

Case report:

A patient with HF and cardiac arrhythmias

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29 y old female patient
family medical history of sudden cardiac death (SCD)



mother died unexpectedly at the age of 35 y
(two kids adopted by host family)

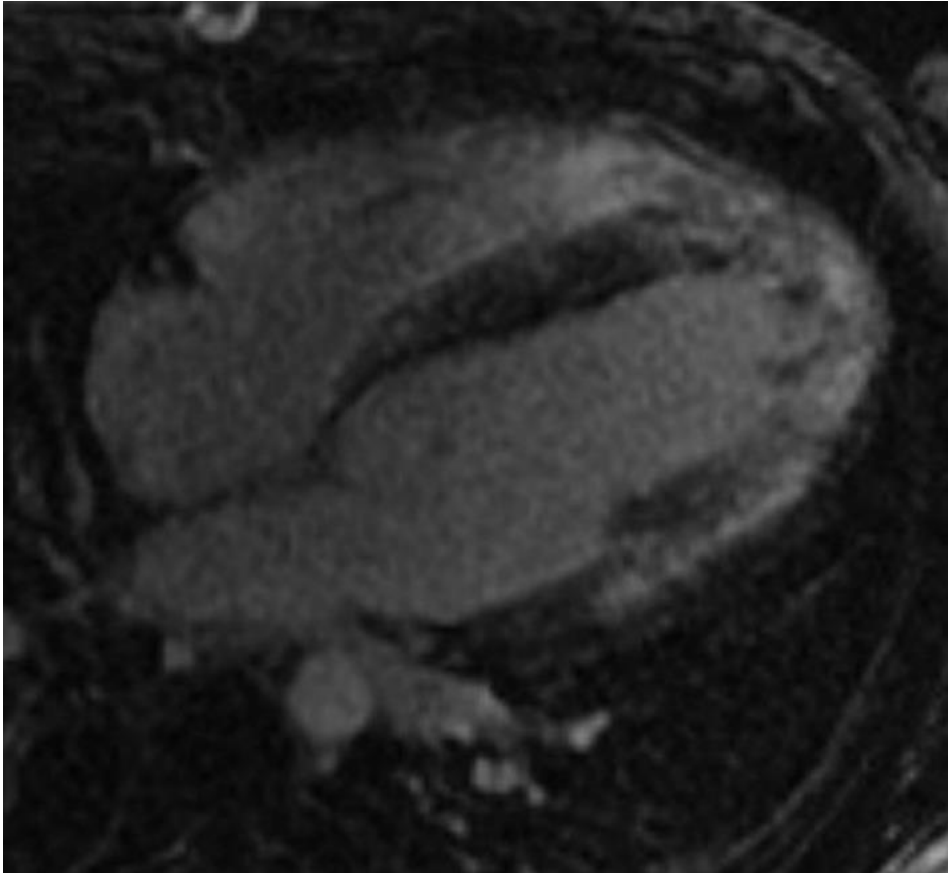


**sister with primary survived SCD (single chamber ICD)
finally died in Age of 29 y by heart failure with fatal VA**

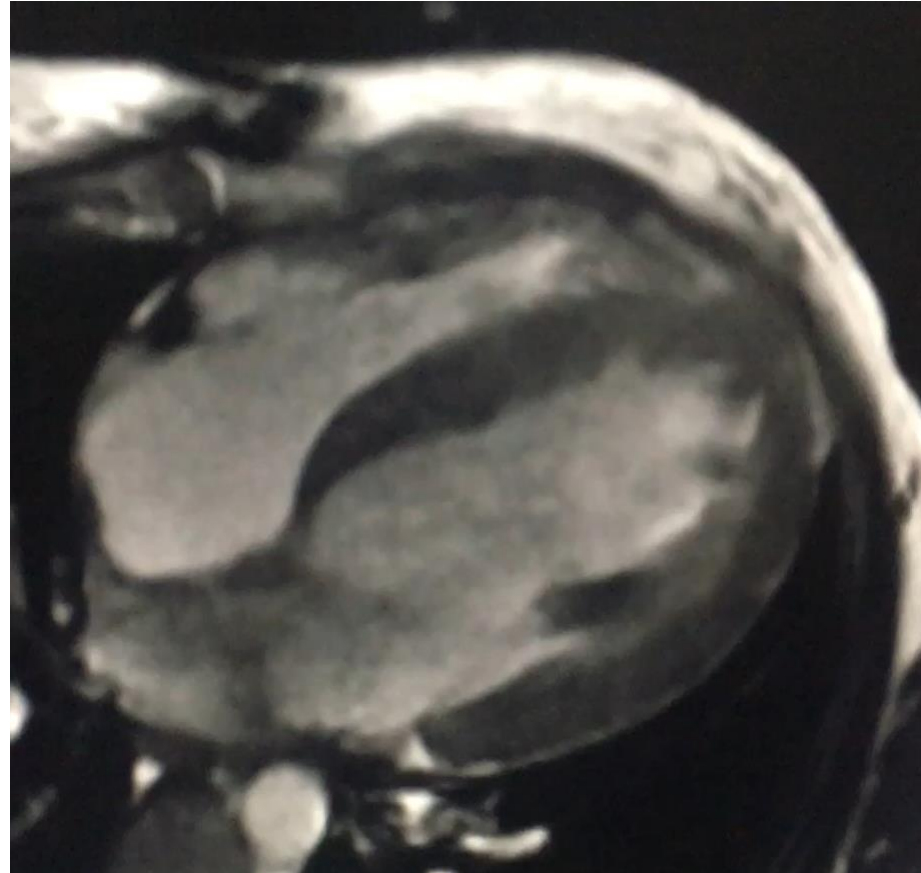


**hypertrophic non obstructive Cardiomyopathy
(HNOCM)**

2012



LGE enhancement



LVEF 50% and eccentric hypertrophy

Medical History

- 2012: Syncope
- 2012: 24h-ECG with PVCs (Couplets) and paroxysmal atrial tachycardia (145 bpm)
- 2014: Single-Chamber ICD-Implantation
- **2015 genetic test**



Mutation of LMNA Gene (laminin A/C)

= LAMINOPATHY

Mutation of LMNA-Gens (laminin A/C)



conduction disorder

- early onset AV-Block
- SVT and Afib
- Ventricular Arrhythmia (VA)

