPCM-Br patients (n=1, 10%) experienced the composite end point of poor outcome defined as death, New York Heart Association functional class III/IV, or left ventricular ejection fraction <35% at 6 months compared with the PPCM-Std patients (n=8, 80%; $P=0.006$). Cardiac magnetic resonance imaging revealed no intracavitary thrombi. Infants of mothers in both groups showed normal growth and survival.

Conclusions—In this trial, the addition of bromocriptine to standard heart failure therapy was associated with improved left ventricular ejection fraction and a composite clinical outcome in women with acute severe PPCM. The number of patients studied was small and the results cannot be considered definitive. Larger randomized studies are in progress to test this strategy more robustly. (***Circulation***. 2010;121

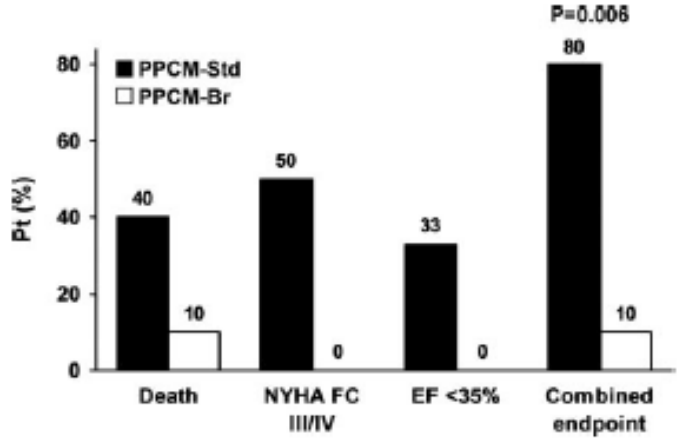

Figure 3. Comparison of 6-month prespecified poor outcome, including death, NYHA functional class (FC) III/IV, and LVEF <35% among survivors, and the combined end point including all 3 of these end points for PPCM-Br vs PPCM-Std patients (Pt).

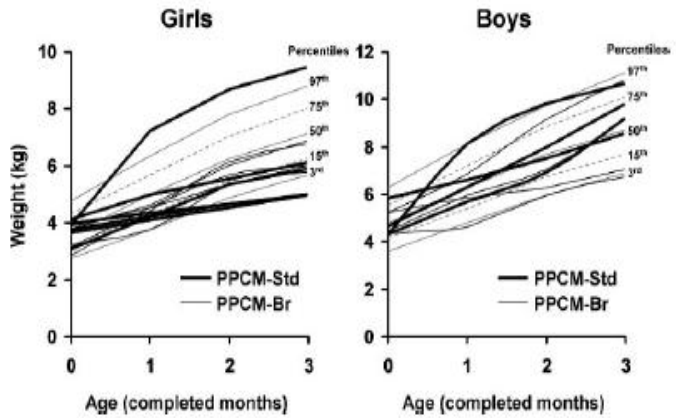

Figure 5. Growth and survival of children of PPCM study mothers from birth to 3 months plotted on World Health Organization growth charts.

