

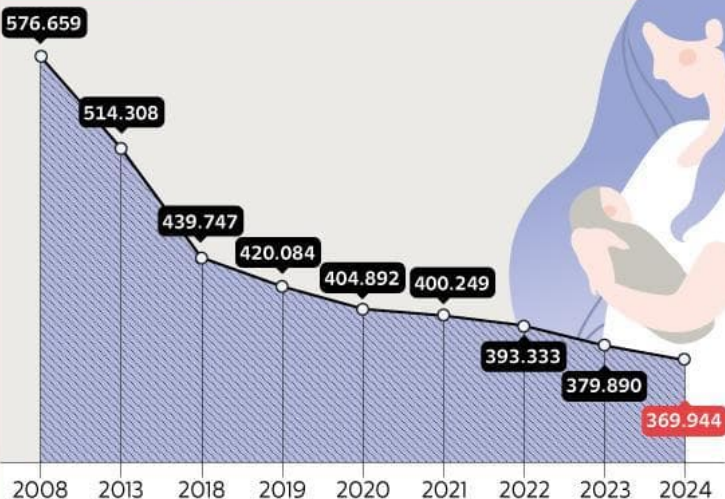
Gravidanza e Patologie Cardiovascolari

La gravidanza nelle pazienti con cardiomiopatia aritmogena

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IL CALO DELLE CULLE



1,11

Numero medio di figli delle donne di cittadinanza italiana nel **2024**

Era **1,14** nel **2023**

43,2%

Percentuale di nascite fuori dal matrimonio

Era **42,4%** nel **2023**

31,9

Età media delle madri alla nascita del primo figlio

31,7 anni nel **2023**

Fonte: ISTAT

The population frequency of
Arrhythmogenic
Cardiomyopathy has been
estimated at 1:1000 to 5000.

Corrado, Domenico et al. "Arrhythmogenic
Cardiomyopathy." *Circulation research* vol. 121,7
(2017): 784-802.



Table 1 Obstetric, fetal, and neonate outcomes

First author	Year	Women (n)	Pregnancies (n)	Live birth (n)	Miscarriage (n; %)	Cesarean section (n; %)	Preterm delivery (n; %)	Birth weight (mean g)
Bauce [5]	2006	6	6	6	0	4 (67)	0	3490 g
Boulé [9]	2014	2	2	2	0	1 (50)	1 (50)	NR
Hodes [31]	2016	26	39	39	6 (15)	11 (28)	2 (5)	3400 g
Billebeau [7]	2018	3	3	3	0	0	0	NR
Gandjbakhch [27]	2018	23	60	50	10 (17)	8 (13)	2 (3)	3229 g
Castrini [13] ^a	2019	58	88	86	2 (2)	6 (7)	NR	NR
Luo [36]	2020	9	9	9	0	4 (44)	0	3118 g
Wu [69]	2020	120	224	192	32 (14)	44 (20)	10 (5)	NR
Platonow [48]	2020	120	261	261	NR	NR	NR	NR
<i>n</i>		367	692	648	50/431	78/431	15/343	104/648
%		–	–	–	11.6	18.1	4.4	16
<i>g</i>		–	–	–	–	–	–	3292

mean g Mean grams, *NR* not reported

^aReference [48] includes 58 patients

Wichter et al, Herzschr Elektrophys 2021



Table 6 Modified World Health Organization 2.0 classification of maternal cardiovascular risk