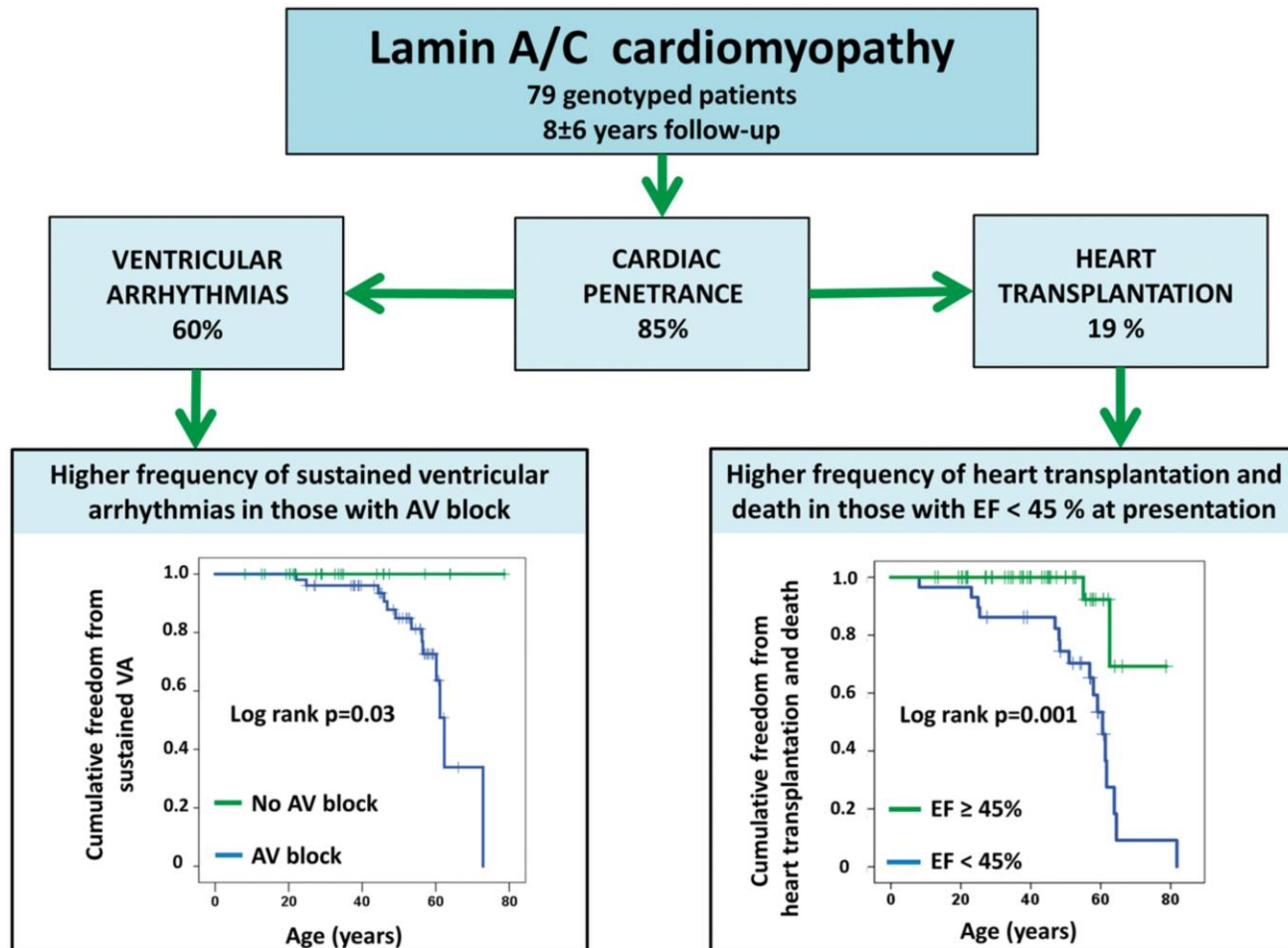


Dilative Cardiomyopathy (DCM)

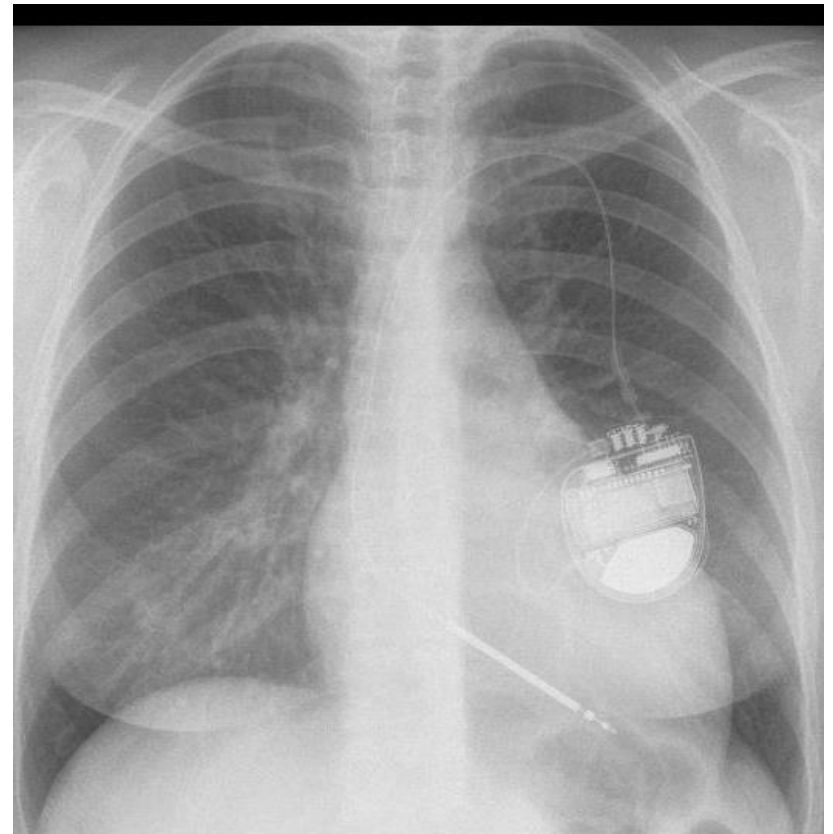
Laminopathy

- 6-8 % of familial DCM
- autosomal dominant
- variable penetration of phenotype

- 2003-2015 genetic testing of 561 patients with familial DCM
- 35 patients with LMNA-Gene mutation (6,5%) and 97 further genotype positive family members



2016 paroxysmal atrial fibrillation,
reduced EF of 40%, NYHA II



2018 persistend tachycardic atrial fibrillation
worsening heart failure, EF 20 % , NYHA III-IV



Troponin T hs	< 14	ng/l	H 97 *5	H 140 *5
NT-proBNP	< 116	ng/l	H 7072 *6	H 7166 *6

existing medical therapy

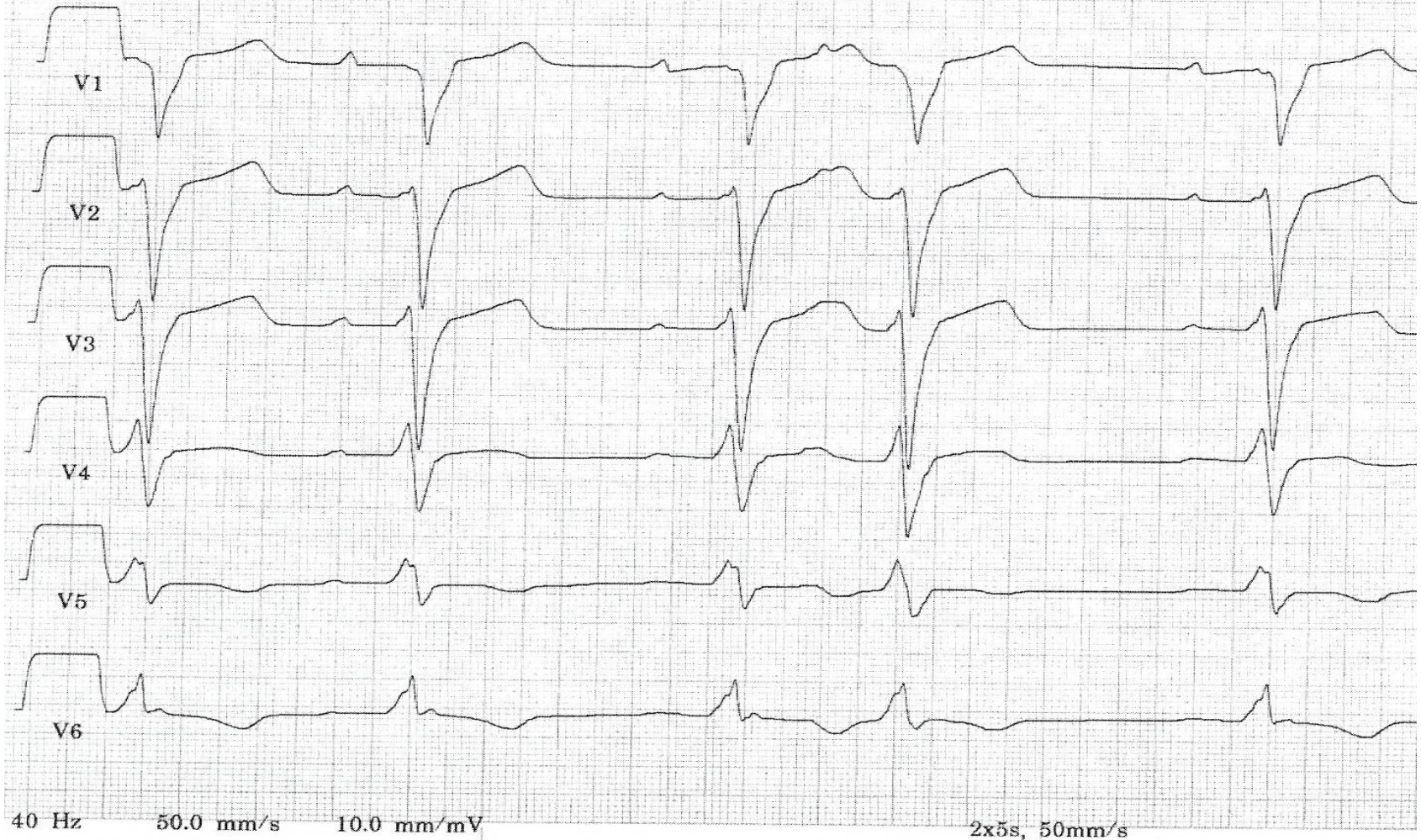
- β -Blocker
- Entrestor (Sacubatil/Valsartan)
- Aldosteronantagonist
- Diuretics

limited by systolic blood pressure of 75-80 mmHg

right and left heart catheterisation

- no coronary heart disease
- cardiac output 2.5 l/min, CI 1.5 l/min/m², PA 41/20/30 mmHg, PCWP 26 mmHg, LVEDP 26 mmHg

improved clinical status after cardioversion
But: 24 h later Afib recurrence despite amiodarone