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- **Better prognosis**
- Higher incidence
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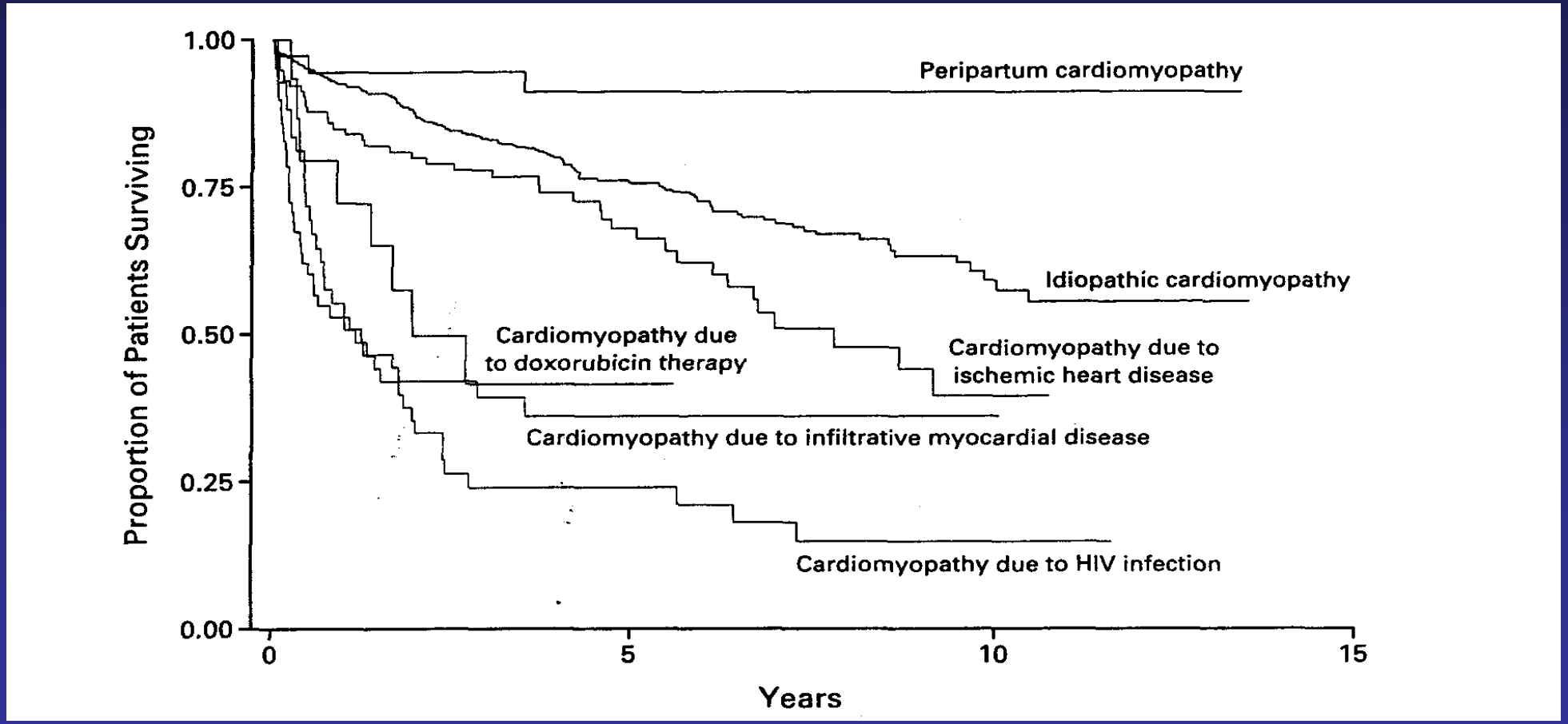
Pradipta Bhakta et al, Yonsei Med J 2007

Adjuste Kaplan-Meier estimates of survival according to the underlying cause of cardiomyopathy

