

# HCM & Pregnancy

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Davidai Arrhythmia Center  
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# L.P

- A 34 y.o patient w/ HOCCM
- Diagnosed at age 26
- Asymptomatic until the second pregnancy → mild DOE
- FHx: Familial CMP with hypertrophic and restrictive patterns:
  - Mother & Father: HOCCM
  - Brother: HOCCM
  - Brother: ICD
  - No h/o SCD

# L.P

- G3P2, Currently @ 12 week of gestation
- 1<sup>st</sup> Pregnancy – normal; Labor – vacuum
- 2<sup>nd</sup> Pregnancy – CHF -> Diuretics, BB; Labor-C/S

# L.P

## • ECG:

\*\*\* NO ADT RECORD FOR PATIENT \*\*\*

06-MAR-1983 (34 yr)

Female

Room:

Vent. rate	63	BPM
PR interval	216	ms
QRS duration	114	ms
QT/QTc	436/446	ms
P-R-T axes	50 20	111

Sinus rhythm with 1st degree A-V block with occasional Premature ventricular complexes

Possible Left atrial enlargement

ST & T wave abnormality, consider lateral ischemia

Abnormal ECG

When compared with ECG of 19-NOV-2017 10:10,

Premature ventricular complexes are now Present

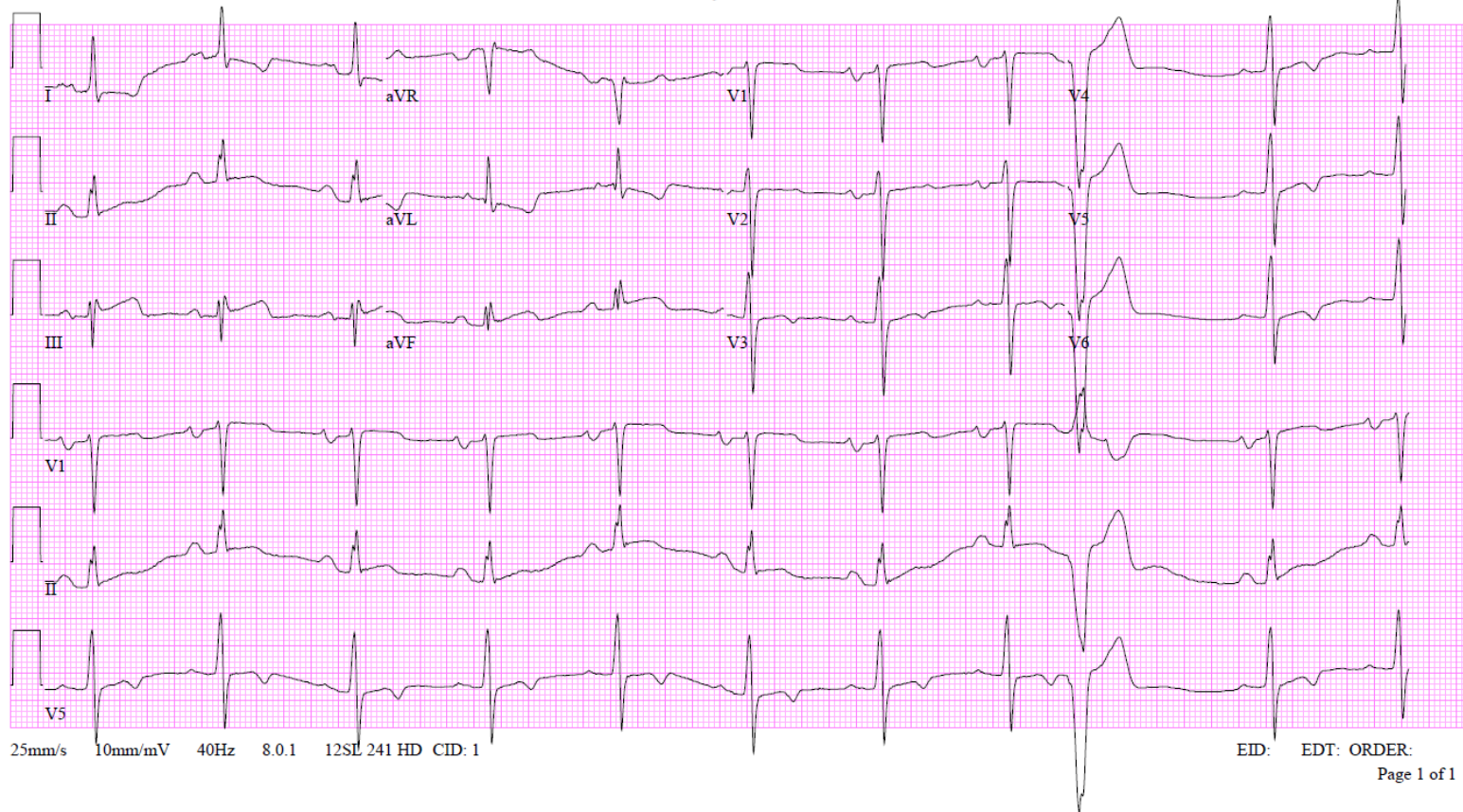
T wave inversion more evident in Lateral leads

Technician:

Test ind:

