

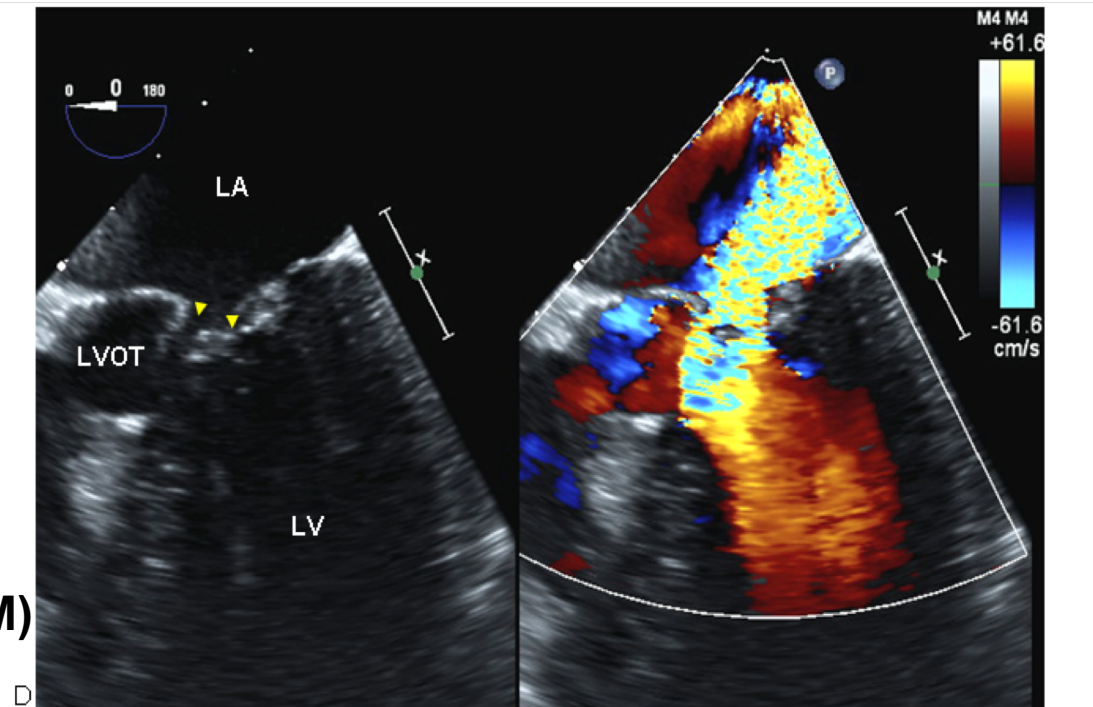
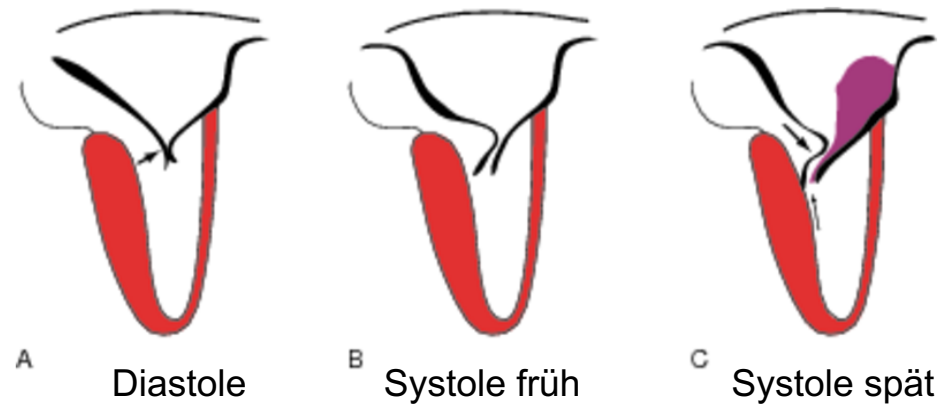
PATHOPHYSIOLOGIE LVOT-OBSTRUKTION

PHYSIK

- Normale Flussgeschwindigkeit im LVOT = 1-1.5m/s
- Kontinuitätsgleichung ($dP_{max} = 4(V_{max}^2 - V_{prox}^2)$)
- Obstruktiv > 30mmHg