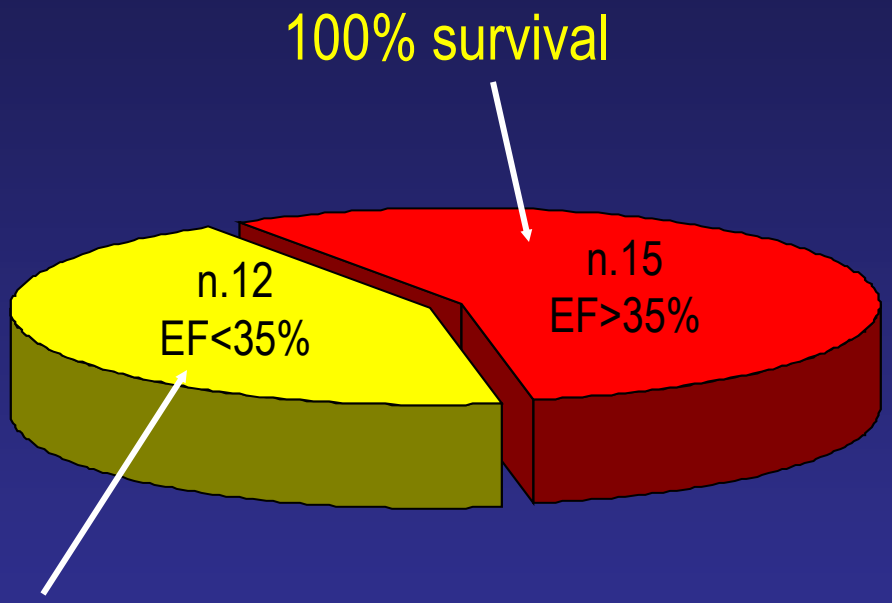


Felker M et al, NEJM 2000

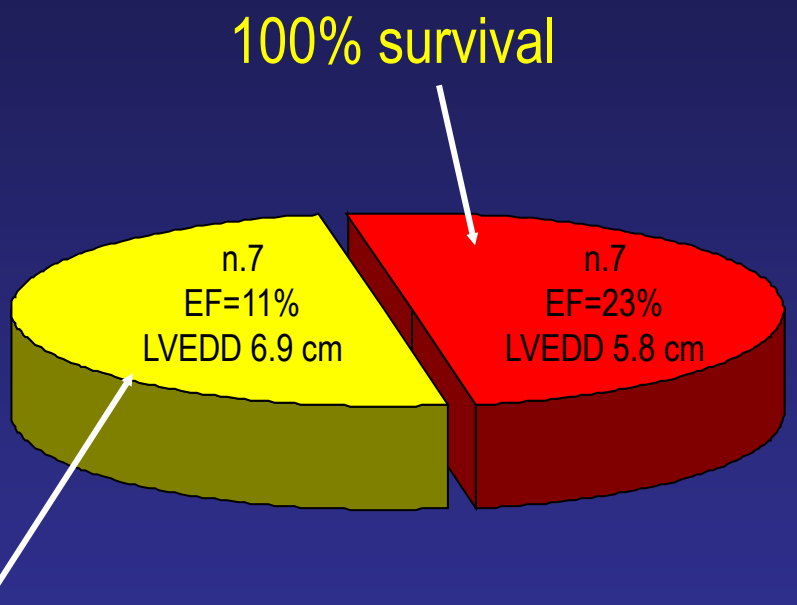
PERIPARTUM CARDIOMIOPATHY: PROGNOSIS

- The prognosis of PPCM depends on the normalization of LV function < 6 mo. after delivery.
- If EF% is < 35% the 10-year actuarial survival rate has been estimated to be 42% (Davis MD, 1992)
- The mortality rate of PPCM has been reported to be 25-50% and most deaths < 3 mo. after diagnosis for progressive CHF, arrhythmias and thromboembolism.
- Patients with PPCM whose LV function recovers have a significanly improved survival.

LONG-TERM PROGNOSIS OF PPCM AND LV FUNCTION



Demakis et al, 1971



St John-Suttom MS, 1991

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Recovery from severe heart failure following peripartum cardiomyopathy ☆

James D. Fett *, Herriot Sannon, Emmeline Thélisma, Therese Sprunger, Venkita Suresh

Department of Adult Medicine, Hôpital Albert Schweitzer, Deschapelles, Haiti

Characteristics of recovered and nonrecovered Haitian peripartum cardiomyopathy patients

Characteristics	Recovered (n = 32)	Nonrecovered (n = 84)	*P* value
Mean age at diagnosis, y (range)	33.8 (17–47)	31.6 (16–50)	NS
Mean parity at diagnosis (range)	4.7 (1–9)	4.3 (1–11)	NS
Mean LVEF at diagnosis (range)	0.28 (0.15–0.40)	0.23 (0.15–0.35)	0.002
NYHA Functional Class III/IV at diagnosis	94% 30/32	92% 77/84	NS

Abbreviations: NYHA, New York Heart Association; LVEF left ventricular ejection fraction.