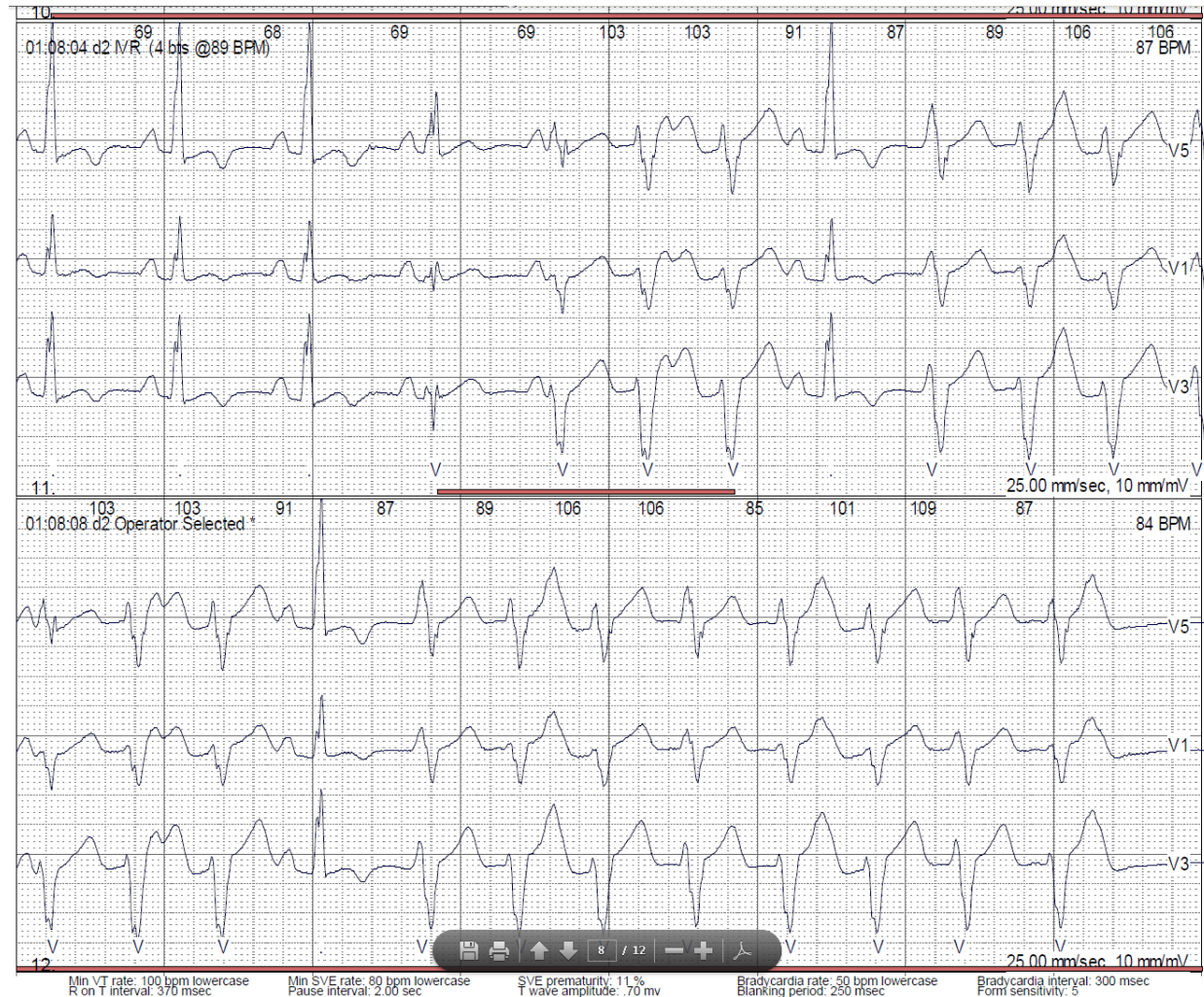
Referred by:

Unconfirmed

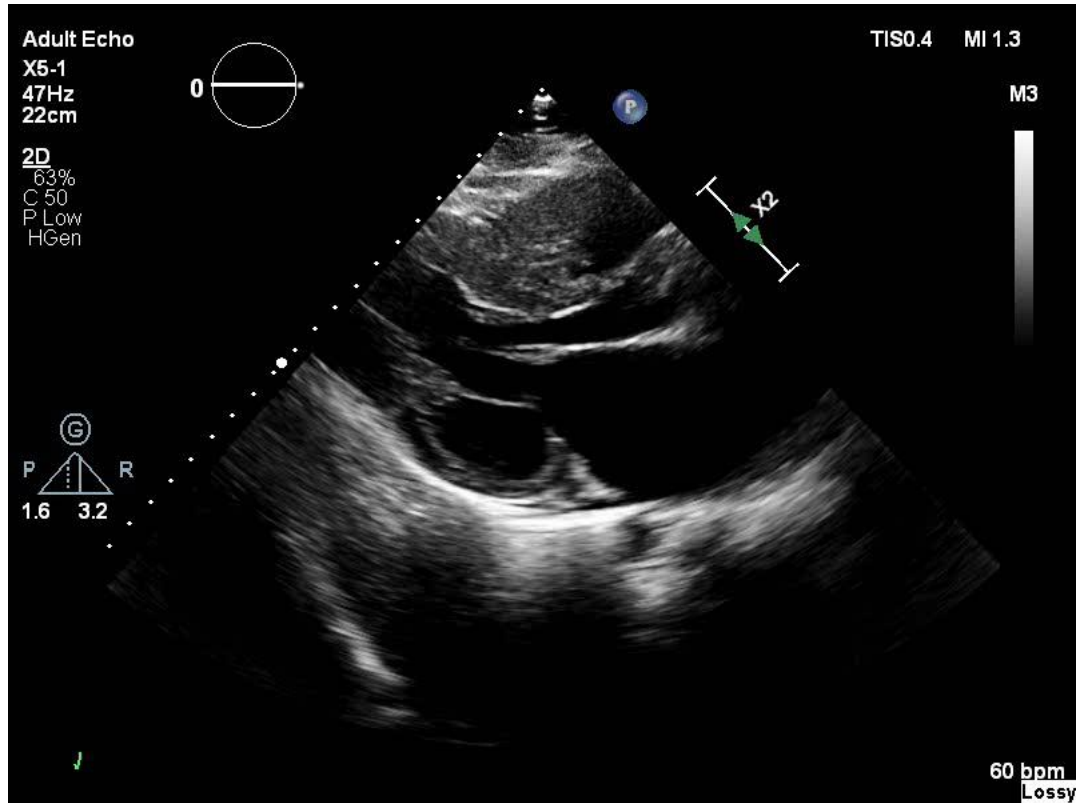


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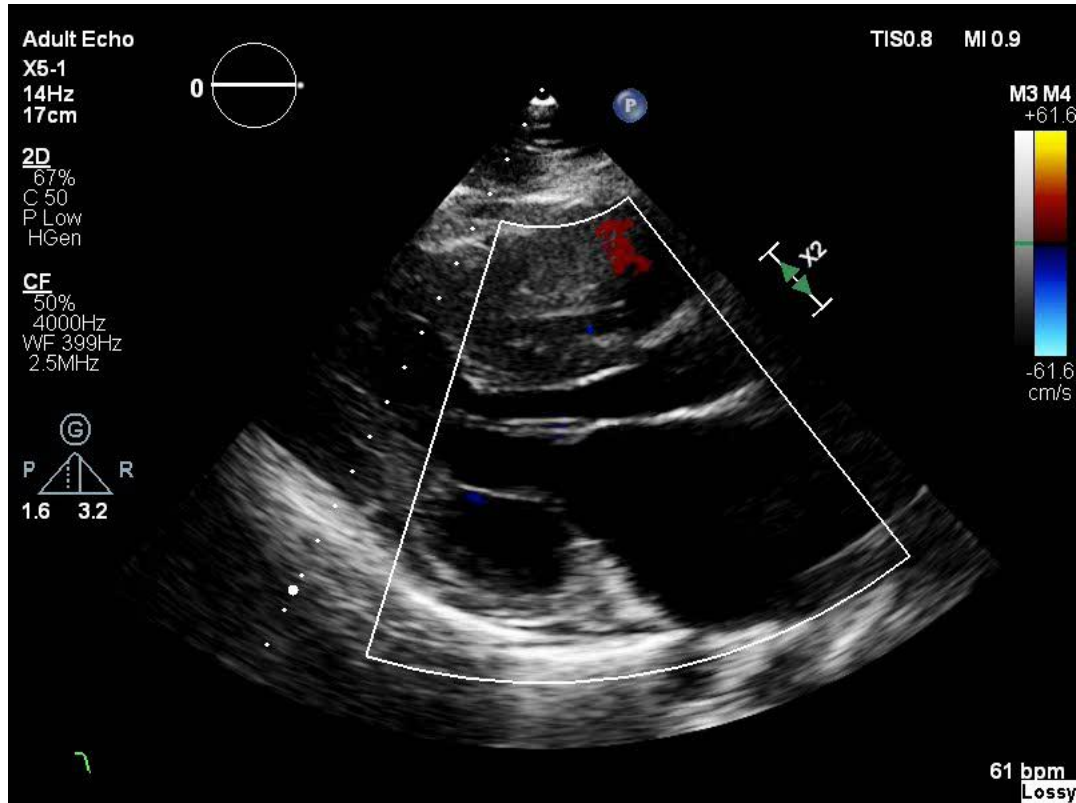
- Holter:



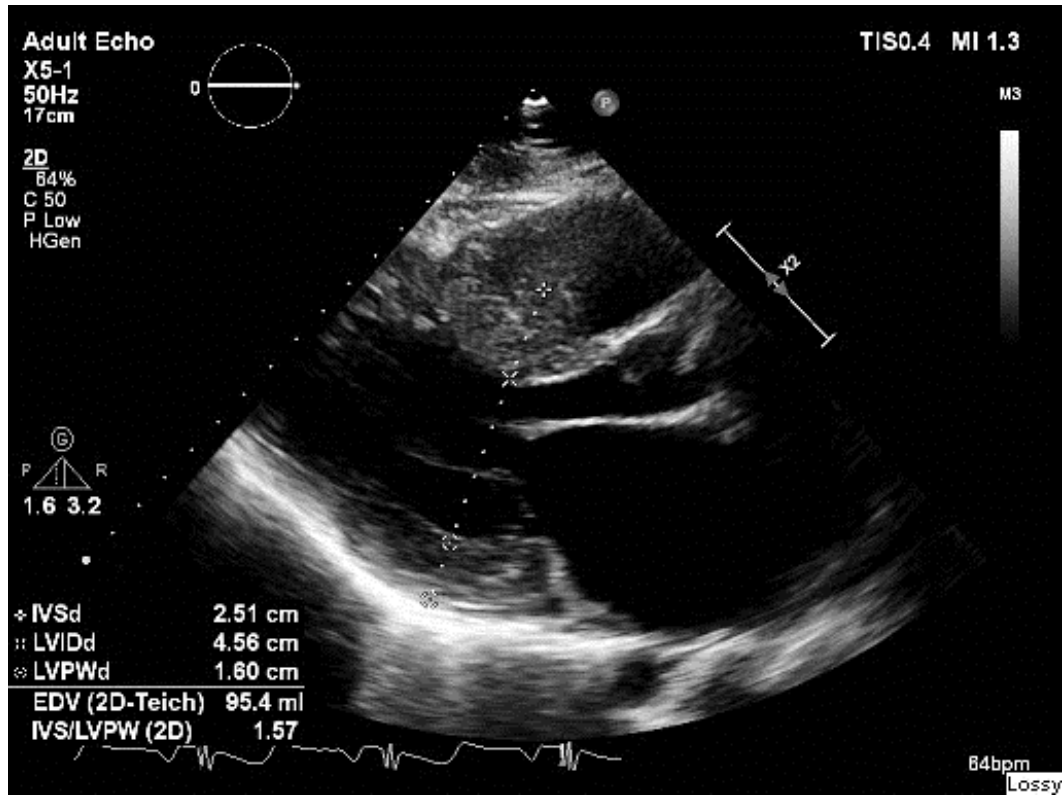
# L.P



# L.P

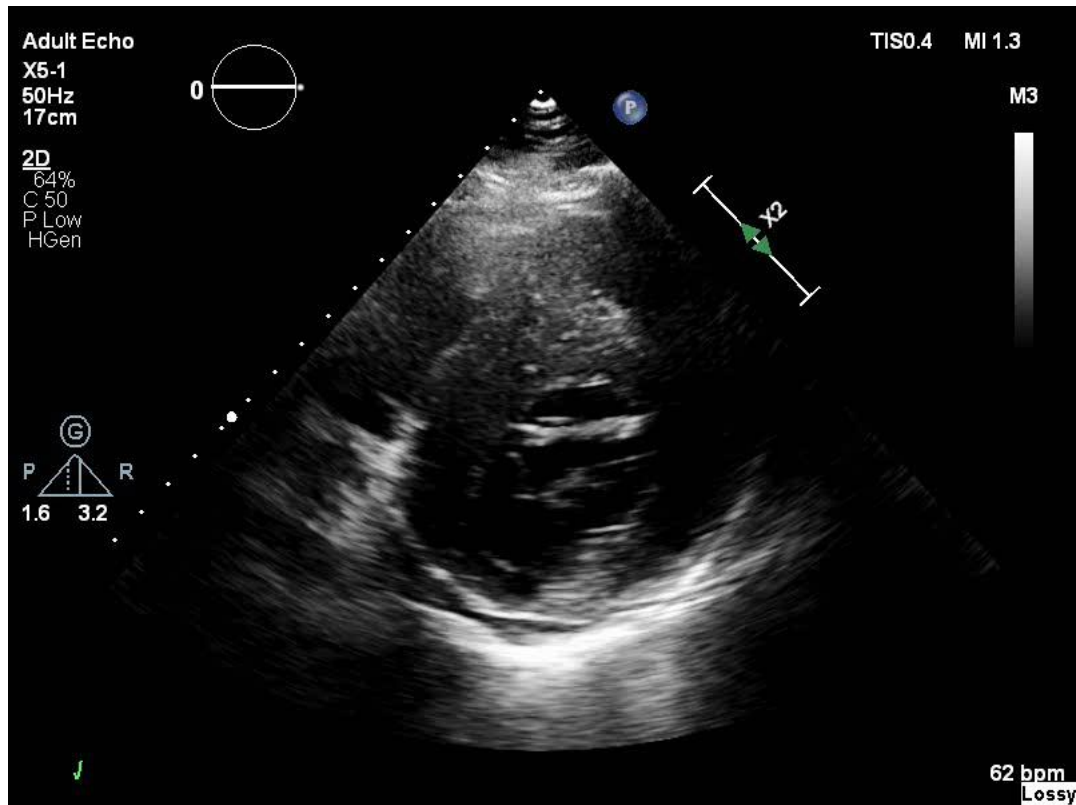


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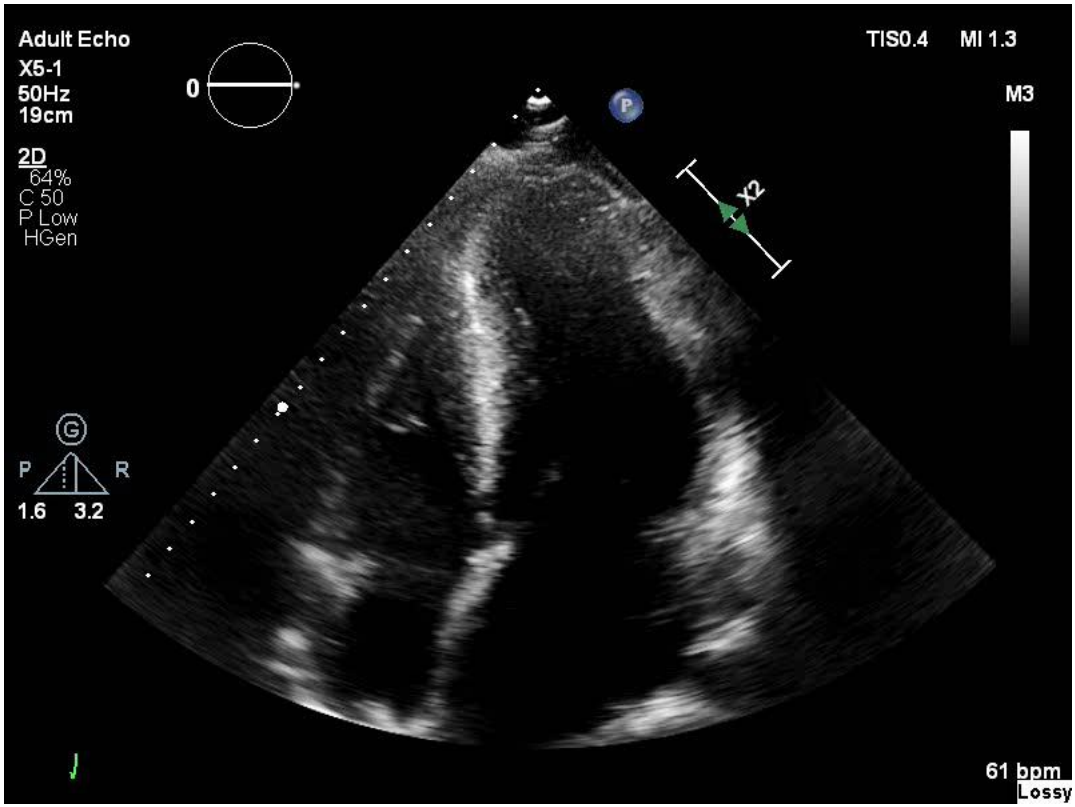




# L.P



# L.P



# L.P

- TTE:



NONINVASIVE CARDIOLOGY UNIT  
HEART INSTITUTE  
Leviev Heart Center  
SHEBA MEDICAL CENTER  
TEL HASHOMER, ISRAEL

טל 03-5302433  
פקס 03-5302407  
\*\* חסוי רפואי \*\*

היחידה לקרדיולוגיה לא פולשנית  
מכון הלב  
מרכז הלב ע"ש לבייב  
המרכז הרפואי ע"ש שיבא  
תל השומר, ישראל



Peak E-wave:	108 cm/s	Peak A-wave:	33.5 cm/s
E/A ratio:	3.2	Deceleration time:	198 msec
Comments:	Restrictive filling pattern		

### Tissue Doppler

S' velocity (septal):	4.9 cm/s	S' velocity (lateral):	7.29 cm/s
E' velocity (septal):	6.31 cm/s	E' velocity (lateral):	10 cm/s
E/e' ratio (septal):	17.12	E/e' ratio (lateral):	10.8

### Intraventricular pressure gradient

Location of obstruction:	LV outflow	Valsalva:	64 mmHg
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### Summary

### סיכום

עליה שמאלית מורחבת. חדר ימין תקין. חדר שמאל ברוחב תקין עם תפקוד גלובלי ואזורי היפרדינמי, היפרטרופיה ספטלית בולטת. הודגמה תנועה סיסטולית קידמית של המסתם המיטרלי עם מפל לחצים של 64 מ"מ"כ לאחר וולסלווה.. אס"ק מיטרלית קלה עד בינונית. אס"ק טריקוספידלית קלה. לחץ ריאתי תקין. תבנית מילוי רסטריקטיבית.

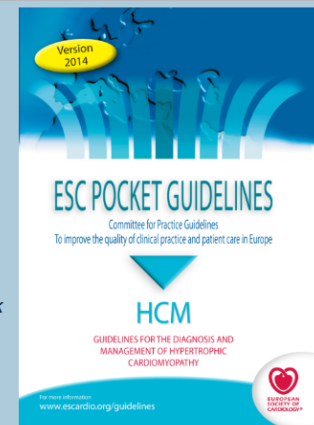


## HCM Risk-SCD Calculator

**Age**  **Years** *Age at evaluation*  
**Maximum LV wall thickness**  **mm** *Transthoracic Echocardiographic measurement*  
**Left atrial size**  **mm** *Left atrial diameter determined by M-Mode or 2D echocardiography in the parasternal long axis plane at time of evaluation*  
**Max LVOT gradient**  **mmHg** *The maximum LV outflow gradient determined at rest and with Valsalva provocation (irrespective of concurrent medical treatment) using pulsed and continuous wave