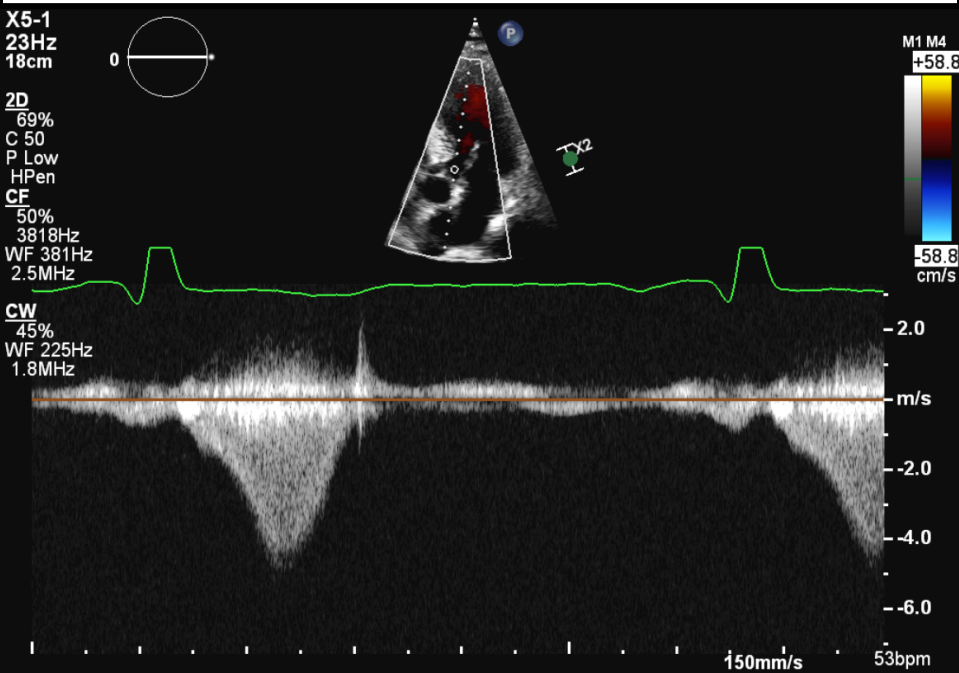
ANATOMIE

- Septumhypertrophie
- Mitralklappenanomalien, grosse und lange Segel
- Papillarmuskelanomalien
- Flussbeschleunigung im LVOT – Venturi – Effekt – Systolic Anterior Movement der Mitralklappensegel (SAM)



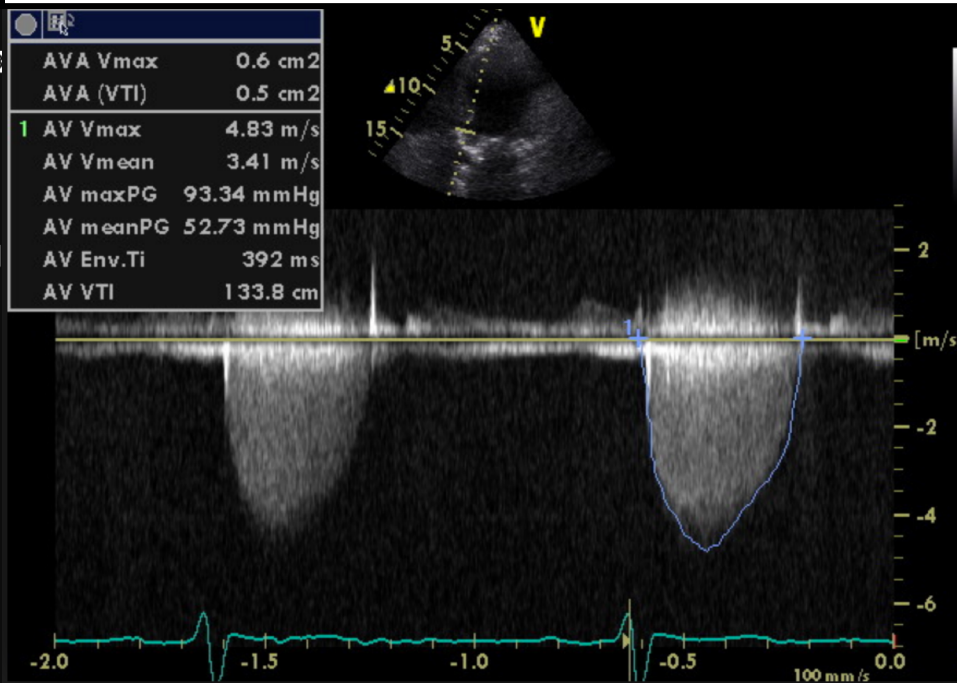
LVOT-OBSTRUKTION – DYNAMISCH vs FIX

DYNAMISCH



‘dagger-shape’