Length of time required for recovery of left ventricular function in 32 Haitian peripartum cardiomyopathy patients

Months post diagnosis	6	12	18	24	30	36	48
Patients (no.)	2	6	7	8	5	2	2
Cumulative	2	8	15	23	28	30	32
Total (%)	6.3	25	46.9	71.9	87.5	93.8	100

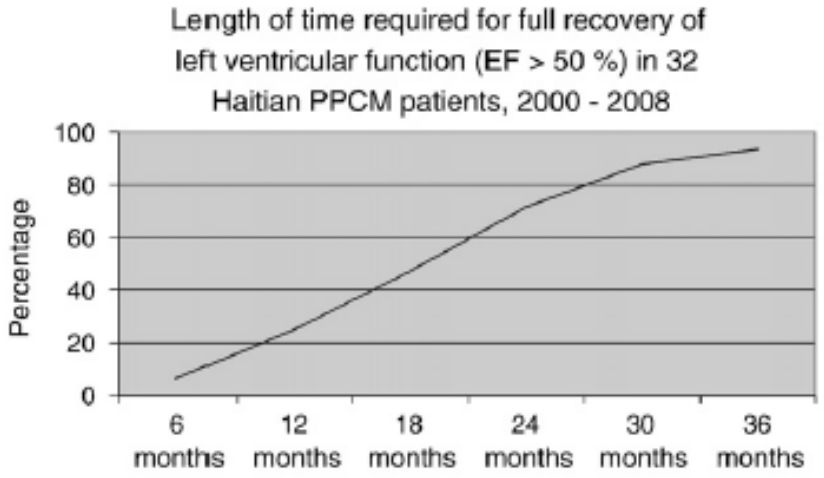
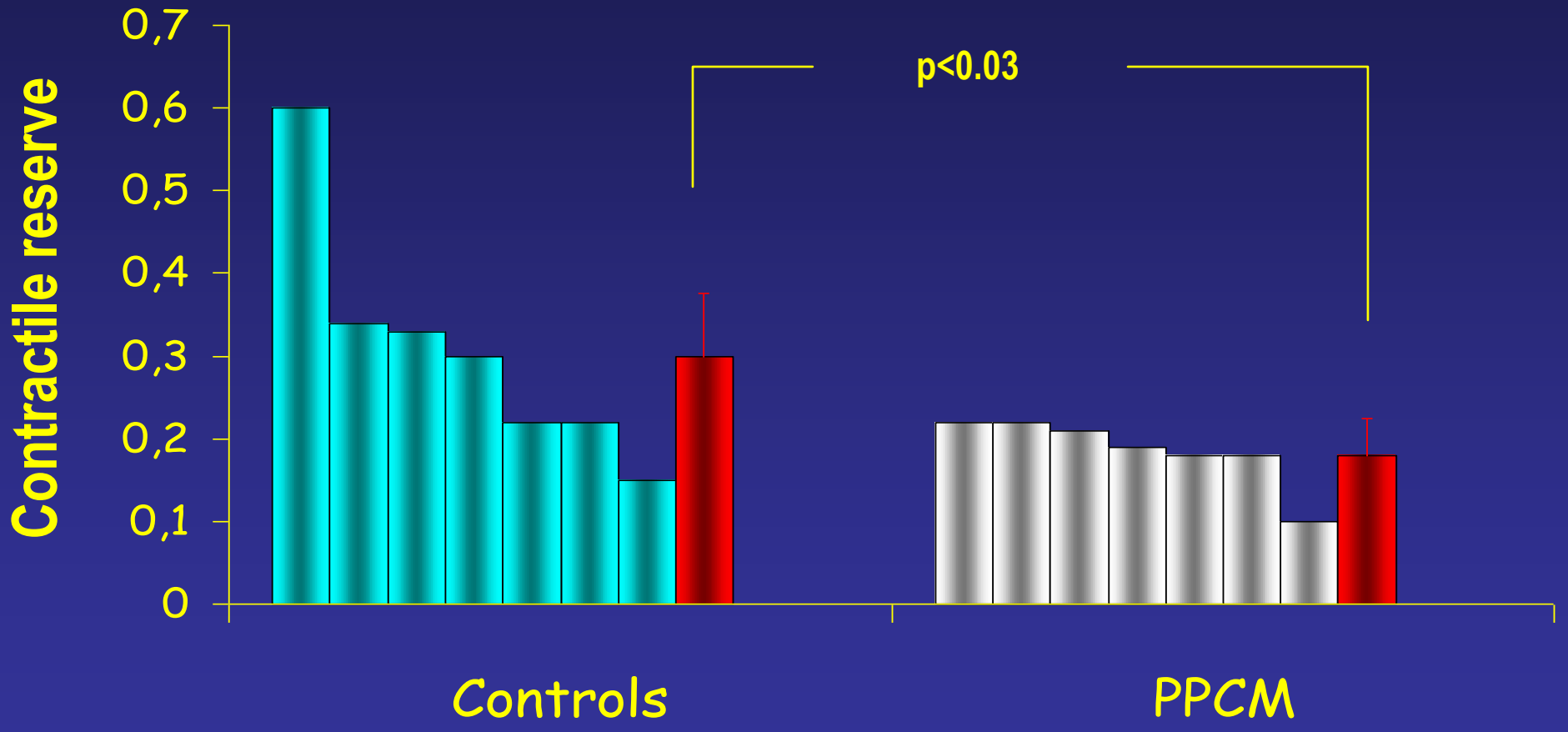