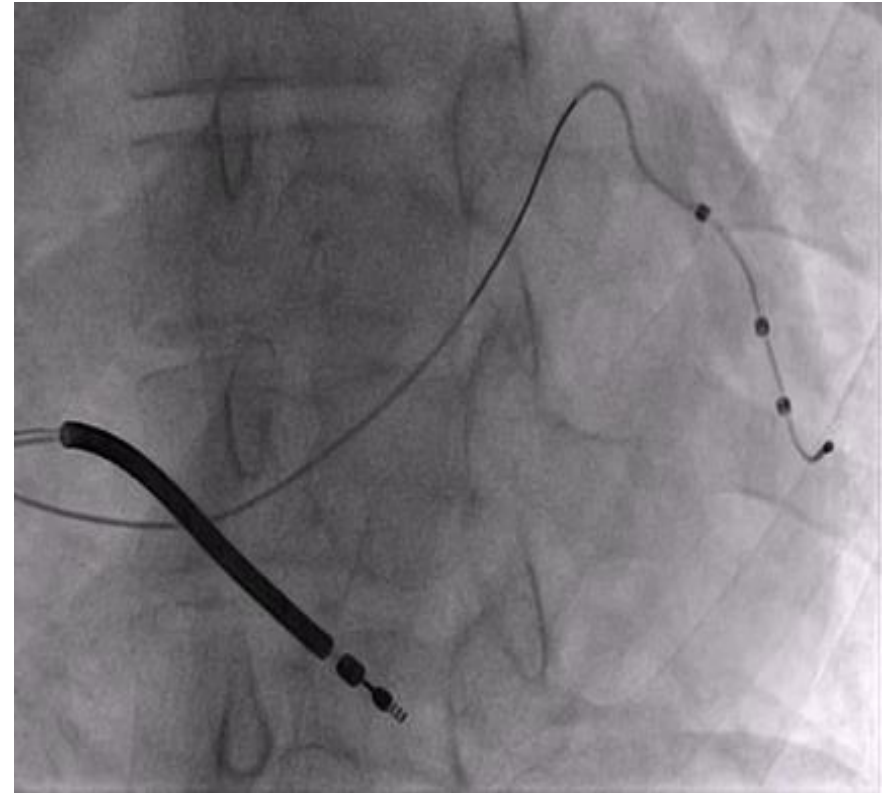
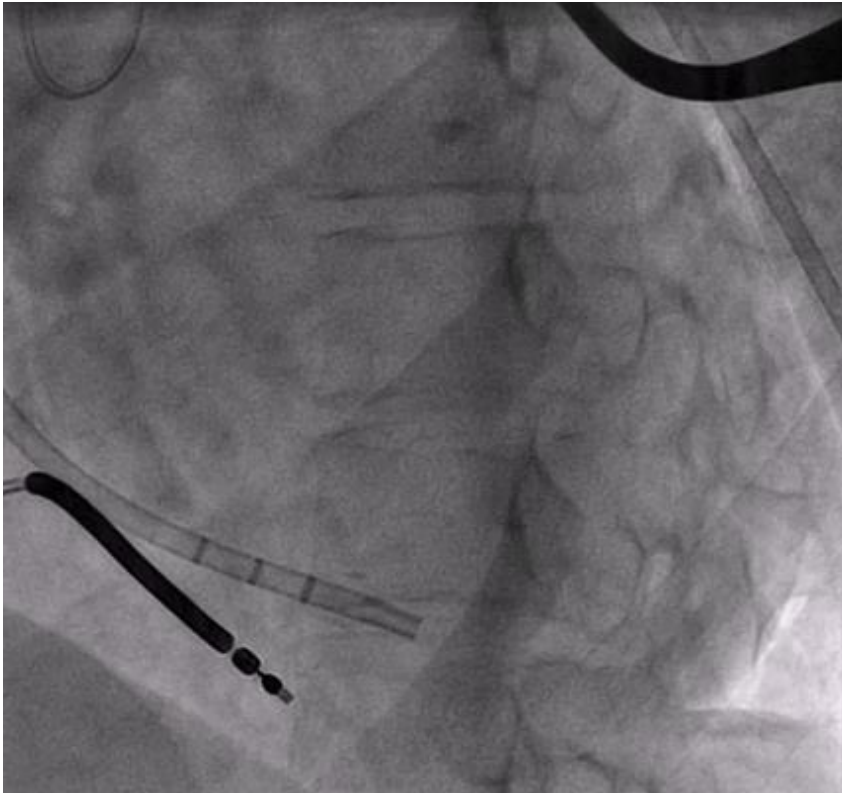


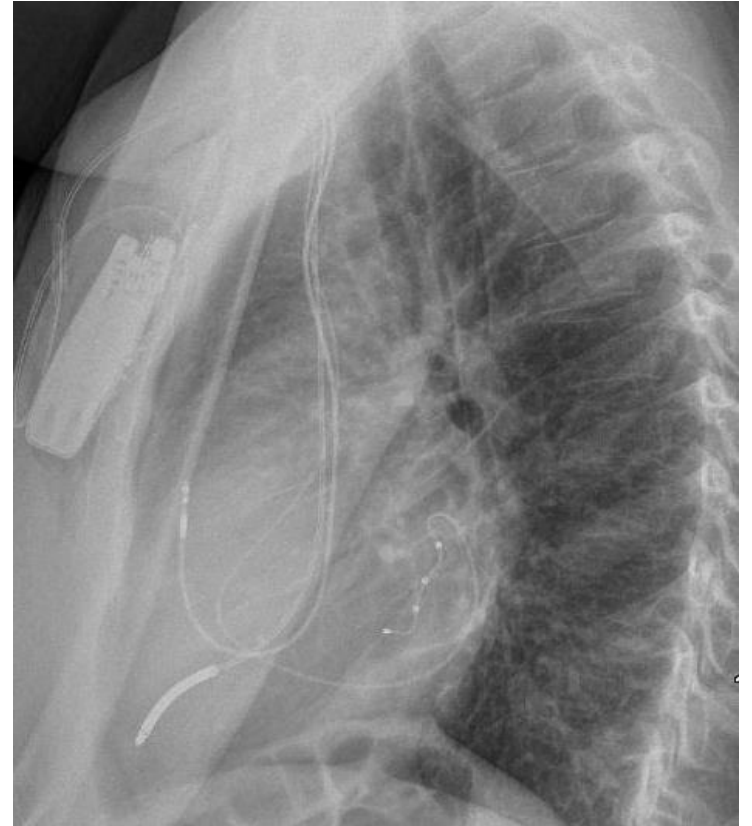
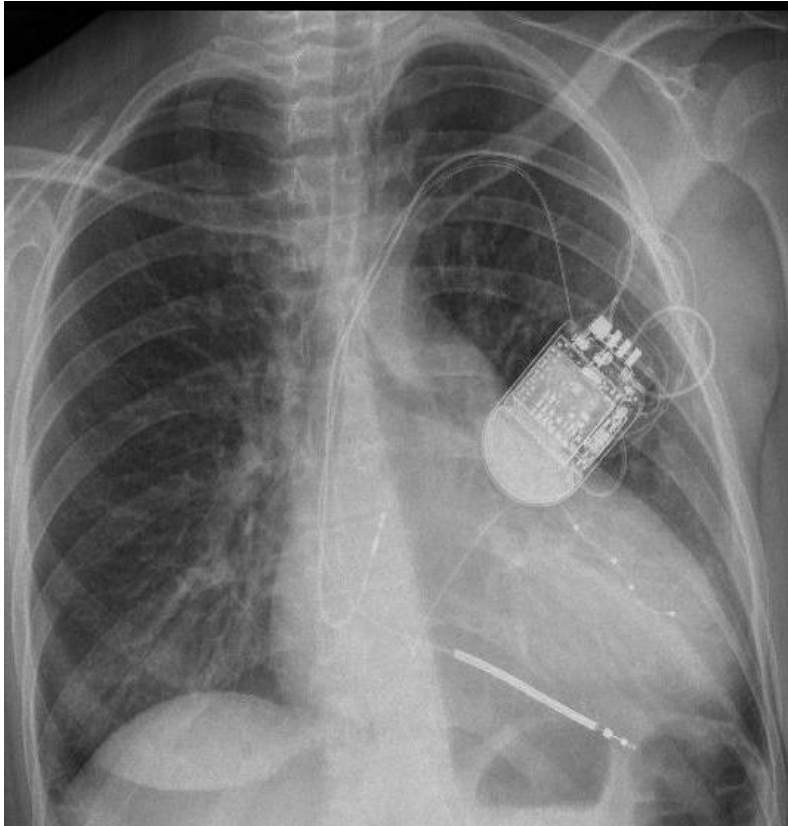
What to do next ?

