Syncope after Mustard repair of transposition

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Antecedents

- 30 year old female
- Born with
  - Transposition of the great arteries
  - Subpulmonary stenosis
- Treated with
  - Rashkind procedure
  - Mustard repair

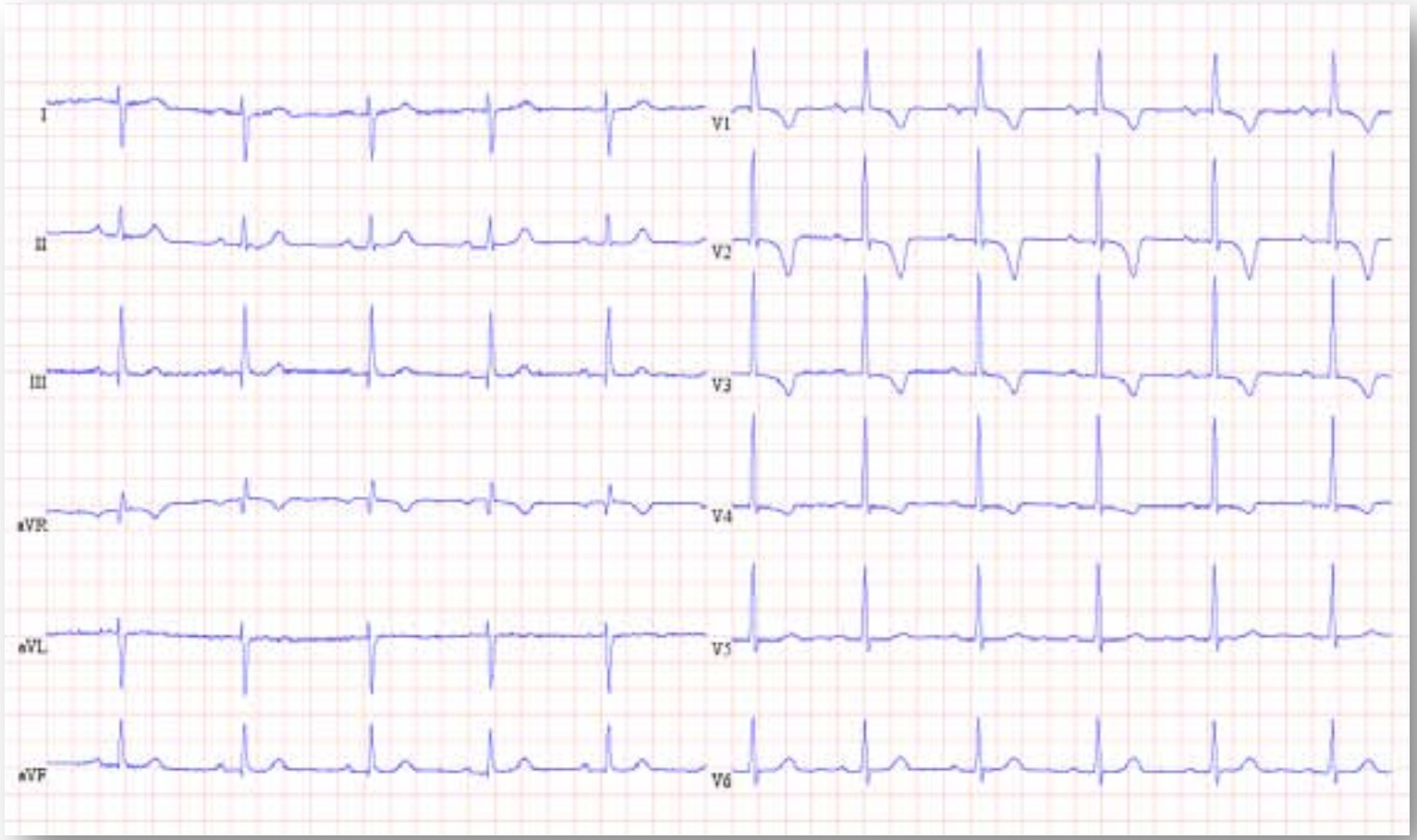
Further interventions

- At the age of 13 years
  - Epilepsy, R/ temporarly with Depakine®
- At the age of 16 years
  - Wall stent implantation in SVC – “LA”
  - Wall stent implantation in IVC – “LA”
- At the age of 21 years
  - Flutter ablation (1:1 conduction, 242 b/min)
  - R/ verapamil 240 mg/d (intol. β-blockers)