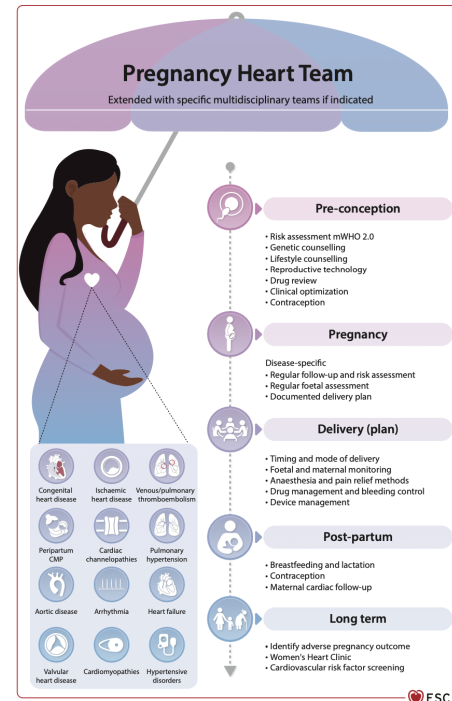
	mWHO 2.0 I	mWHO 2.0 II	mWHO 2.0 II-III	mWHO 2.0 III	mWHO 2.0 IV
Diagnosis	Ventricular (dys)function + pulmonary hypertension				
			Mild left ventricular impairment: EF >45%. Significantly impaired RV (subpulmonary) function.	Moderate left ventricular impairment: EF 30%–45%. Previous PPCM with not more than mild residual left ventricular impairment.	Severe left ventricular impairment: EF <30% or NYHA class III/IV. Previous PPCM with more than mild left ventricular impairment. PAH.
	Arrhythmias				
	Atrial or ventricular ectopic beats, isolated.	Most supraventricular arrhythmias. Bradycardia requiring pacemaker.	Low-risk LQTS: no previous events + on full dose beta-blocker therapy. Low-risk CPVT: well controlled by medical therapy. BrS with no previous events.	Sustained ventricular tachycardia from any aetiology. LQT2 (post-partum). Symptomatic CPVT and LQTS not adequately controlled by therapy. BrS with previous events.	
	Cardiomyopathy				
	HCM: genotype-positive + phenotype-negative.		Low-risk ARVC: genotype-positive + no or mild phenotype. HCM without complications. DCM/NDLVC with normal or mild left ventricular impairment: EF >45%.	ARVC with moderate/severe disease. HCM with arrhythmic and/or moderate haemodynamic complications. DCM/NDLVC with moderate left ventricular impairment: EF 30%–45%.	DCM/NDLVC with severe left ventricular impairment: EF <30% or NYHA class III/IV. HCM with symptomatic severe outflow tract obstruction: ≥50 mmHg. HCM with severely symptomatic LV dysfunction (EF <50%).

Specific expertise and collaborative management by a Pregnancy Heart Team is mandatory for all women with a condition of mWHO 2.0 class II–III or above

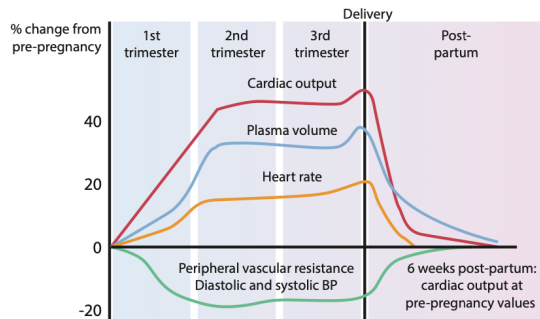


	mWHO 2.0 I	mWHO 2.0 II	mWHO 2.0 II-III	mWHO 2.0 III	mWHO 2.0 IV
Involvement of the Pregnancy Heart Team	No	No	Yes	Yes	Yes
Counselling	Yes (by regular healthcare professional)	Yes (by regular healthcare professional)	Yes: expert counselling by Pregnancy Heart Team is required	Yes: expert counselling by Pregnancy Heart Team is required	Yes: expert counselling by Pregnancy Heart Team is required, with clear and thorough discussion of very high pregnancy risk and shared decision-making process for termination if pregnancy occurs
Obstetric and cardiac care during pregnancy	Local hospital	Local hospital	Shared care with local hospital + Pregnancy Heart Team	Care led by Pregnancy Heart Team	Care led by Pregnancy Heart Team
Location of delivery	Local hospital	Local hospital	Shared care with local hospital + Pregnancy Heart Team. Location depends on CV status and evolution of pregnancy	Expert centre, care led by Pregnancy Heart Team	Expert centre, care led by Pregnancy Heart Team