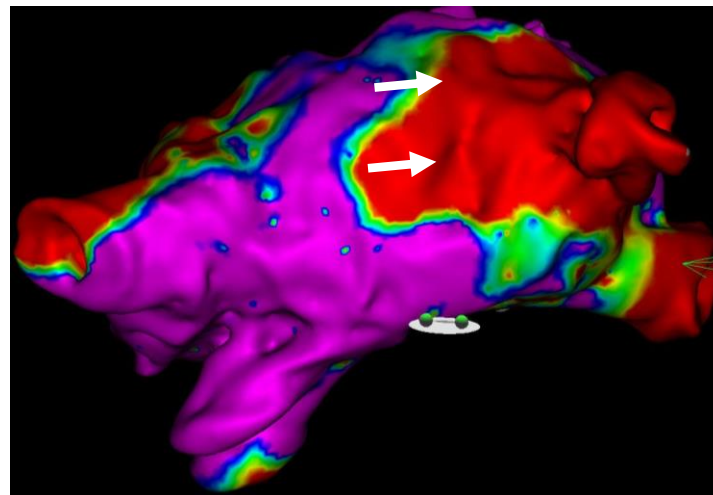
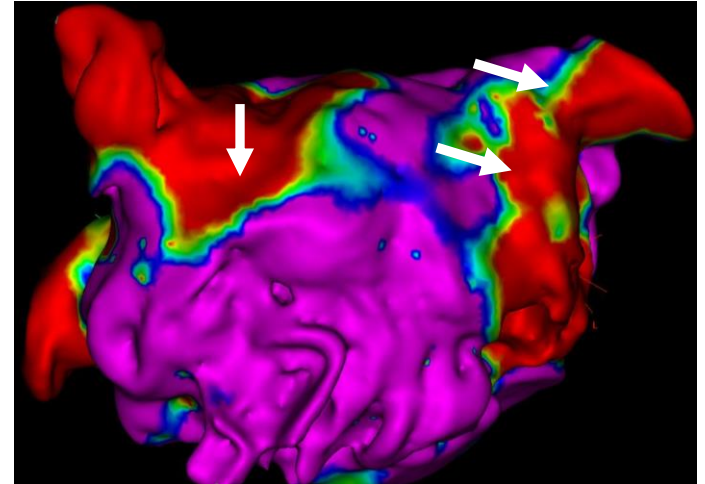
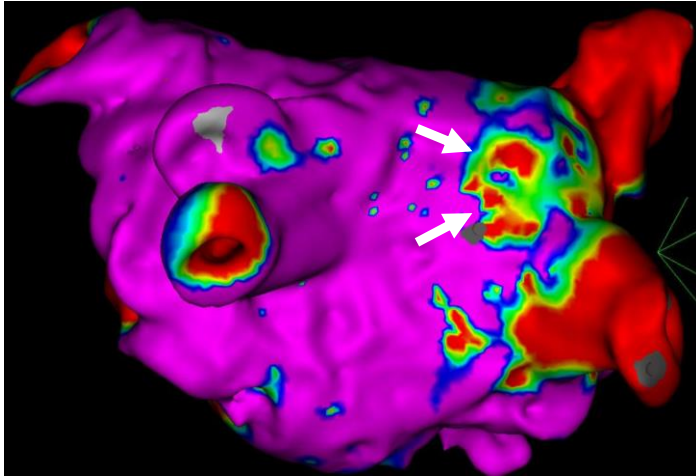
1. Urgent listing for cardiac transplantation ?
2. Scheduling for LVAD implantation ?
3. CRT-D upgrade with AV-nodal-ablation ?
4. CRT-D upgrade with rhythm control (AF ablation) ?

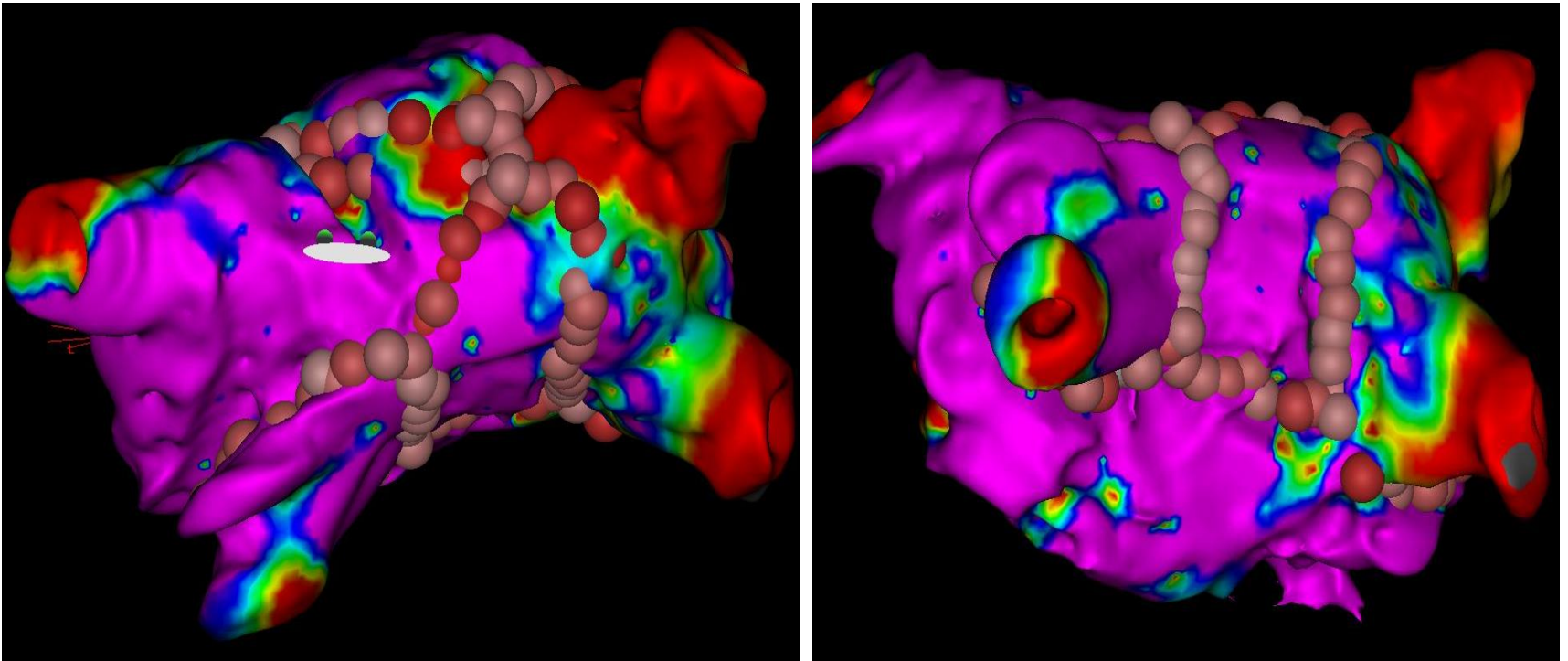
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low voltage zone: anterior wall, roof, patchy fibrosis posterior wall

ablation strategy: PVI, anterior line, roof line, posterior box

Grundfrequenz (GF)	70 min ⁻¹	Detektionsparameter	150 min ⁻¹	184 min ⁻¹	214 min ⁻¹
Ob Grenzfr MTR	95 min ⁻¹	Therapie (AKTIVIERT)	Monitor	ATP x10	ATP x1
Stimuliertes AV-Intervall	140 ms			x10	36,0 J
Wahrg. AV-Int.	100 ms			Aus	40,0 J
V. Stimulation	LV → RV, 80 ms				40,0 J x4

Stimulation und Wahrnehmung

	A	RV	LV
Cap Confirm	Monitor	Aus	Aus
Impulsamplitude (Marge)	2,0 V (3.2:1)	2,5 V	2,5 V
Impulsdauer	0,5 ms	0,5 ms	1,5 ms
SenseAbility™	Ein	Ein	
Empfindlichkeit	Auto A	Auto A	