Current problem

- Sudden syncope, while sitting and during a phone call; sometimes palpitations
  - Broken tooth and collarbone
- Physical examination
  - Blood pressure 105/70 mmHg, RR 103/min
  - Right ictus
  - Systolic murmur 3-4/6, 2_{nd} ICS left

ECG



Chest X-ray

Transthoracic echocardiography

Well preserved systemic ventricular function, moderate tricuspid valve regurgitation, pulmonary venous baffle: no obstruction, turbulence in systemic venous baffle, subpulmonary stenosis of 70 mmHg

Further tests

- 24h Holter registration
  - Sinus rhythm 46-150/min, rare (S)VES
  - One episode of supraventricular tachycardia (148 b/min for one minute) (mildly symptomatic)
- Bicycle test
  - 100 Watt, maximal heart rate 171 b/min
  - No arrhythmia, no conduction disorders

“What is the next step?”

1. Medical approach, add bètablocker to verapamil
2. Tilt testing
3. Electrophysiological evaluation (bilateral femoral vein thrombosis)
4. Hemodynamic evaluation
5. Combination of 3 and 4

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Motivation to go invasively…

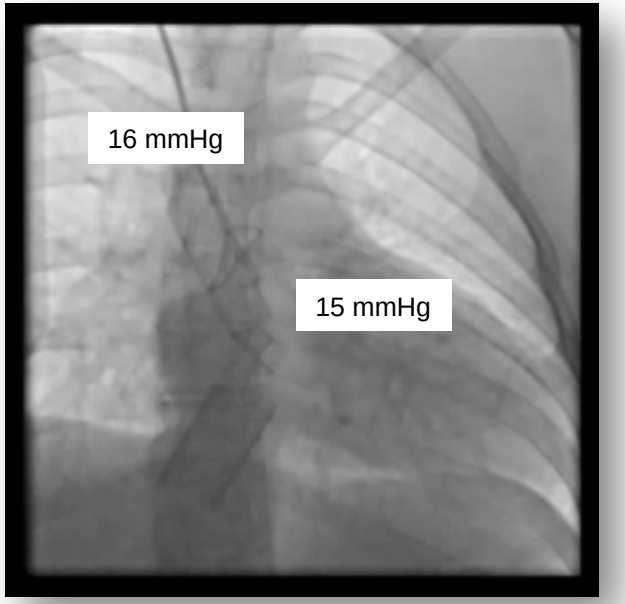
Table 2. Variables for Which Conditional Logistic Regression Analysis Was Performed, Noted in Mean OR With 95% CI