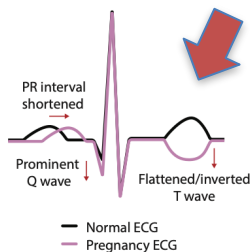
© ESC 2025



Haemodynamic changes



ECG changes



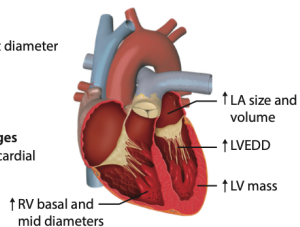
Echocardiographic changes

Unchanged

- Aortic root diameter
- LVEF
- RVEF
- SPAP

Small changes

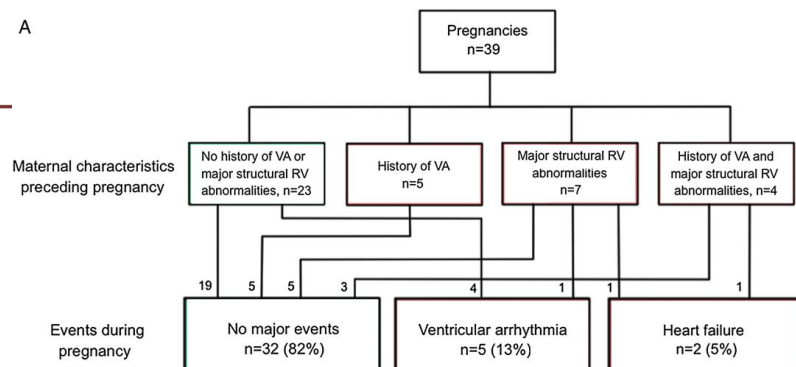
- Small pericardial effusion



- Arrhythmic risk?
- Heart Failure occurrence?



A



Most pregnancies were uneventful



Excellent obstetric outcomes



Optimize management before conception:

B



Cardiomyopathies (see specific indications)

++ Metoprolol, propranolol, nadolol, flecainide

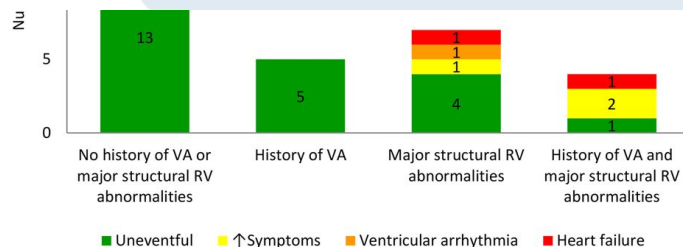
+- Sotalol

x ACE-I, ARB, ARNI, disopyramide, direct renin inhibitors, MRA, SGLT2-I, mavacamten, atenolol

++ Metoprolol, propranolol, nadolol, flecainide, spironolactone

+- Sotalol, candesartan

x ARB^a, disopyramide, direct renin inhibitors, SGLT2-I, mavacamten

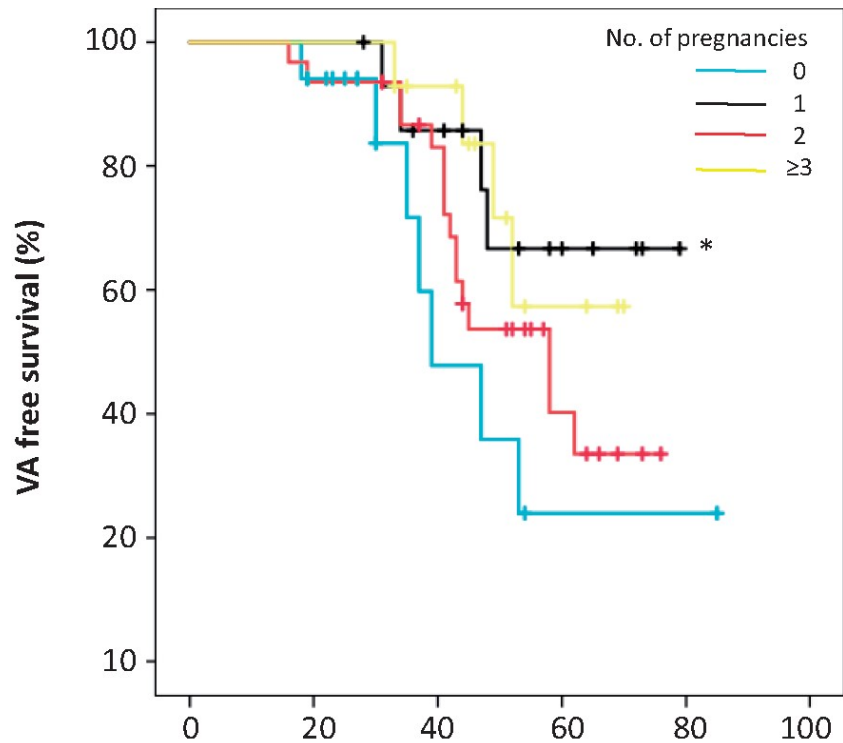


Assess ventricular function before pregnancy



Do not discontinue β -blockers





Number of pregnancies

Number at risk

Age (years)