Doppler from the apical three and five chamber views. Peak outflow tract gradients should be determined using the modified Bernoulli equation: Gradient= 4V<sup>2</sup>, where V is the peak aortic outflow velocity*  
**Family History of SCD**  No  Yes *History of sudden cardiac death in 1 or more first degree relatives under 40 years of age or SCD in a first degree relative with confirmed HCM at any age (post or ante-mortem diagnosis).*  
**Non-sustained VT**  No  Yes *3 consecutive ventricular beats at a rate of 120 beats per minute and <30s in duration on Holter monitoring (minimum duration 24 hours) at or prior to evaluation.*  
**Unexplained syncope**  No  Yes *History of unexplained syncope at or prior to evaluation.*

<b>Risk of SCD at 5 years (%):</b>	<input type="text" value="10.93"/>
<b>ESC recommendation:</b>	<input type="text" value="ICD should be considered"/>

Reset



<p>ICD implantation should be considered in patients with an estimated 5-year risk of sudden death <math>\geq 6\%</math> and a life expectancy <math>&gt; 1</math> year following detailed clinical assessment that takes into account the lifelong risk of complications and the impact of an ICD on lifestyle, socioeconomic status and psychological health.</p>	<b>IIa</b>	<b>B</b>	116,368
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# L.P

## סיכום יעוץ:

"בת 34 עם HOCM עיבוי ספטום משמעותי NSVT בהולטר, ירידת ל"ד במאמץ, עליה מוגדלת ומפל עד 64 מ"מ"כ במוצא החדר, כעת בשבוע 12 להריון.

**ע"פ הקוים המנחים ישנה אינידקציה ל ICD למניעה ראשונית עם חישוב של 10.93% ל SCD ב-5 שנים.**

לאור ההריון ולאחר דיון עם חברי הצוות, ממליץ על מתן **טו** עד סיום ההריון ולאחר מכם להשתיל ICD למניעה ראשונית (וזאת על מנת להימנע מקרינה).

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תשובת קופ"ח (טלפונית):

"בשיחה טלפונית עם XXXXX הובהר שהקופה לא תאשר וסט למטופלת.