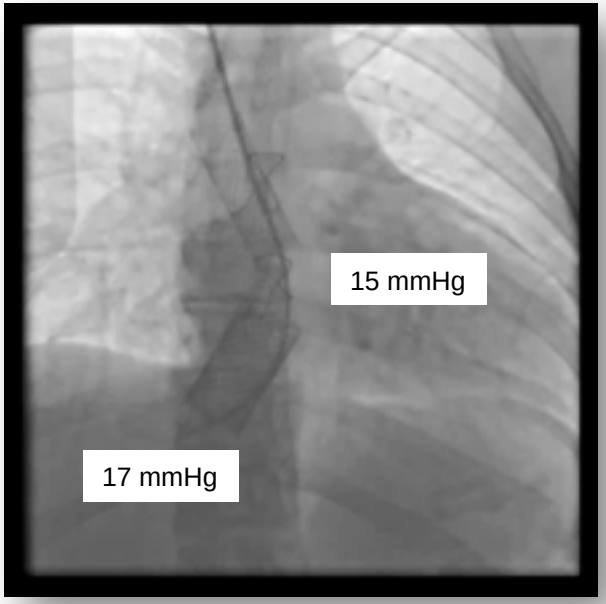
	p Value	OR (95% CI)
Symptoms		
Symptomatic*	<0.0005	6.45 (2.42–17.24)
Arrhythmic symptoms*	0.003	21.60 (2.80–166.79)
Heart failure symptoms*	0.001	4.44 (1.85–10.62)
ECG		
QRS duration	0.723	0.32 (0.001–175.66)
QT interval	0.668	0.16 (0.000–734.27)
QTc interval	0.193	1084.50 (0.029–4.1E + 07)
QRS duration >100 ms	0.251	1.980 (0.618–6.324)
QT dispersion	0.126	0.988 (0.973–1.003)
Heart rate	0.054	1.017 (1.000–1.035)
Basal heart rhythm nonsinus	0.790	1.112 (0.509–2.427)
Chest X-ray		
Enlarged heart size	0.053	2.227 (0.989–5.000)
24-h Holter		
Basal heart rhythm nonsinus	0.037	5.260 (1.10–25.00)
Documented episodes of arrhythmia	0.431	1.770 (0.44–7.25)
Mean heart rate	0.527	0.980 (0.919–1.044)
Minimum heart rate	0.952	1.001 (0.956–1.050)
Maximum heart rate	0.803	0.996 (0.965–1.028)
History of arrhythmia in follow-up		
Documented arrhythmia*	0.005	3.473 (1.451–8.310)
Documented SND in follow-up	0.035	2.405 (1.065–5.432)
Documented AFL/AF in follow-up*	0.001	4.866 (1.900–12.462)
Arrhythmia treatment		
Pacemaker implantation	0.550	0.641 (0.149–2.758)
Medication treatment*	0.002	5.159 (1.863–14.283)

*Statistically significant risk factor ($p < 0.005$).
AFL/AF = atrial flutter/atrial fibrillation; CI = confidence interval; OR = odds ratio; SND = sinus node disease.

Kammeraad et al. J Am Coll Cardiol 2004

Hemodynamic data





Electrophysiological data

- Induction of atrial flutter 1:1 (> 200 b/min)
- No induction of ventricular arrhythmias

Ventricular testing useful?

Table 2. Predictors of Appropriate ICD Shocks

Variable	Hazard Ratio	95% CI	*P* Value
Univariate analysis			
Secondary prevention indication	5.1	1.1,45.5	0.0375
Ventricular septal defect	4.3	0.9,20.8	0.0742
At least moderate tricuspid regurgitation	4.1	0.8,20.5	0.0912
QTc, ms	1.02	1.00,1.05	0.0767
Lack of β-blockers	11.3	1.3,100.1	0.0303
Multivariate analysis			
Secondary prevention indication	18.0	1.2,261.0	0.0341
Lack of β-blockers	16.7	1.3,185.2	0.0301

EP study was not predictive for appropriate shocks in primary prevention for dTGA

Khairy et al. Circ Arrhythm Electrophysiol 2008

“What is the next step?”

1. Flutter ablation (low succes rate expected)
2. Percutaneous hemodynamic optimization (stent implantation baffles)
3. First flutter ablation and then stent implantation
4. First stent implantation and then flutter ablation