0	19	15	4	1	0
1	16	15	10	4	0
2	30	29	24	6	0
≥3	12	11	10	3	0

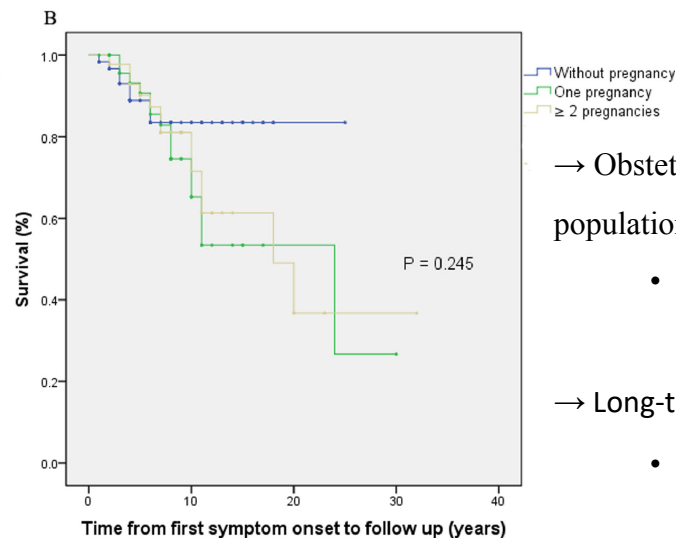
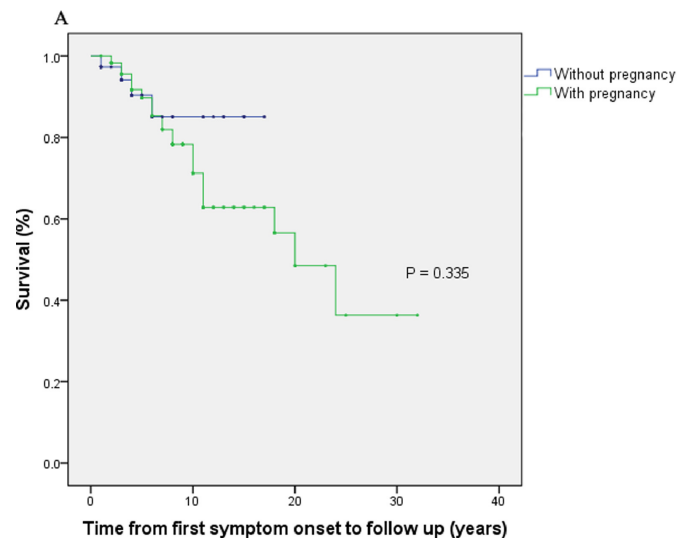
Table 4 Serial investigations in five AC mutation positive female relatives before pregnancy and within 6 months post-partum

	Before pregnancy	Within 6 months post-partum	P-value
RVOT PSAX (mm)	33±6	32±7	0.52
RVFAC (%)	46±7	41±7	0.08
RV free wall LS (%)	-24.3±3.1	-21.9±2.3	0.55
LVEF (%)	54±2	54±2	0.36
LVGLS (%)	-19.5±2.7	-17.9±4.6	0.44

Eur Heart J Cardiovasc Imaging, Volume 20, Issue 2, February 2019
Pages 192–198



224 pregnancies in 120 patients with ARVC



→ Major cardiac events were uncommon during pregnancy

- Sustained VT: 0.8%
- HF requiring hospitalization: 0.8%

→ Obstetric outcomes comparable to the general population

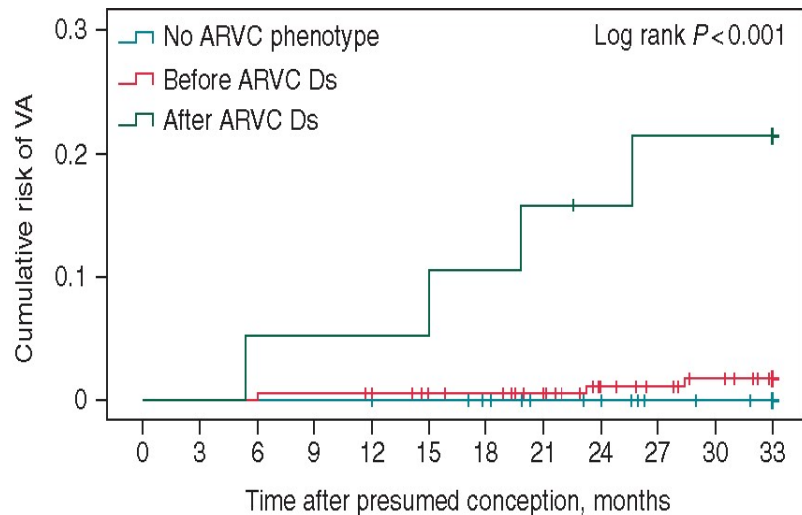
- Spontaneous abortion rate: 13.4%

→ Long-term prognosis driven by disease severity

- Earlier symptom onset: HR 1.046 (p=0.002)
- Lower LVEF: HR 1.127 (p=0.041)

American Journal of Cardiology 2020 125613-617DOI: (10.1016/j.amjcard.2019.11.008)





The risk of VA is primarily related to the degree of phenotypical manifestations of ARVC rather than pregnancy itself.