המטופלת תועלה לוועדת חריגים **לאישור וסט רק לחודש האחרון להריון.**

מבחינת קופ"ח אין מניעה להשתיל ICD למטופלת בהריון החל **מטרימסטר שני**, ובלבד שהקרינה תהיה מוגבלת ל 5 rad “

# L.P

## תשובת קופ"ח (דוא"ל):

"חברים נכבדים ,

בהמשך למכתבכם בנושא של XXXXX מבקש להעמידכם על אי  
דיוקים בנוסח מכתבכם :

1. קופ"ח אישרה למרות אי הסכמה המקצועית שלי בוועדת חריגים מתן  
ווסט לגברת !!!!

2. הוויכוח היה על בסיס רפואי לחלוטין כאשר בבדיקתי את הנושא מול  
מנהלי תחום נשים ודימות בקופ"ח , הובהר כי **הסיכון להשתלה דפיניטיבית  
של הדפיברילטור לאחר טרימסטר ראשון של הריון מזערי**, במידה וקיימת  
התוויה מלאה להשתלת דפיברילטור כמו במקרה זה . יתרה מזה **אין ספק כי  
ההגנה בפני מוות פתאומי עדיפה לאין ערוך ע"י הדפיברילטור המושתל  
מאשר הווסט.**

3. שמחתי לשמוע כי לאחר שביקשתי ממכם לבדוק נושא זה אצלכם כך  
עשיתם והגעתם למסקנה זחה וכל זאת ללא כל שיקול כלכלי של קופ"ח אלא  
בריאות החולה ויעילות הטיפול בלבד .

4. מאושרת השתלת ICD."



# L.P

ניהול סיכונים:

**"1. קרינה לא מכוונת ישירות לרחם ולעובר אינה מהווה בעיה בהריון. כמובן בהנחה שהחשיפה לקרינה נדרשת למניעת חבלה חמורה לאם.**

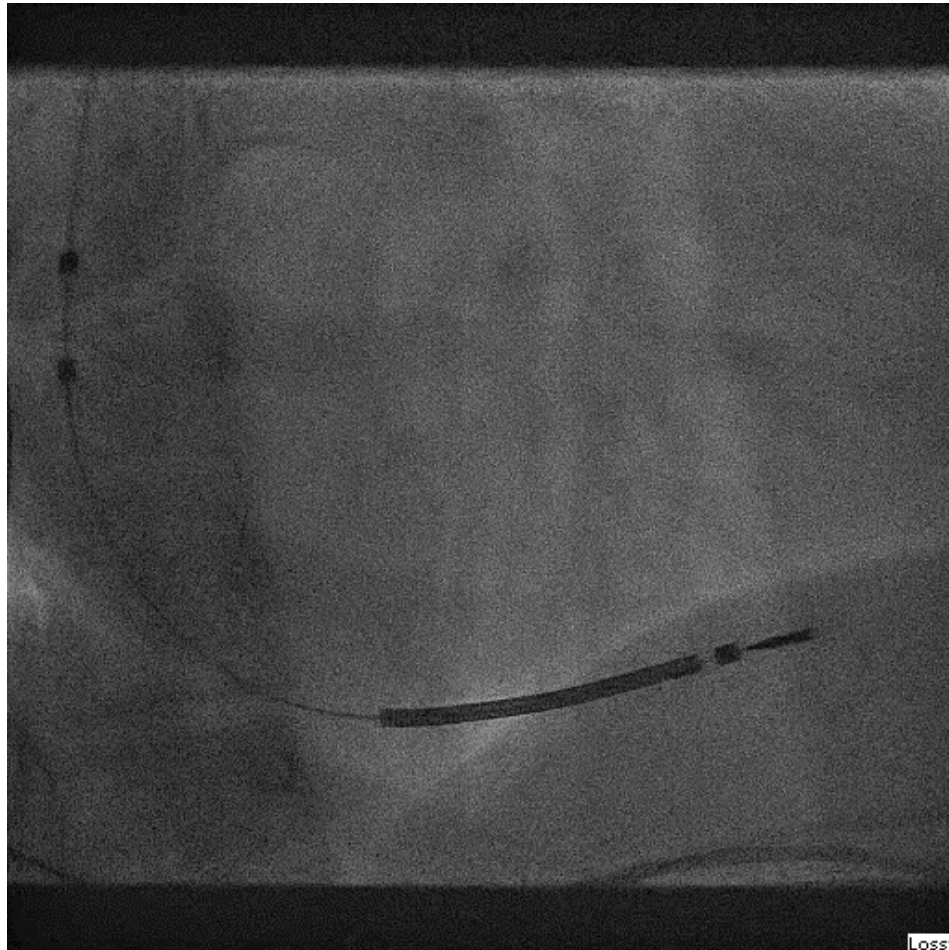
**2. קרינה בשליש השני להריון אינה שונה משמעותית מקרינה המופעלת בהדמיית פגים. למרות שאין דין שבוע 14 כדין שבוע 24.**

**3. כיסוי בסינר עופרת של אזור הבטן עונה בהחלט. השילוב של השניים - קרינה לא מכוונת עם כיסוי הרחם ממזער הסיכון מהקרינה למינימום לא מדאיג.**

**4. כמובן שטופס ההסכמה צריך לכלול ההסבר הזה של תועלת מירבית לעומת סיכון זניח לעובר ולהריון".**

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גישה צפאלית:



# L.P

- אין ספק כי ההגנה בפני מוות פתאומי עדיפה לאין ערוך ע"י הדפיברילטור המושתל מאשר הווסט.
- הסיכון להשתלה דפיניטיבית של הדפיברילטור לאחר טרימסטר ראשון של הריון מזערי

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## Chapter 5: Individualization of Treatment

FDA:

### Specific patient population

Patient groups expected to use the LifeVest device are those whose SCD risk is temporary (hours to months) or those who have limited expected life span (less than one year). Patients who have an elevated SCD risk include the following:

- Patients awaiting cardiac transplant or patients having equivalent heart status (New York Heart Association Class III or IV heart failure) and an ejection fraction below 30%.
- Patients having an acute myocardial infarction (MI), or patients immediately post coronary artery bypass graft surgery with any of the following: a VT/VF event within 48 hours of the MI or surgery, an ejection fraction below 30% at least three days after MI or surgery, or patients having a sudden cardiac arrest or syncopal VT event at least 48 hours after MI or surgery and not receiving an ICD. Also included are patients having an acute MI and are Killip Class III or IV at least three days after the MI.
- Patients having viral, chemical, or metabolic cardiomyopathy who are expected to recover.
- Patients beginning pro-arrhythmic medications.

Patient groups not expected to use the LifeVest device are:

- Patients with mental, visual, physical, or auditory deficits that could impair their ability to properly interact with the LifeVest device.
- Patients taking medication that would significantly impair their ability to activate the response buttons.
- Patients who are unwilling or unable to comply with usage requirements such as wearing the device continuously, except when bathing or showering.
- Patients, who for anatomic or other non-correctable reasons, have excessive amounts of electrode noise corrupting the detection algorithm.
- Female patients who are pregnant, breast-feeding, or who are not taking adequate contraceptive measure if they are of childbearing age.
- Patients under 18 years of age.
- Any patient with an advance directive prohibiting resuscitation.