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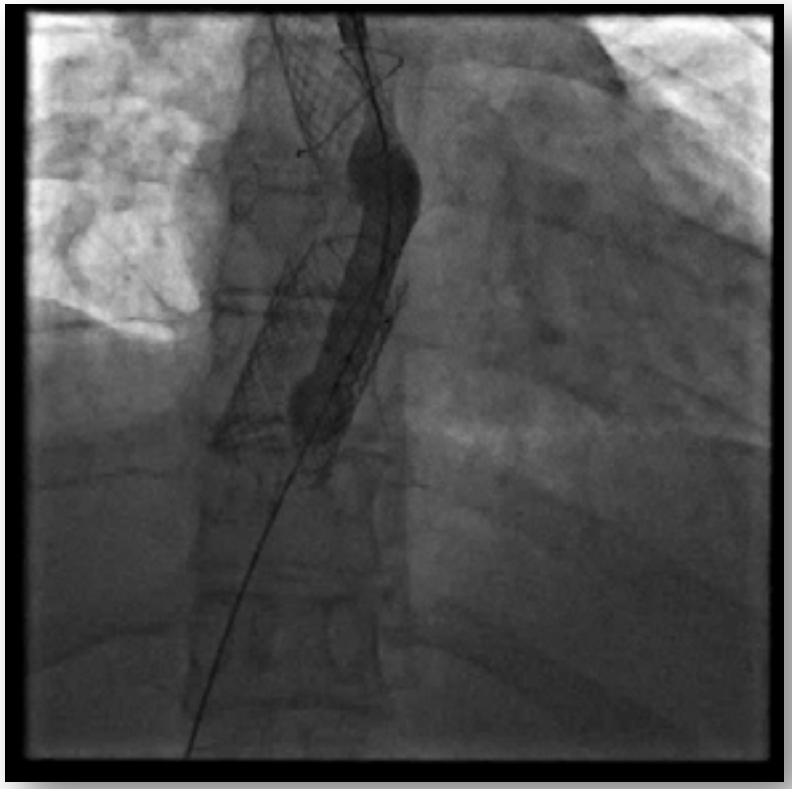
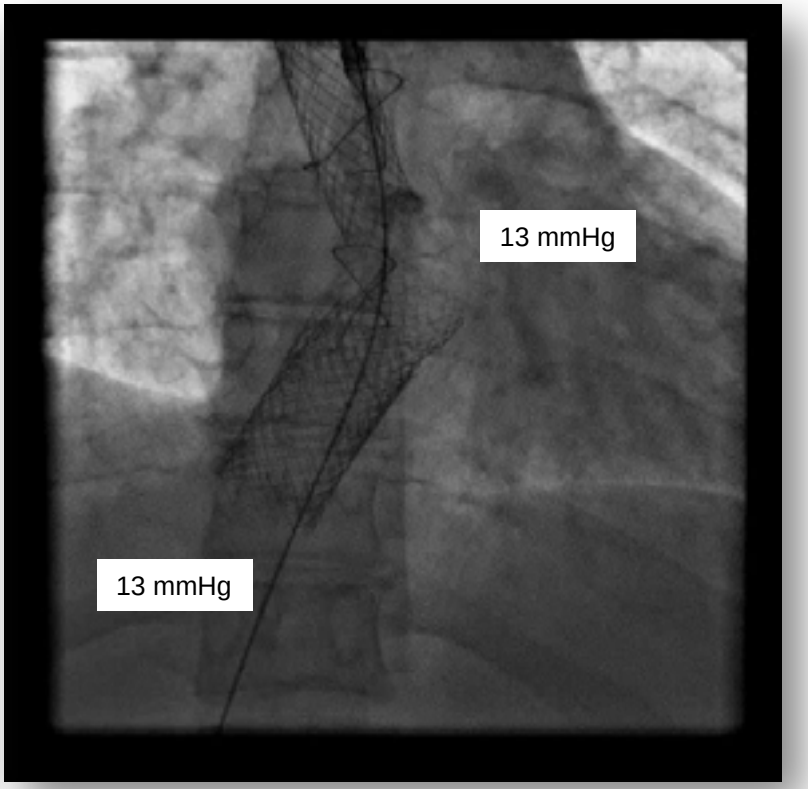
IVC – LA stenting (Intrastent maxi LD 20 mm)





Further follow-up

- Persistent intermittent palpitations without syncope
  - Several times per week, 30 minutes
  - Progressive adynamic behaviour
  - New syncope with clinical documented bradycardia (under verapamil 240 mg/d)
- Electrophysiological testing
  - No atrial tachycardia inducible – no ablation

“What is the next step?”