Pregnancies at risk	0	Birth	1 y	2 y
No ARVC	51	51	43	35
Before ARVC	191	190	178	145
After ARVC	19	18	16	14

Donna di 34 anni

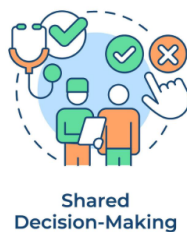
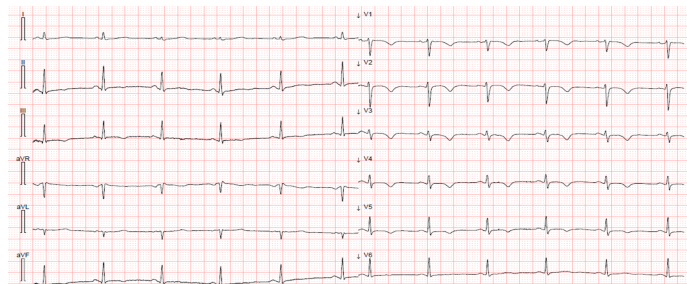
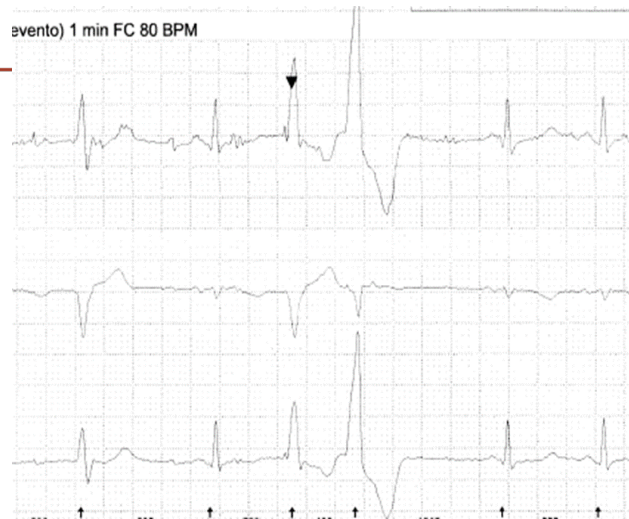
Diagnosi di ACM da variante PKP2 all'età di 27 anni

Asintomatica NYHA I[^]