Diagnostikübersicht

Seit 27 Mai 2018

AP	36 %
BP	75 %
AMS-Episoden	2666
Mode-Switch	27%
AT/AF-Burden	15%

VT/VF-Episoden: 0

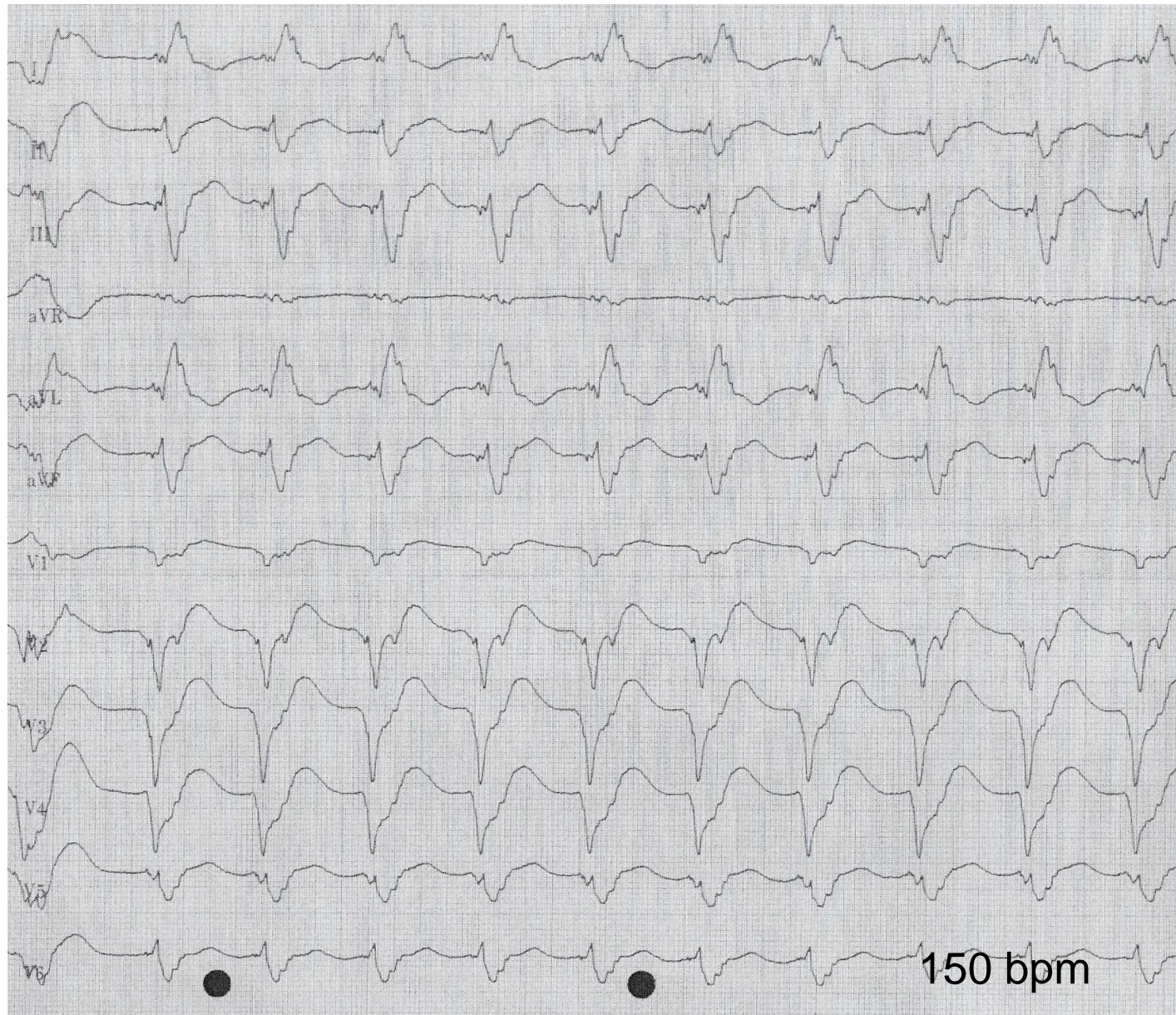
Seit 4 Jun 2018

	VT-1	VT-2	VF
Episoden	0	0	0
ATP abgegeben		0	0
Abgegebene Schocks		0	0
SVT-Episoden: 0			

Warnhinweis



AT/AF-Burden überschritten
Lange AT/AF-Episode(n)
BIV Stimationsanteil unter der unteren Grenze

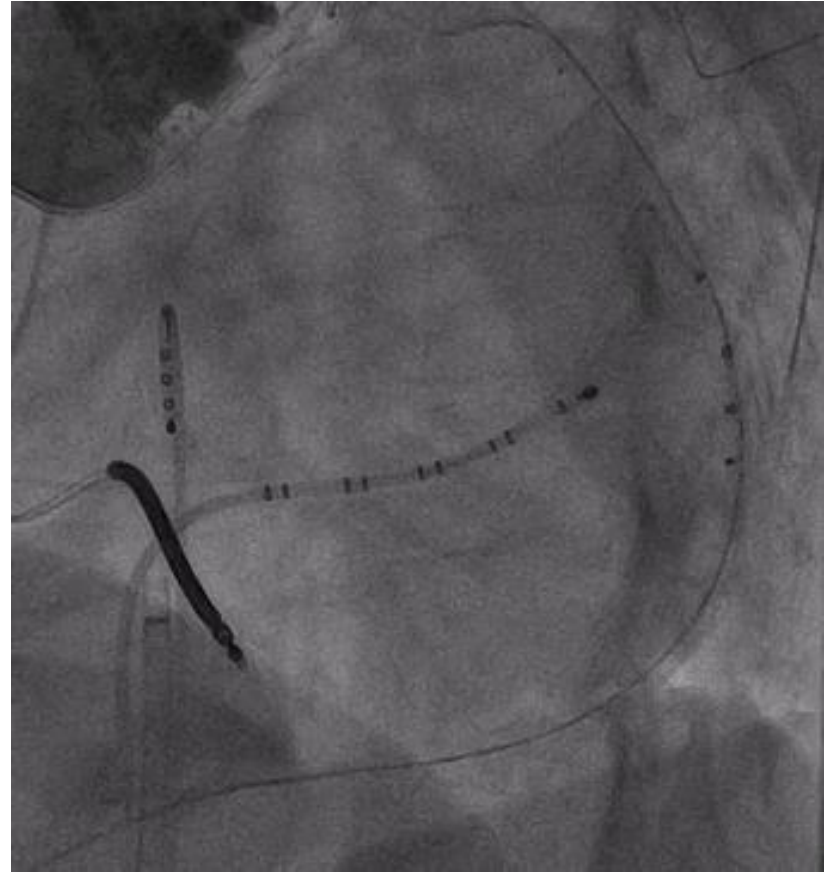
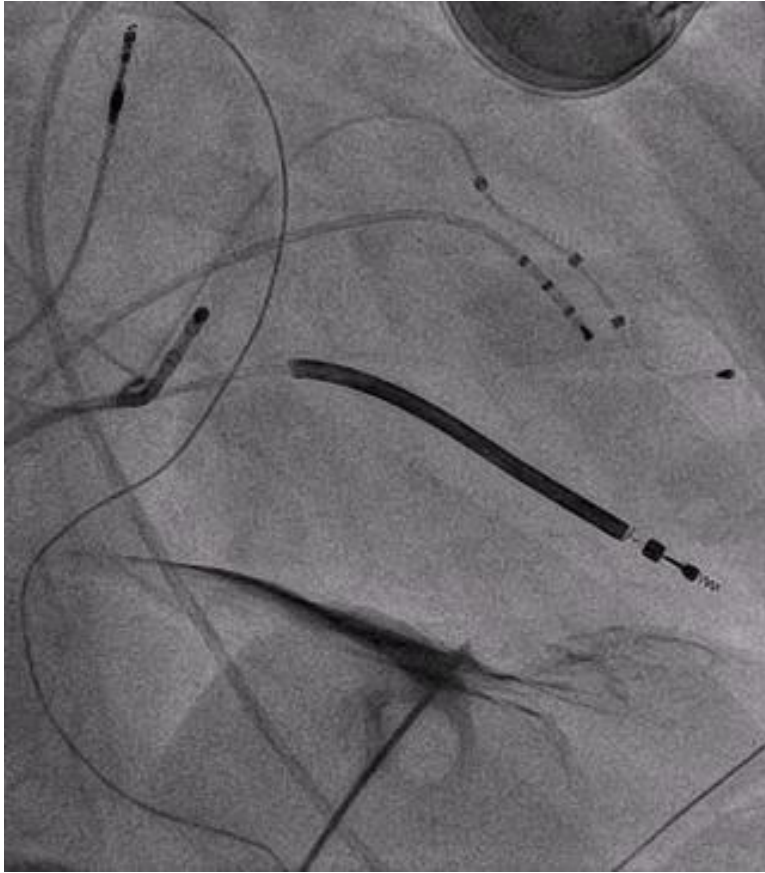