FIX

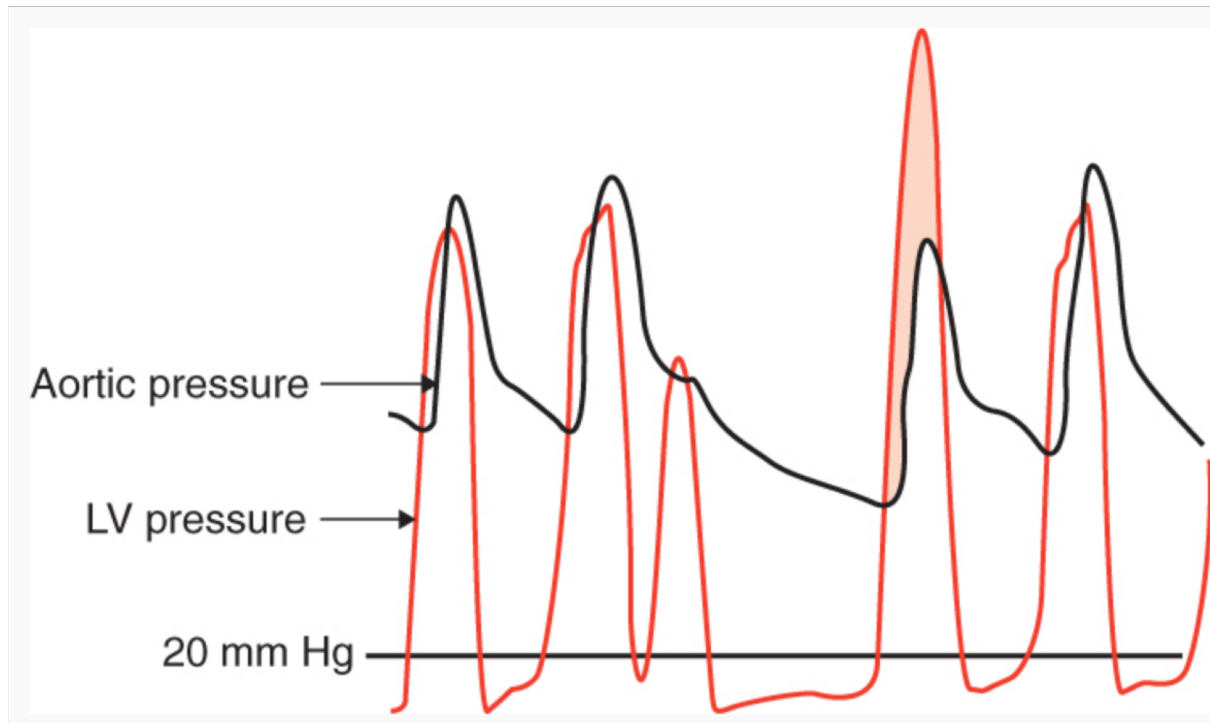


- Hypertroph obstruktive Kardiomyopathie
- Hypertensive Herzkrankheit
- Jegliche anderen Hypertrophieformen
- Lange Mitralklappensegel

- Valvuläre Aortenstenose
- Subvalvuläre Aortenstenose (Membran)