RV moderatamente dilatato, RVEF: 40%

All'Holter-ECG: 547 BEV/24 h, polimorfi e 5 coppie

Terapia con sotalolo 80 mg 1 cp x2 + ½ cp

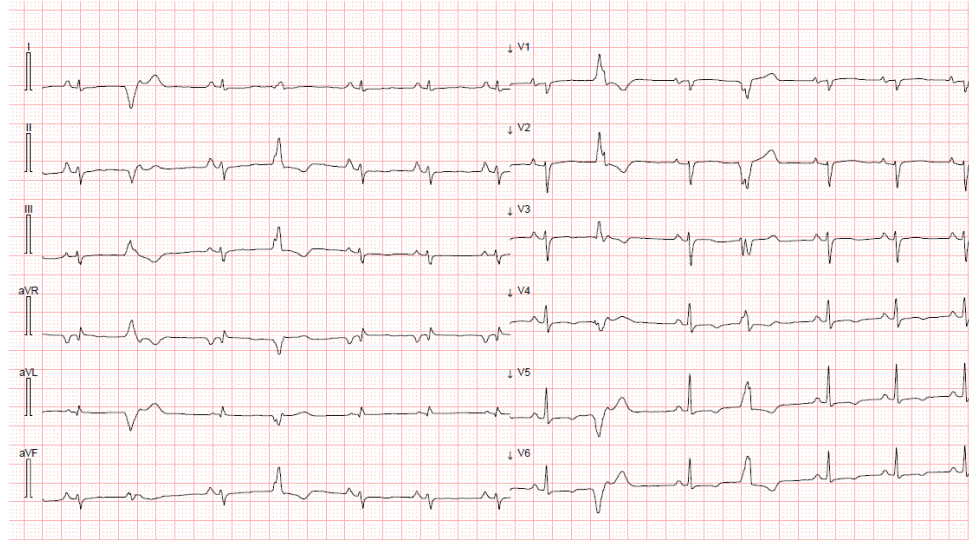


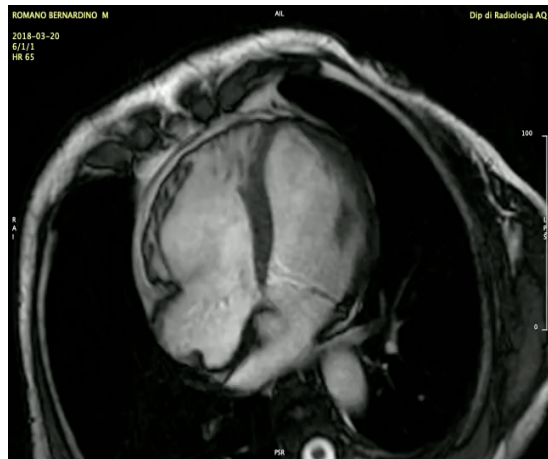
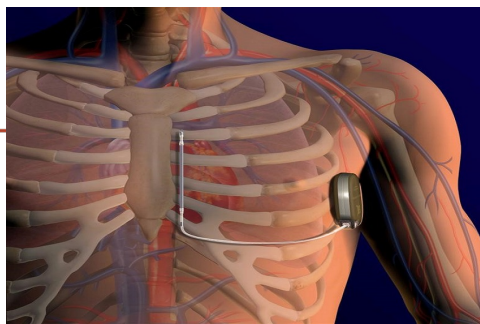
Gravidanza previo
impianto ICD in
prevenzione
primaria



Donna di 29 anni

Diagnosi nel 2019 per extrasistolia ventricolare in sportive con elevato burden extrasistolico anche organizzato al monitoraggio Holter





Impianto di s-ICD in prevenzione primaria

ICD implantation should be considered in patients with definite ARVC and severe RV or LV systolic dysfunction.^{675,691}

Ila

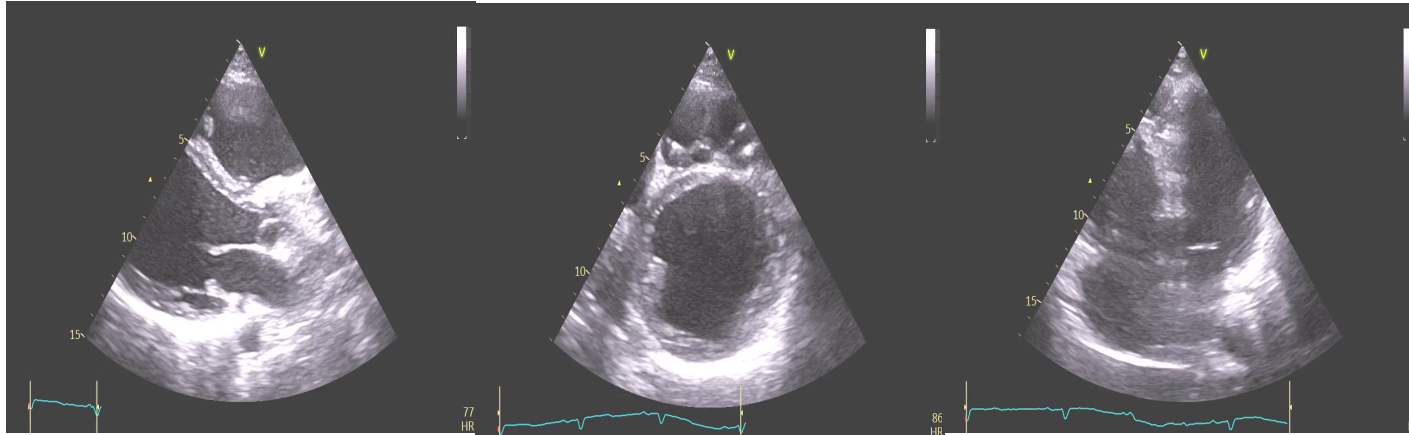
C



Mutazione : ***DSP*** → ACMG 5 (c.939+1G>A)



- **Prima valutazione presso il nostro ambulatorio alla 19^a sg**
- No sintomi
- NYHA I
- Normale crescita fetale
- ECG Holter: SR, numerosissimi BEV polimorfi (15796), 4 RIVA and 3 NSVT.