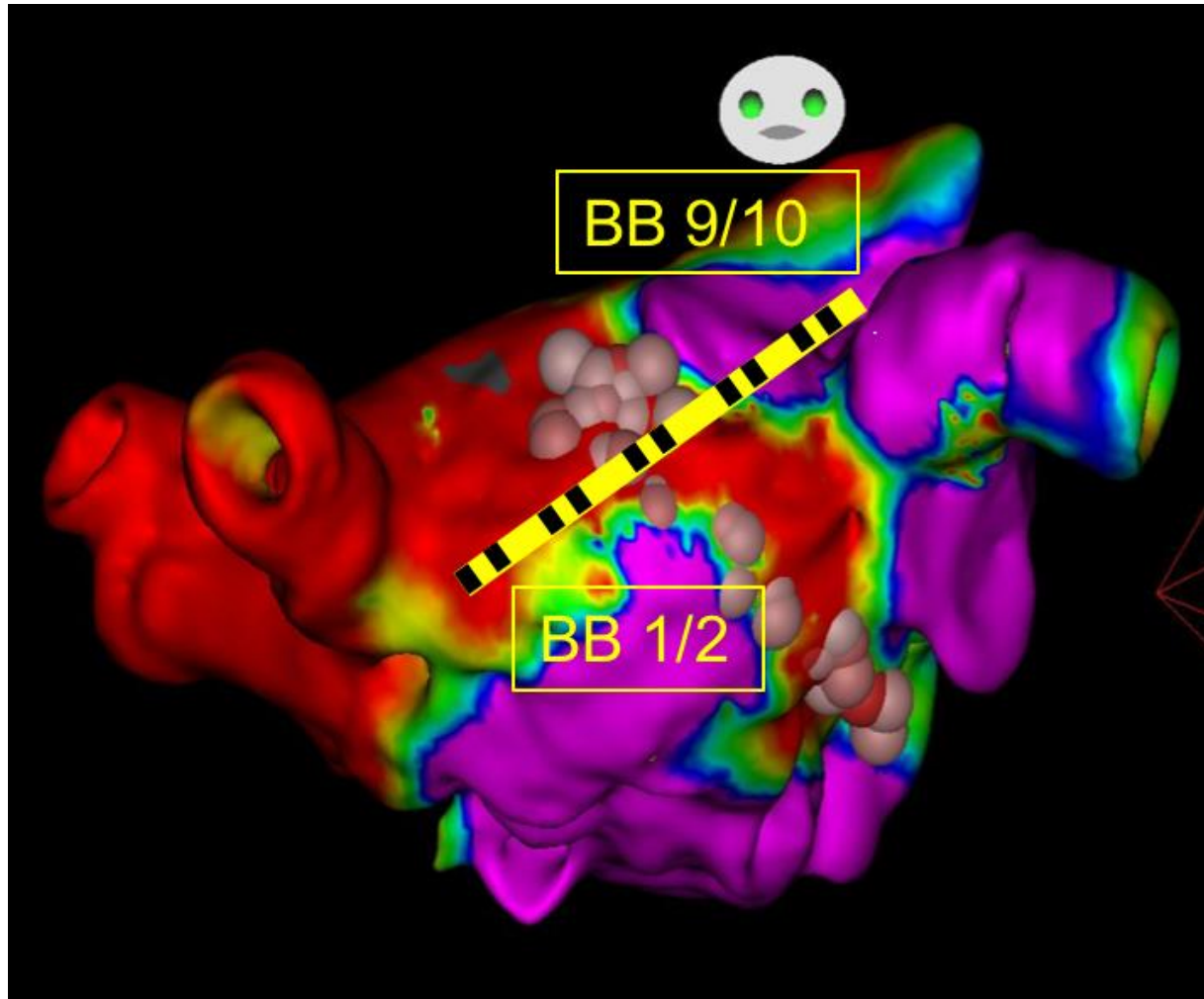
What to do now ?

1. Now LVAD or Transplant ?
2. Re-do catheter ablation ?
3. AV-node Ablation ?
4. Rate control with Digitoxin and β -Blocker ?

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II

V6



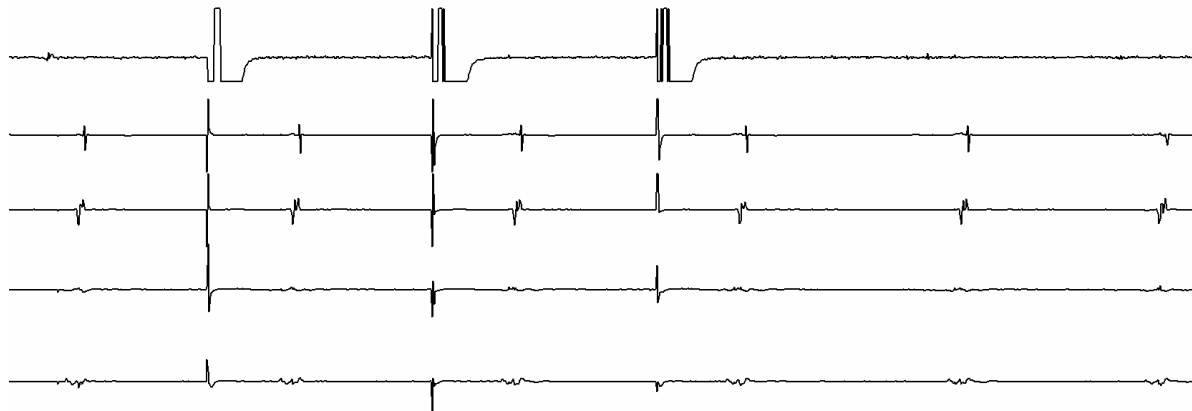
BB 1/2

BB 3/4

BB 5/6

BB 7/8

BB 9/10



CS 9/10

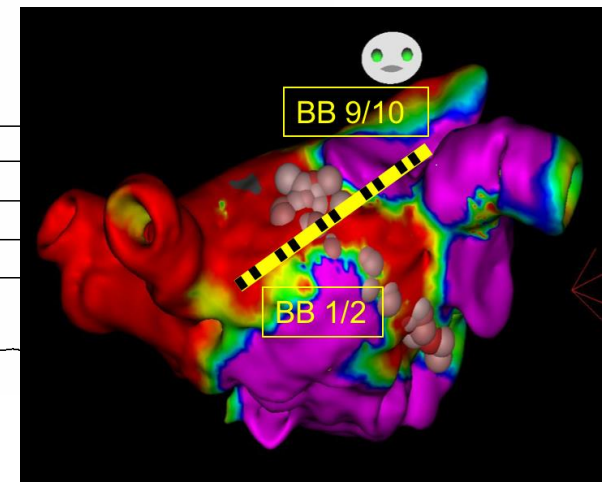
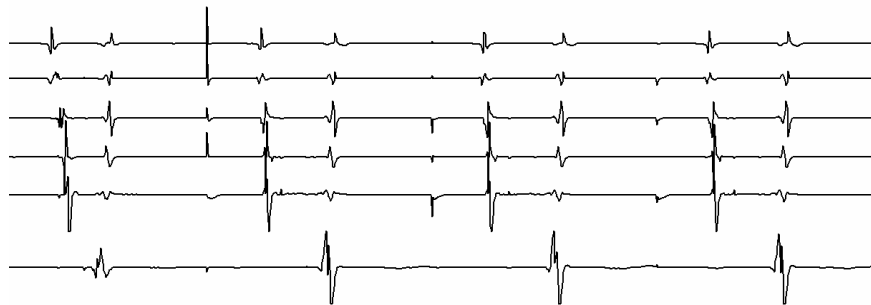
CS 7/8