Fig. 1. Progression from shortest to longest time required for left ventricular systolic function recovery in peripartum cardiomyopathy.

International Journal of Gynecology and Obstetrics 104 (2009) 125–127

Contractile reserve of the LV among patients recovered from PPCM and matched controls



Lampert MB et al, Am J Obstet Gynecol,1997

CARDIOMIOPATIA PERIPARTUM : caso clinico

- *33 anni, 2 anni fa gravidanza con 2 episodi di gastroenterite e dispnea con ortopnea nell'ultimo mese*
- *Travaglio piuttosto lungo, con necessità di ossitocina*
- *Tentativo di parto vaginale con anestesia spinale (fentanil)…*
- *Cesareo con epidurale complicato da dispnea acuta e parestesie diffuse (anche arti superiori)…terapia non meglio precisata, prurito al volto…dispnea, shock, perdita di coscienza*
- *Bambina 3340 g, Apgar 1 min 9, Apgar 5 min 9*
- *Ecocardiogramma il giorno dopo il parto: DTDVsn 56 mm, DTSVsn 48 mm, AF 14%, FE 27%, IM lieve, IT lieve, versamento pericardico lieve*
- *Ecocardiogramma 5 giorni dopo il parto: DTDVsn 65 mm, DTSVsn 46 mm, AF 29%, FE 34%, IM minima, IT lieve, pattern restrittivo, versamento pericardico lieve*
- *Rapido miglioramento clinico e strumentale (terapia con ACE-I, beta-bloccante gradualmente "autoridotta")*

Diagnosi di PPCMP, successivamente messa in dubbio in altra sede x la rapidità di comparsa e risoluzione (shock da fentanil?)