Parto pretermine programmato (36^{sg}) con taglio cesareo : peso normale alla nascita (2.75 kg)

Monitoraggio in UCIC post partum → non complicazioni nel post partum → dimissione.

POST-PARTUM

Dopo alcuni giorni dalla dimissione : compare dispnea (NYHA II-III) and edemi declivi colonnari → **SCOMPENSO CARDIACO ACUTO** → **UCIC**

BNP 930 pg/mL; NT-proBNP 2526 ng/L

- Terapia diuretica infusiva and ottimizzazione della terapia scompenso (Diuretici , Ace inibitori, Rismarmiatori di potassio e glifozine e beta bloccanti) - Inibita la lattazione (Dostinex)



FOLLOW-UP

Test Cardiopolmonare

Test Submassimale (79% FC massimale)

VO₂ peak 20.0 ml/Kg/min → 64% del predetto

BEV durante esercizio e recupero

Normale risposta pressoria allo sforzo



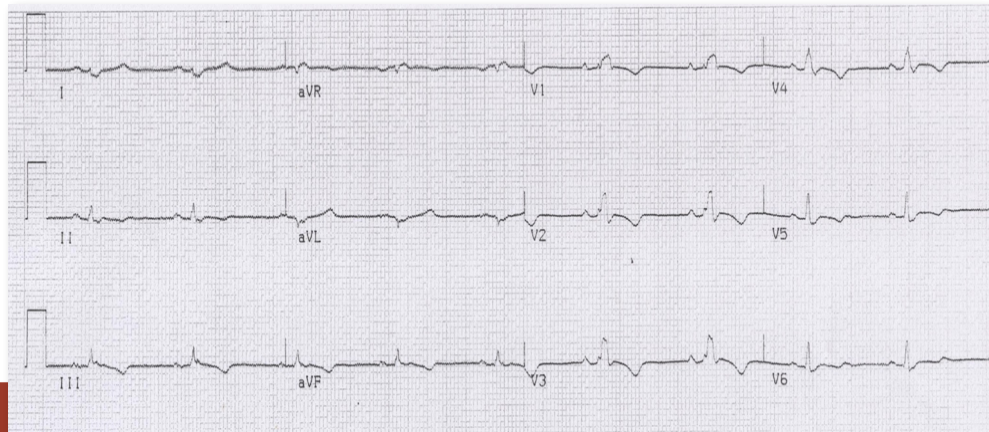
Donna di 28 anni, NYHA II

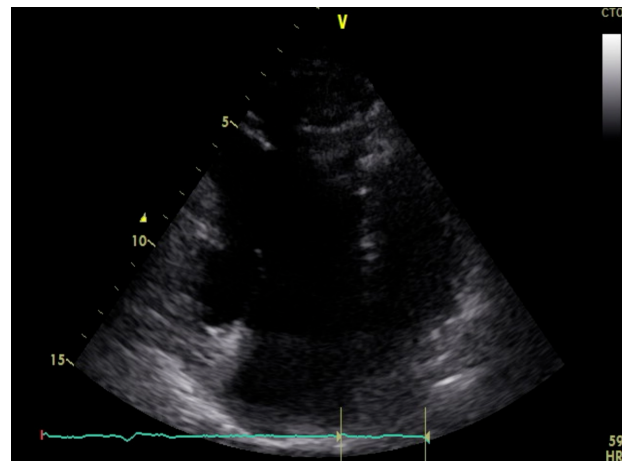
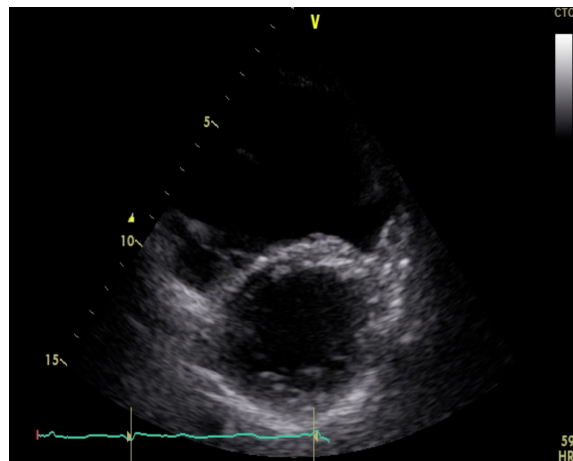
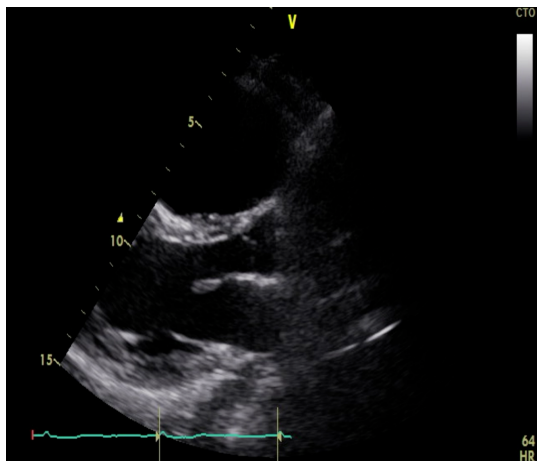
Diagnosi di **ARVC** all età di 18 aa per anomalie ECG in sincope da sforzo