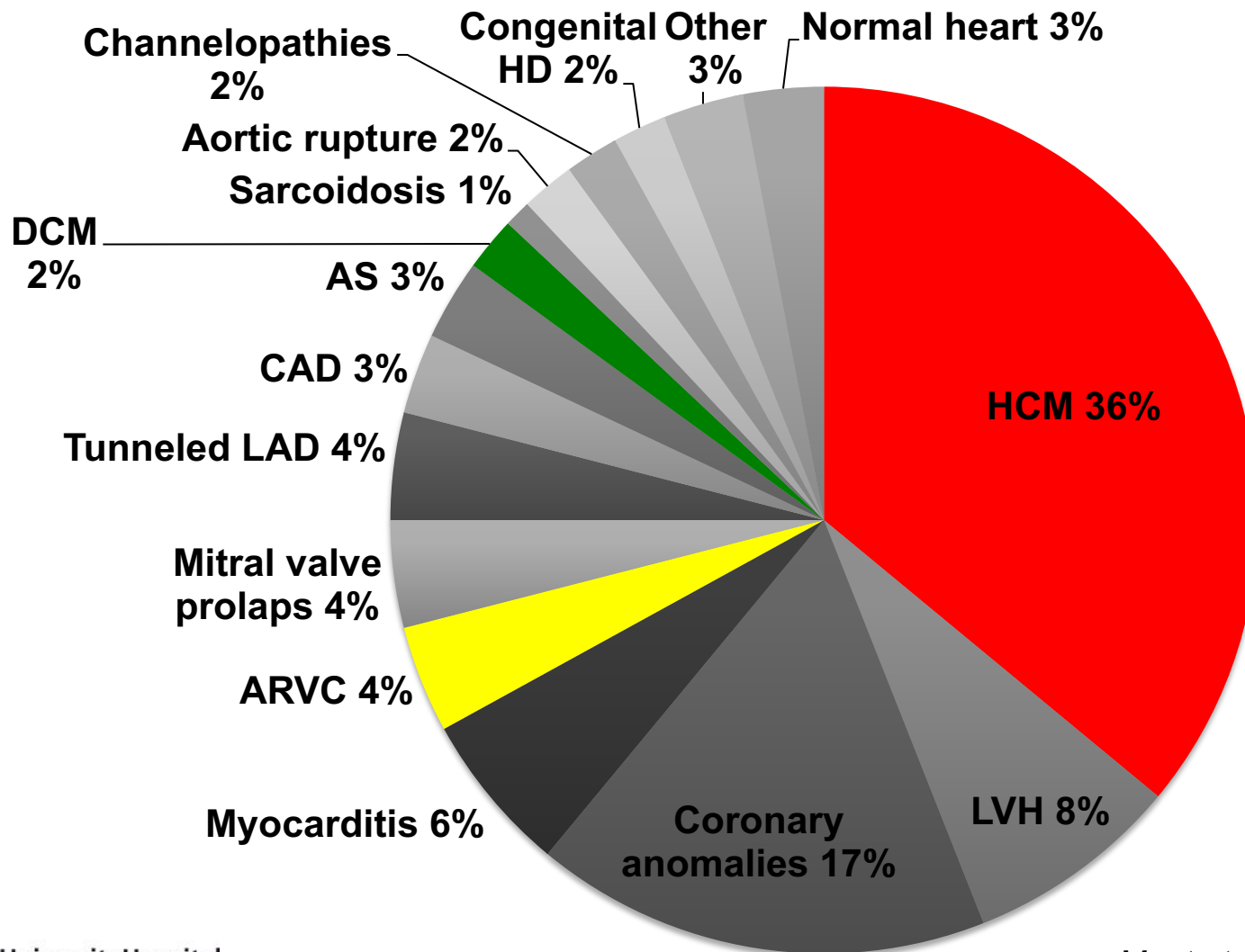
LVOT-OBSTRUKTION – PROVOKATIONSMANÖVER

- Valsalvamanöver
- Sitzen - Stehen
- Körperliche Anstrengung – Stressecho (physikalischer Stress)
- Post-extrasystolisch