CS 5/6

CS 3/4

CS 1/2

RV 1/2



I
II
V1

BB 1/2

BB 3/4

BB 5/6

BB 7/8

BB 9/10

CS 9/10

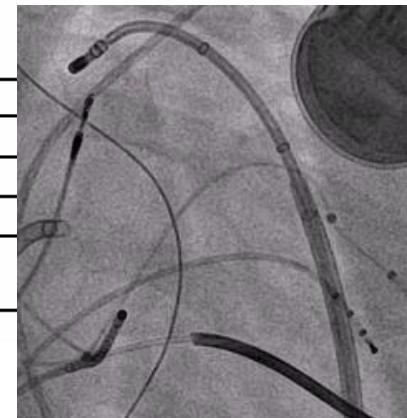
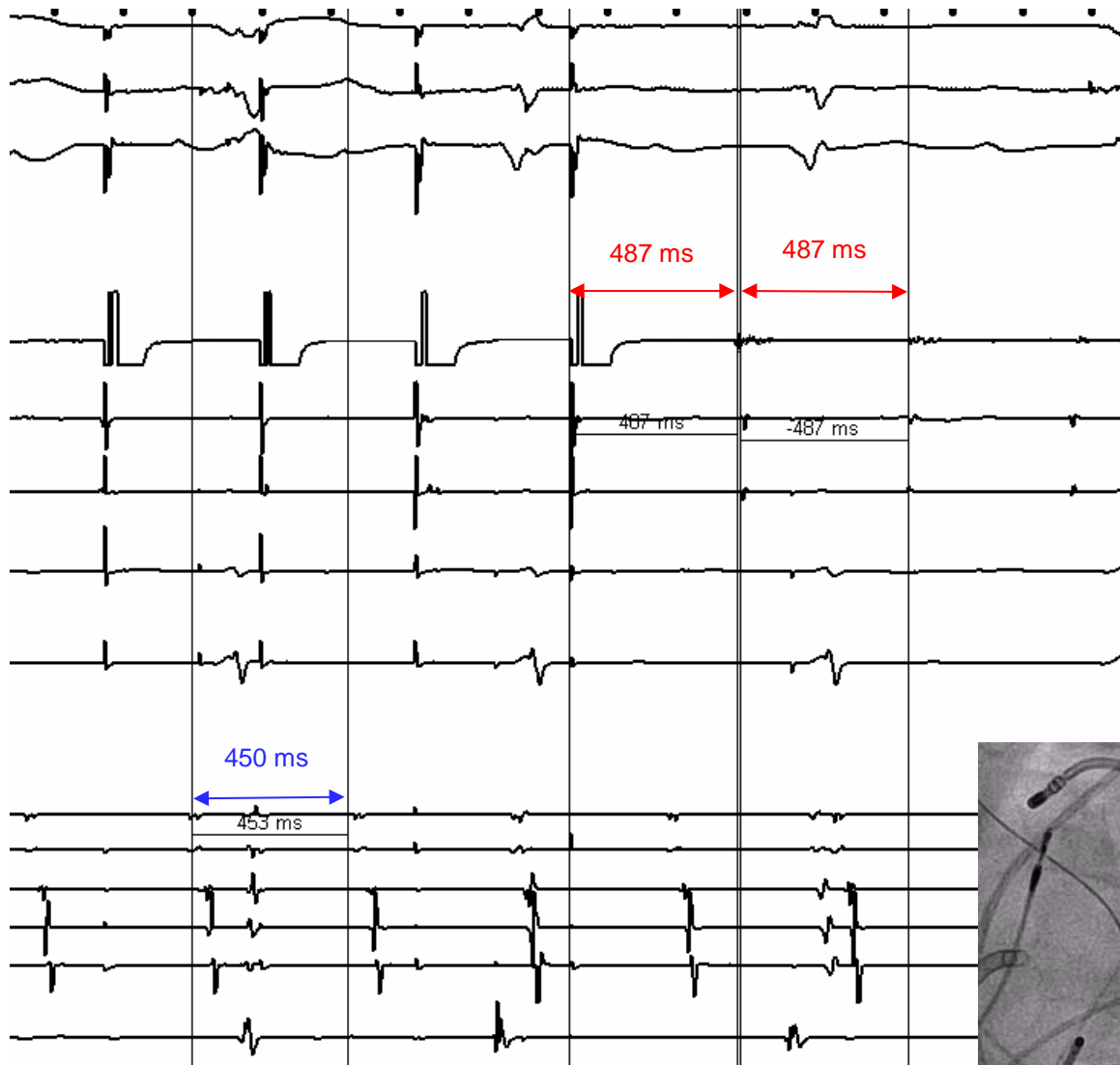
CS 7/8