## Clinical Investigations

# Characteristics and Outcomes of Peripartum Versus Nonperipartum Cardiomyopathy in Women Using a Wearable Cardiac Defibrillator

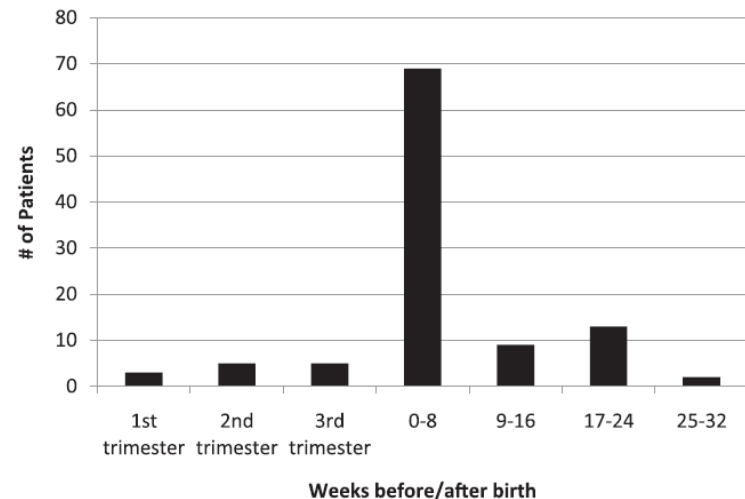
MITCHELL T. SALTZBERG, MD,<sup>1</sup> STEVEN SZYMKIEWICZ, MD,<sup>2</sup> AND NICOLE R. BIANCO, PhD<sup>2</sup>

*Newark, Delaware; Pittsburgh, Pennsylvania*

13 (12%) of the PPCM patients were prescribed the WCD prepartum.

None of the PPCM patients referred for WCD received an appropriate shock during the time the WCD was used.

No inappropriate WCD shocks were observed.



**Fig. 2.** Timing of wearable cardioverter defibrillator (WCD) prescription. The known timing of WCD prescription is in regard to stage of pregnancy in the 107 peripartum cardiomyopathy (PPCM) women.

## Risk for ventricular fibrillation in peripartum cardiomyopathy with severely reduced left ventricular function—value of the wearable cardioverter/defibrillator

David Duncker<sup>1\*</sup>, Arash Haghikia<sup>1</sup>, Thorben König<sup>1</sup>, Stephan Hohmann<sup>1</sup>, Klaus-Jürgen Gutleben<sup>2</sup>, Ralf Westenfeld<sup>3</sup>, Hanno Oswald<sup>1</sup>, Helmut Klein<sup>4</sup>, Johann Bauersachs<sup>1</sup>, Denise Hilfiker-Kleiner<sup>1</sup>, and Christian Veltmann<sup>1</sup>

**Table 2** Characteristics and follow-up of the seven patients treated with a wearable cardioverter/defibrillator (WCD)

	Patient #6	Patient #7	Patient #8	Patient #9	Patient #10	Patient #11	Patient #12
Age, years	37	36	30	41	32	27	35
Time of diagnosis after delivery	2 weeks	7 days	4 weeks	4 weeks	5 months	4 weeks	3 months
Number of pregnancies	2	2	3	3	2	2	4
LVEF at diagnosis, %	20	10	15	15	10	30	10
NYHA at diagnosis	IV	III	IV	IV	IV	II	IV
Longest follow-up, months	13	15	6	5	15	5	12
LVEF at longest follow-up, %	45	35	53	41	48	47	34
ΔLVEF	25	25	38	26	38	17	24
NYHA at longest follow-up	II	II	I	II	I	II	II
WCD days	176	183	81	42	25	80	345
WCD wear time, h/day	23.2	23.1	16.3	23.0	21.8	23.1	23.6
Number of WCD shocks	0	0	0	0	1	1	2
ICD	Not indicated	Primary prophylactic CRT-D implanted	Not indicated	Not indicated	Secondary prophylactic CRT-D implanted	Secondary prophylactic ICD implanted	Secondary prophylactic ICD refused

N=12



## Correspondence

## Interdisciplinary management of left ventricular hypertrabeculation/ noncompaction during pregnancy with a wearable defibrillator



E. Reuschel <sup>a,\*,1</sup>, A. Baessler <sup>b,\*,1</sup>, C. Stöllberger <sup>c</sup>, J. Finsterer <sup>d</sup>, L. Maier <sup>b</sup>, M. Fischer <sup>b</sup>, F. Poschenrieder <sup>d</sup>,  
F. Heissenhuber <sup>e</sup>, K. Kurzidim <sup>e</sup>, C.P. Schepp <sup>f</sup>, G. Badelt <sup>f</sup>, B. Seelbach-Göbel <sup>a</sup>

<sup>a</sup> Klinik für Gynaekologie und Geburtshilfe, Krankenhaus Barmherzige Brüder Regensburg, Germany

<sup>b</sup> Universitäres Herzzentrum Regensburg, Regensburg, Germany

<sup>c</sup> Krankenanstalt Rudolfstiftung, Wien, Austria

<sup>d</sup> Institut für Röntgendiagnostik, Universitätsklinikum Regensburg, Germany

<sup>e</sup> Klinik für Herzrhythmusstörungen, Krankenhaus Barmherzige Brüder Regensburg, Germany

<sup>f</sup> Klinik für Anästhesiologie, Krankenhaus Barmherzige Brüder Regensburg, Germany

**A case report: A 27 y.o woman with LV-non compaction** presented within the **16th gestational week**.

**TTE** showed a mildly reduced LVEF and **pronounced trabeculation of LV**

The cardiac **MRI** performed subsequently confirmed **hypertrabeculation of LV**

FHx: sister with HCM died due to LV failure

48-hour **Holter** monitoring disclosed 2 **NSVT**

Implantation of an **ICD was recommended but refused by the patient**.

At 24 weeks of gestation, 24-hour monitoring showed multiple ventricular ectopic beats and short ventricular ectopic runs.

A **WCD was suggested**. Cardiac monitoring by the WCD did not disclose any significant arrhythmias during the further pregnancy.