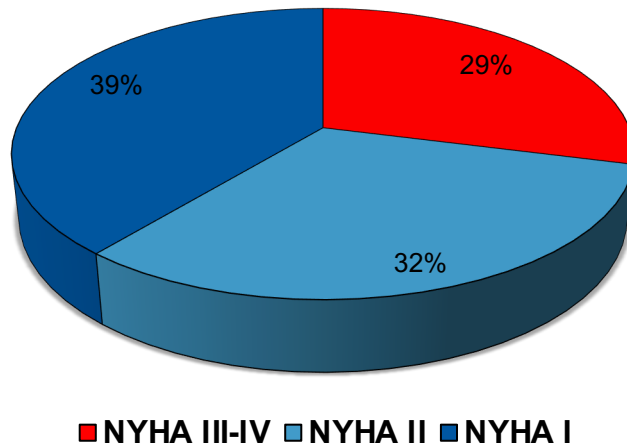
Brockenbrough-Morrow-Braunwald-Sign

KARDIOMYOPATHIEN / SCD BEI ATHLETEN

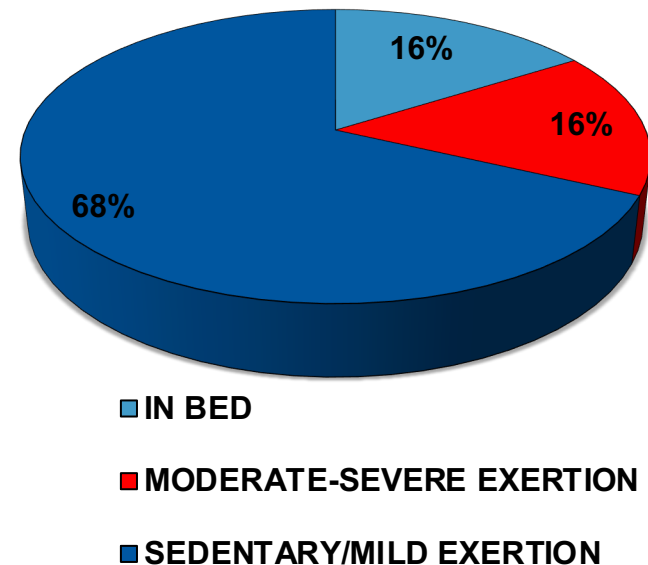


HYPERTROPHE KARDIOMYOPATHIE: WANN KOMMT DER PLÖTZLICHE HERZTOD?

SYMPTOMS



ACTIVITY DURING SCD



3 centers, n = 744, HCM-death 86/744 (12%), SCD 44/744 (6%), FU 8±7 years

HYPERTROPHE KARDIOMYOPATHIE: RISIKOSTRATIFIZIERUNG FÜR SCD

Indications For ICD

- **Aborted sudden death**
- **Sustained VT**

Major Risk Factors

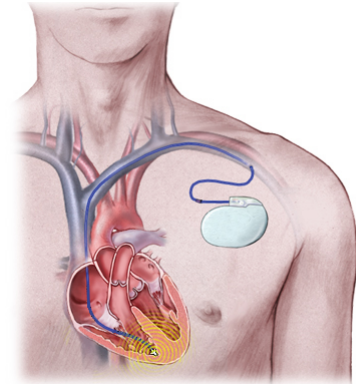
- **LV wall thickness 30mm or greater**
- **First degree family member SCD**
- **Recent unexplained syncope**

Minor Risk Factors

- **Abnormal BP response to exercise**
- **Non sustained VT on Holter**

Modifiers

- **CMR: LGE**
- **LVOT obstruction**
- **Apical LV aneurysm**
- **Genetic mutations (double and compound)**



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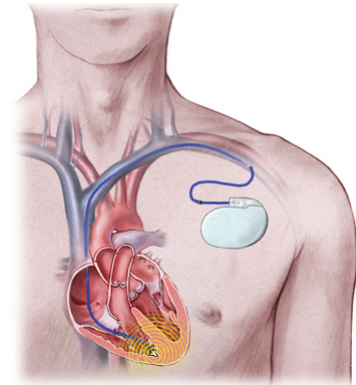
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HYPERTROPHE KARDIOMYOPATHIE: ESC RISK CALCULATOR

HCM Risk-SCD Calculator

Age	52	Years	<i>Age at evaluation</i>
Maximum LV wall thickness	20	mm	<i>Transthoracic Echocardiographic measurement</i>
Left atrial size	39	mm	<i>Left atrial diameter determined by M-Mode or 2D echocardiography in the parasternal long axis plane at time of evaluation</i>
Max LVOT gradient	21	mmHg	<i>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: $\text{Gradient} = 4V^2$, where V is the peak aortic outflow velocity</i>
Family History of SCD	☒ No	☐ Yes	<i>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).</i>
Non-sustained VT	☐ No	☒ Yes	<i>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.</i>
Unexplained syncope	☒ No	☐ Yes	<i>History of unexplained syncope at or prior to evaluation.</i>