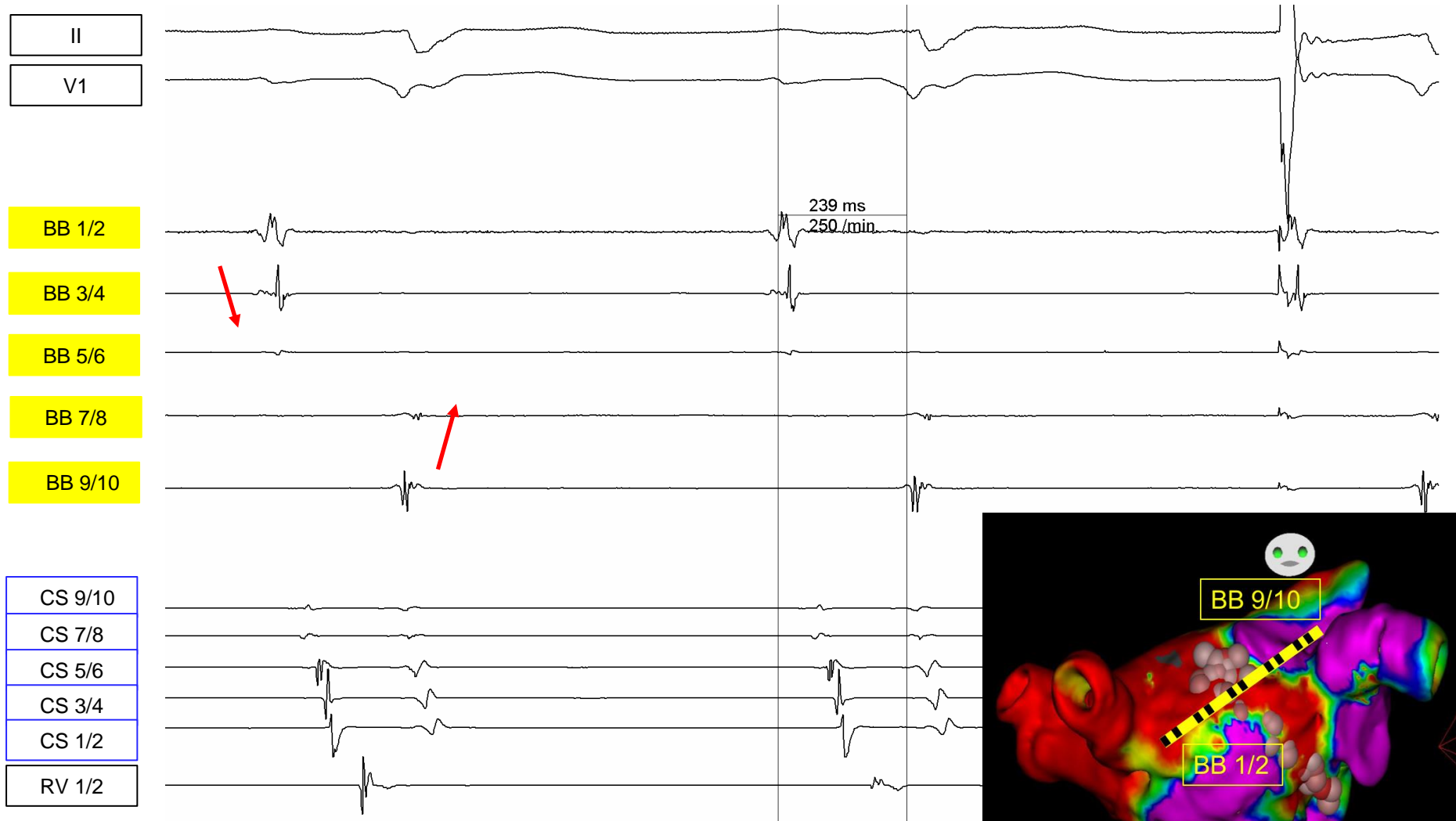
CS 5/6

CS 3/4

CS 1/2

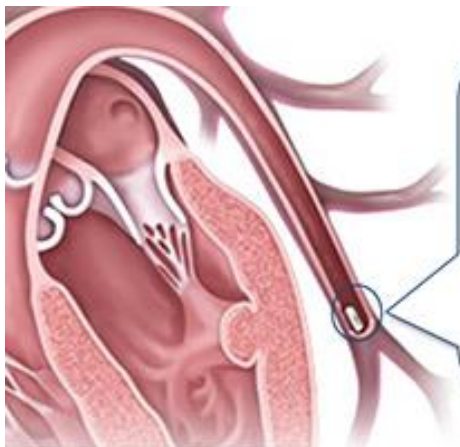
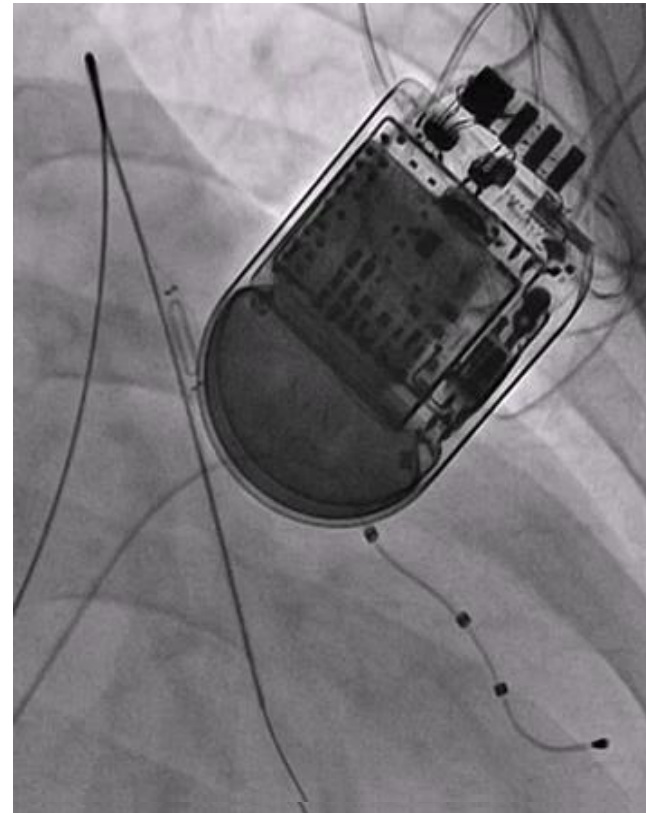
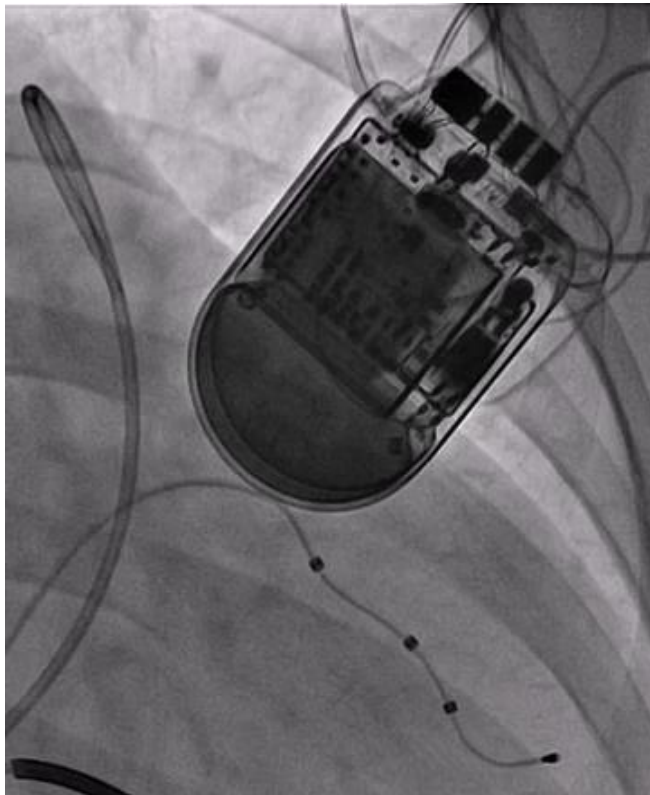
RV 1/2



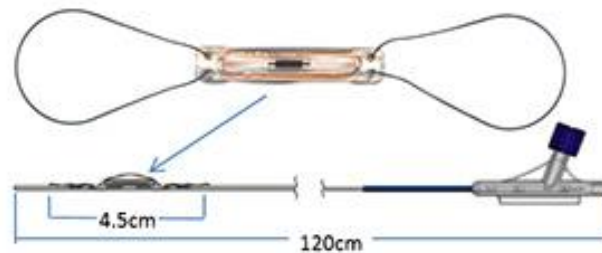




NT-proBNP	< 116	ng/l	H 3867 *1	H 4004 *1	H 2669 *1	H 2668 *1
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PA Sensor and Delivery System

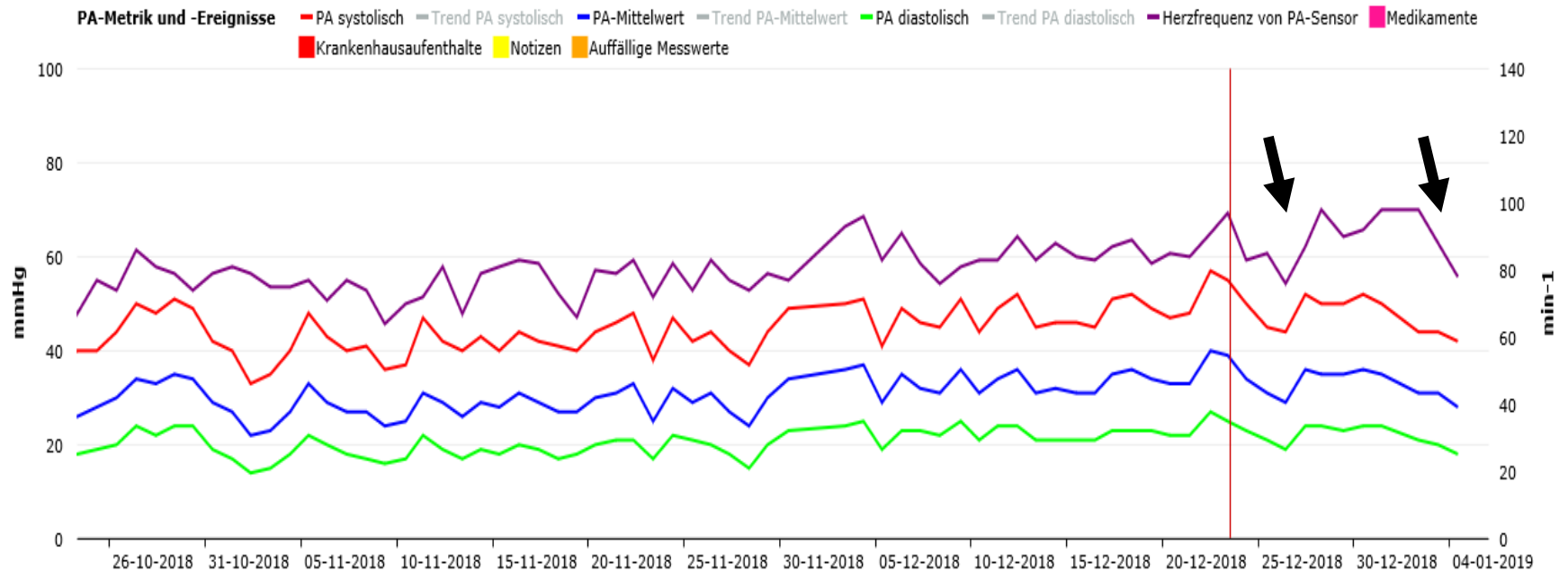


EMB 2019

Konstant Auto

Von: 24-10-2018 Nach: 06-01-2019

Datumsbereich: 30 Tage 90 Tage 180 Tage Alle



12/2018: BNP level: 1500 ng/l

Conclusion

- A case of cardiac laminopathy with manifestation of DCM over a period of 6 years
- **Think about:** young patients with new AV Block, AF / Aflutter, and family history of DCM or SCD – potential laminin A/C mutation
- **Consider:** the three pillars of heart failure therapy - OMT, early rhythm control, and resynchronisation
- **Interesting:** hemodynamic surveillance through a pulmonary artery sensor (Cardio-MEM)



THANK YOU FOR YOUR ATTENTION