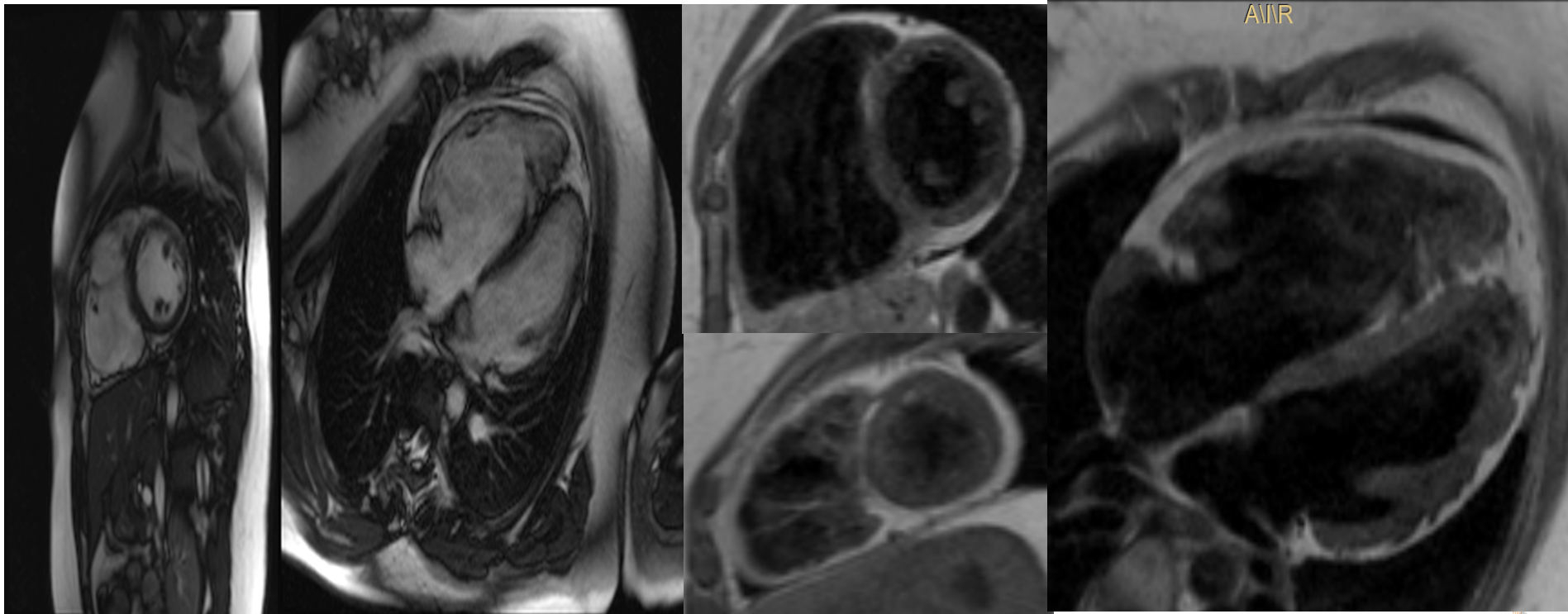
Test genetico negativo

Impianto di ICD in prevenzione primaria

Severa dilatazione e disfunzione ventricolare destra , moderata IT









Gravidanza ad alto rischio mWHOM2.0 III-IV
(mortalità materna 28.9% → 50.3%)

Paziente trapiantata all'età di 33 anni
Gravidanza senza complicanze all'età di 35 anni



- ✓ Pregnancy is usually well tolerated in ACM
- ✓ Pre-conception counselling and Pregnancy Heart Team management are key
- ✓ Do not discontinue beta-blockers (attention to sotalol)
- ✓ Outcomes depend more on phenotypic severity than on pregnancy itself