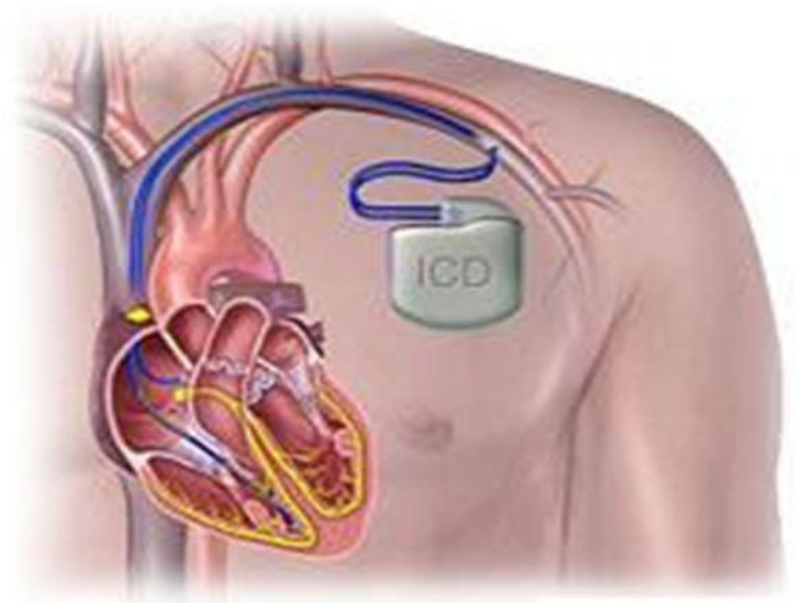
**C/S @ 37+1**



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European Society  
of Cardiology

Europace (2017) **19**, 1909–1922

doi:10.1093/europace/eux252

**EHRA CONSENSUS DOCUMENT**

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# **Occupational radiation exposure in the electrophysiology laboratory with a focus on personnel with reproductive potential and during pregnancy: A European Heart Rhythm Association (EHRA) consensus document endorsed by the Heart Rhythm Society (HRS)**

**Andrea Sarkozy<sup>1\*</sup>, Tom De Potter<sup>2</sup>, Hein Heidbuchel<sup>1</sup>, Sabine Ernst<sup>3</sup>,  
Jedrzej Kosiuk<sup>4</sup>, Eliseo Vano<sup>5</sup>, Eugenio Picano<sup>6</sup>, Elena Arbelo<sup>7</sup>, and Usha Tedrow<sup>8</sup>**

**Table 3** Typical patient' and physician's radiation doses in electrophysiology

Type of study	Effective dose to patient in mSv median and range per procedure <sup>9</sup>	Effective dose to operator in $\mu$ Sv mean and mean range per procedure <sup>a 23</sup>
Diagnostic electrophysiological study	3.2 1.3–23.9	UR <sup>b</sup>
Ablation procedure	15.2 1.6–59.6	2.7 0.24–9.6
Atrial fibrillation	16.6 6.6–59.6	3.3 UR <sup>b</sup>
AT/AVNRT/AVRT	4.4 1.6–25	2.6 0.2–9
Ventricular tachycardia	12.5 3– $\geq$ 45	UR <sup>b</sup>
VVI/DDD PM or ICD implant	4 = 4 mGy 1.4–17	4.8 0.29–17.4
CRT implant	22 2.2–95	UR <sup>b</sup>
Coronary angiography	7 2.0–16	4.4 0.02–38
Percutaneous coronary intervention	15 7–57	4.9 0.17–31

<sup>a</sup>The reported mean doses and mean dose ranges are mean estimates from a small number of studies including low number of procedures performed before 2008 and should be interpreted with caution. Operator doses varied by two to three orders of magnitude for the same type of procedure.

<sup>b</sup>Under-reported: occupational exposure is reported in an insufficient number of procedures to produce representative numbers for operator effective doses.

AT/AVNRT/AVRT, atrial tachycardia, atrioventricular nodal re-entry tachycardia, atrioventricular re-entry tachycardia; CRT, cardiac resynchronization therapy; ICD, implantable cardioverter-defibrillator; PM, pacemaker; UR under-reported.

**Table 5** Probability of a live birth without malformation or without childhood cancer as a function of radiation dose<sup>7</sup>

Dose to conceptus above natural background radiation (mGy)	No malformations (%)	No childhood cancer (%)	No malformations and no childhood cancer (%)
0	96.00	99.93	95.93
0.5	95.999	99.926	95.928
1.0	95.998	99.921	95.992
5.0	95.99	99.89	95.88
10.0	95.98	99.84	95.83

***“Radiation risks are most significant during organogenesis and the early fetal period, somewhat **less in the second trimester, and least in the third trimester**”***

**EXPERT CONSENSUS DOCUMENT**

# 2018 ACC/HRS/NASCI/SCAI/SCCT Expert Consensus Document on Optimal Use of Ionizing Radiation in Cardiovascular Imaging: Best Practices for Safety and Effectiveness

A Report of the American College of Cardiology Task Force on  
Expert Consensus Decision Pathways

*Developed in Collaboration With Mended Hearts*

**TABLE 1****Continued**

<b>Modality</b>	<b>Protocol</b>	<b>Typical Effective Does (mSv)</b>
Fluoroscopy	TAVR, transapical approach	12-23
Fluoroscopy	TAVR, transfemoral approach	33-100
Fluoroscopy	Diagnostic electrophysiological study	0.1-3.2
Fluoroscopy	Radiofrequency ablation of arrhythmia	1-25
Fluoroscopy	Permanent pacemaker implantation	0.2-8 = <b>0.0002-0.008 Gy</b>
Fluoroscopy	Diagnostic invasive coronary angiography	2-20
Fluoroscopy	Percutaneous coronary intervention	5-57

**TABLE 10** Estimates of Adverse Embryonic and Fetal Events as a Function of Fetal Radiation Dose

Acute Radiation Dose*to the Embryo/Fetus	Time Post Conception				
	Blastogenesis (up to 2 wks)	Organogenesis (2-7 wks)	Fetogenesis		
			(8-15 wks)	(16-25 wks)	(26-38 wks)
< 0.05 Gy (5 rads)†	Noncancer health effects NOT detectable				
0.05-0.50 Gy (5-50 rads)	Incidence of failure to implant may increase slightly, but surviving embryos will probably have no significant (noncancer) health effects	<ul style="list-style-type: none"> <li>■ Incidence of major malformations may increase slightly</li> <li>■ Growth retardation possible</li> </ul>	<ul style="list-style-type: none"> <li>■ Growth retardation possible</li> <li>■ Reduction in IQ possible (up to 15 points, depending on dose)</li> <li>■ Incidence of severe mental retardation up to 20% depending on dose</li> </ul>	Noncancer health effects unlikely	
> 0.50 Gy (50 rads) <i>The expectant mother may be experiencing acute radiation syndrome in this range, depending on her whole-body dose.</i>	Incidence of failure to implant will likely be large.‡ depending on dose, but surviving embryos will probably have no significant (noncancer) health effects	<ul style="list-style-type: none"> <li>■ Incidence of miscarriage may increase, depending on dose</li> <li>■ Substantial risk of major malformations such as neurological and motor deficiencies</li> <li>■ Growth retardation likely</li> </ul>	<ul style="list-style-type: none"> <li>■ Incidence of miscarriage probably will increase, depending on dose</li> <li>■ Growth retardation likely</li> <li>■ Reduction in IQ possible (&gt;15 points, depending on dose)</li> <li>■ Incidence of severe mental retardation &gt;20%, depending on dose</li> <li>■ Incidence of major malformations will probably increase</li> </ul>	<ul style="list-style-type: none"> <li>■ Incidence of miscarriage may increase, depending on dose</li> <li>■ Growth retardation possible, depending on dose</li> <li>■ Reduction in IQ possible, depending on dose</li> <li>■ Severe mental retardation possible, depending on dose</li> <li>■ Incidence of major malformations may increase</li> </ul>	<ul style="list-style-type: none"> <li>■ Incidence of miscarriage and neonatal death will probably increase depending on dose§</li> </ul>