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(tassativamente sconsigliata in altre autorevoli sedi)

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- *Sospende terapia*
- *Rivalutazione clinica e strumentale dopo 6 mesi*
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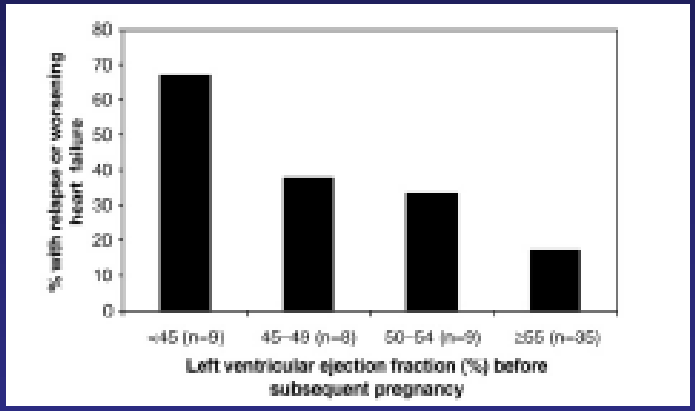
- *Ecostress con dobutamina (fino a 20 gamma/Kg/min): ottima risposta inotropa con incremento di FE fino a 75% e lieve riduzione dei volumi*

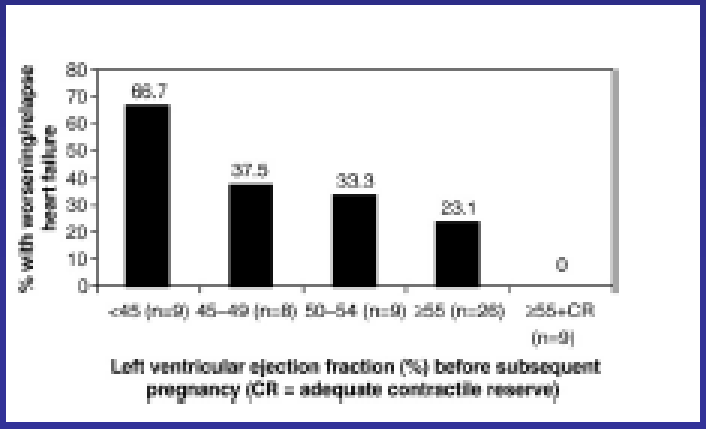
Risk of heart failure relapse in subsequent pregnancy among peripartum cardiomyopathy mothers

James D. Fett [a,b,*], Karie L. Fristoe [b], Serena N. Welsh [b]

[a] Peripartum Cardiomyopathy Research Project, Department of Adult Medicine, Hôpital Albert Schweitzer, Deschapelles, Haiti
[b] A Mother's Heart, Peripartum Cardiomyopathy Support Network



3 criteria were identified to be associated with a lower risk of heart failure relapse in a subsequent pregnancy: (1) regain an LVEF of 0.55 or greater after the index PPCM pregnancy; (2) retain an LVEF of 0.55 or greater after cardiologist-supervised discontinuation of heart failure medications; and (3) demonstrate adequate contractile reserve by exercise stress echocardiography. No relapses were observed when all 3 criteria were met (Table 1 and Fig. 2).



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PERIPARTUM CARDIOMIOPATHY: OPEN PROBLEMS

- Real prevalence and incidence
- Definition of risk factors and prognosis (subsequent pregnancies)
- The role of hypertension/preeclampsia (prolactin)
- The role of genes
- Central serum and tissue bank
- Evaluation of the effects of treatment

International Registry

National Heart, Lung, Blood Institute and Office of Rare Diseases Workshop on PPCMP, JAMA 2000

CMPPP-registro: pazienti arruolate (3 centri dell'Emilia Romagna): esordio precoce o classicamente correlato al parto degli aspetti clinici

	Età (anni)	FE% esordio	FE% follow-up	NYHA esordio	NYHA follow-up	PE associata	Tp follow-up
Esordio classico periparto (15 pz)	33.9 (30-39)	33.05 (15-40)	58 (48-67)	III-IV	I-II	20% (3/15)	87%(13/15)
Esordio precoce (5 pz)	32.5 (27-36)	42.67 (30-50)	50.67 (36-58)	III-IV	I	25% (1/4)	100%

EUROPEAN SOCIETY OF CARDIOLOGY®

EURObservational Research Programme

Long-Term Registry on Patients with Peripartum Cardiomyopathy (PPCM)

Protocol

June 1st, 2012

Registry promoted by the European Society of Cardiology