1. Change of medical treatment: start amiodarone
2. Change of medical treatment: add flecainide to verapamil
3. Focus on hemodynamic optimization: treatment of subpulmonary stenosis
4. Pacemaker implantation and Hiss ablation

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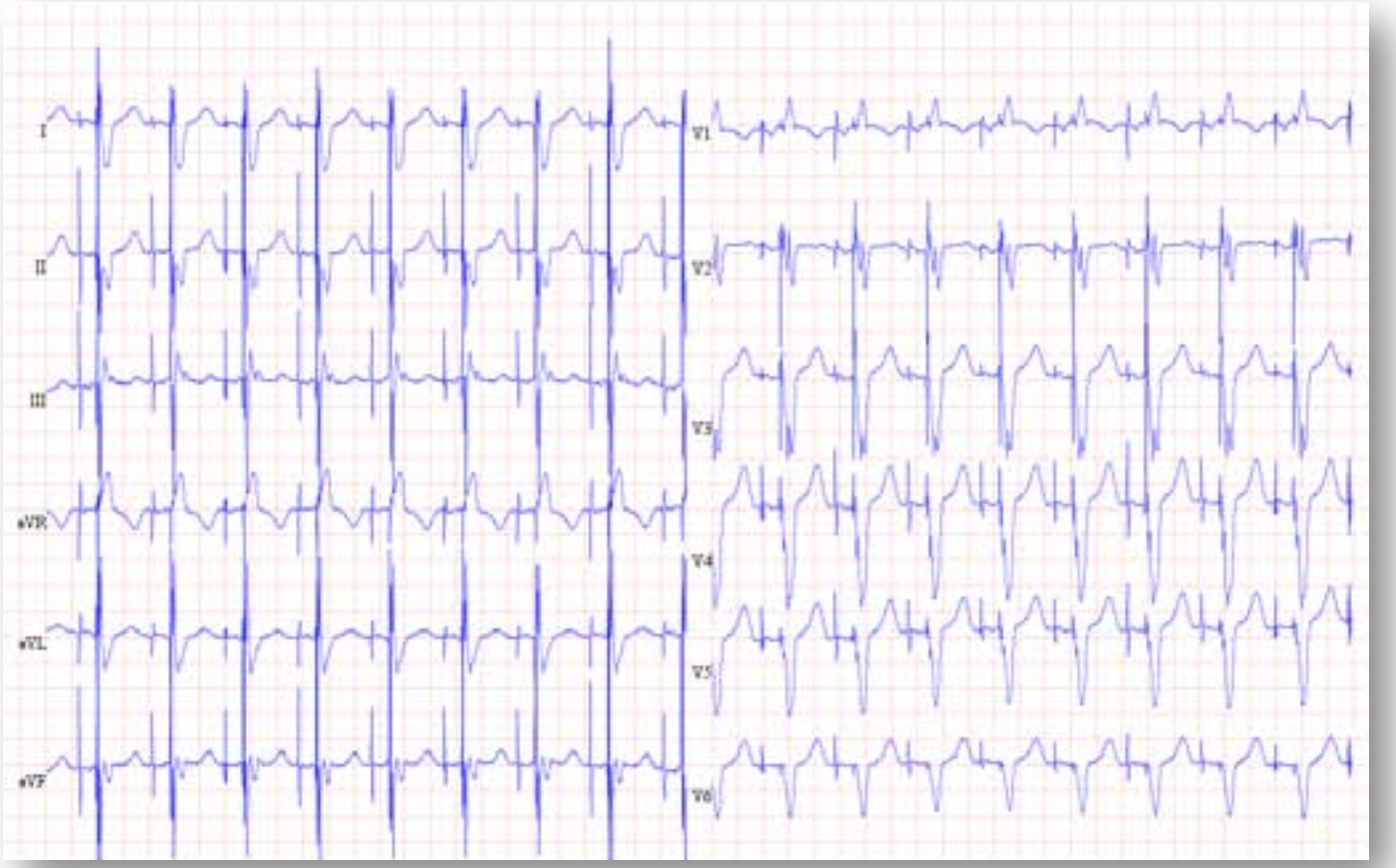
Epicardial PM leads

DDD biventricular pacing 60 – 120/min; mode switch at 160 b/min

ECG



Further follow-up

- No palpitations under verapamil
- Functional class I-II
- No syncope
- Persistent socio-professional integration

Conclusions

- In case of complex heart disease:
  - Arrhythmias might be the first sign of worse outcome
  - Low threshold for invasive measurements (hemodynamic and electrophysiological)
  - Structural anatomical optimization remains important, also in case of electrical problems