# Implantable cardioverter-defibrillator in pregnant women with hypertrophic cardiomyopathy

## *Implante de cardio-desfibrilador em gestantes com cardiomiopatia hipertrófica*

Table 1. Surgical Procedure and ICD Programming

Characteristics	Patient 1	Patient 2
Date of ICD Implantation	April 2007	August 2007
Device implanted	VVIRL	VVIRL
Venous access	Left cephalic vein	Left subclavian vein
Electrode model	Medtronic 6949	Medtronic 6949
Pulse generator manufacturer	Medtronic Entrust D154	Medtronic Entrust D154
ICD Programming		
• Minimum rate	40 bpm	40 bpm
• VT Monitor	130-170 bpm	146-170 bpm
• Anti-Tachycardia Pacing (ATP)	171-194 bpm	171-194 bpm
• Shock Therapy	>194 bpm	>194 bpm

*VVIRL= single chamber ICD; bpm= beats per minute*

**“The low risk of malformations by radiation after the first trimester of pregnancy, which allowed the use of fluoroscopy ... despite the current pregnancy”**



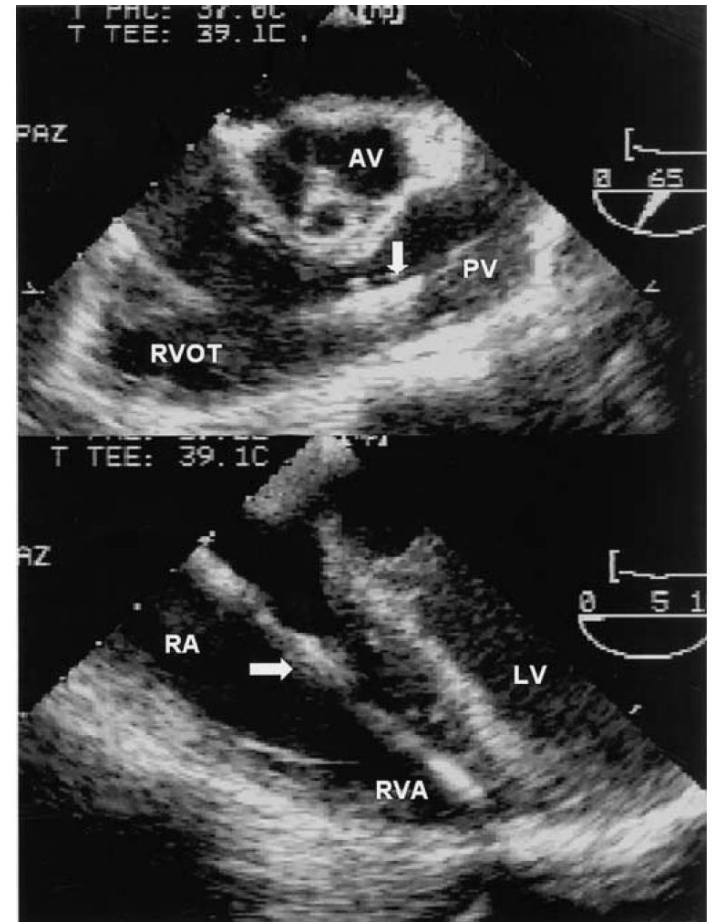
# Cardioverter Defibrillator Implantation in a Pregnant Woman Guided with Transesophageal Echocardiography

MAURICIO ABELLO, RAFAEL PEINADO, JOSÉ LUIS MERINO, MARIANA GNOATTO, MARTA MATEOS, JORGE SILVESTRE, and JOSÉ LUIS DOMINGUEZ

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ABELLO, M., ET AL.: Cardioverter Defibrillator Implantation in a Pregnant Woman Guided with Transesophageal Echocardiography. *This report describes a 28-year-old pregnant woman with mitral valve prolapse and sudden cardiac death due to a ventricular fibrillation who underwent an ICD implantation guided by transesophageal echocardiography. (PACE 2003; 26:1913–1914)*

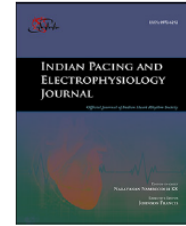
“ICD implantation **under TEE guidance** is feasible”. *(with 4 sec fluoroscopy)*





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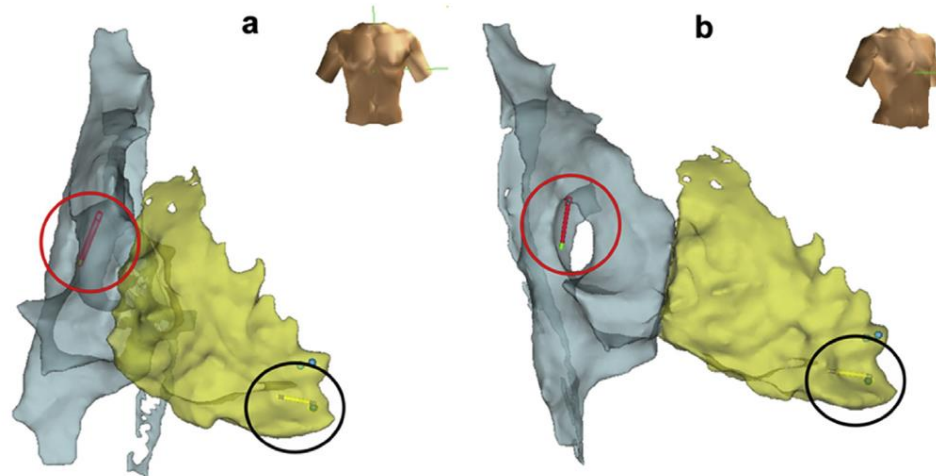
## Implantation of single lead cardioverter defibrillator with floating atrial sensing dipole in a pregnant patient without using fluoroscopy



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**“This technique may be taken into consideration for the few **rare cases** where **fluoroscopy cannot be performed**”**

**Fig. 1.** Three-dimensional reconstruction of the right heart structures in AP view (panel a) and in RAO view (panel B). Blue structure represents right atrial, inferior vena cava, and superior vena cava. Yellow structure represents right ventricle (RV). The implantable cardioverter defibrillator ventricular lead tip can be seen in the black circle, near the apex of the RV; the floating atrial dipole of the lead is visible in the red circle, in the right high region of the atrium.

# Conclusions

- Performing **ICD** implantation after the 1<sup>st</sup> trimester (>13-14 weeks) is **relatively safe**.
- **No data** on the efficacy of **WCD** during pregnancy.
- The **risk-benefit ratio** should be evaluated.
- Pros and Cons should be **discussed with the patient**.

Thank you