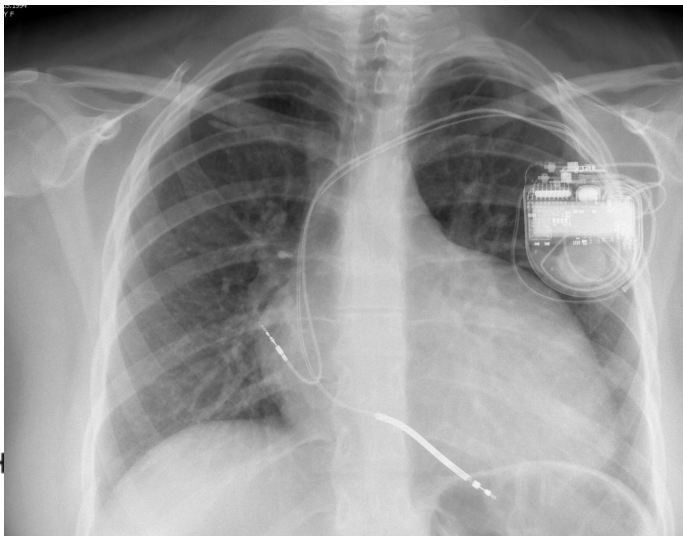
Risk of SCD at 5 years (%):

ESC recommendation:

*** ICD not recommended unless there other clinical features that are of potential prognostic importance and when the likely benefit is greater than the lifelong risk of complications and the impact of an ICD on lifestyle, socioeconomic status and psychological health.*

HYPERTROPHE KARDIOMYOPATHIE: VOR- UND NACHTEILE DES DEFIBRILLATORS

	Jährliche Rate	5-Jahres kumulative Inzidenz	Total (n=334)
Adäquate Schocks	2.3%	13%	28 (8%)
Primärprävention (n=307)	2.0%	12%	21 (7%)
Sekundärprävention (n=27)	4.3%	20%	7 (26%)
Inadäquate Schocks	4.6%	21%	55 (16%)
Implantationskomplikationen	5.1%	21%	60 (18%)



HYPERTROPHE KARDIOMYOPATHIE: VORHOFFLIMMERN

RHYTHMUSKONTROLLE >>>> FREQUENZKONTROLLE

▪ MEDIKAMENTE

- Betablocker, KEIN SOTALOL
- Kalziumkanalblocker (Verapamil-Typ)
- Amiodaron
- Keine Klasse 1C-Antiarrhythmika

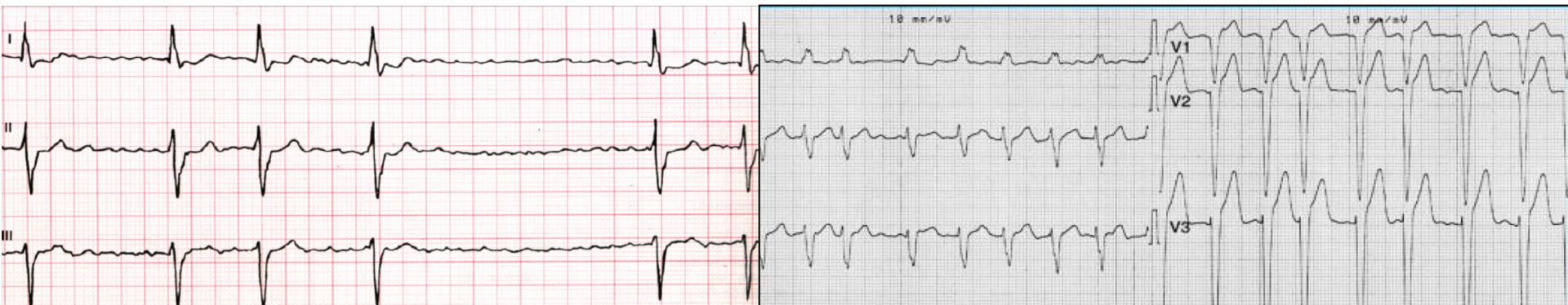
**KEINE 'EXOTISCHEN'
ANTI-ARRHYTHMIKA**

▪ ELEKTRISCHE KARDIOVERSION

▪ KATHETERTECHNISCHE LUNGENVENENISOLATION

▪ BEHANDLUNG DER LVOT-OBSTRUKTION (LA-DRUCK-REDUKTION)

**ORALE ANTIKOAGULATION UNABHÄNGIG VOM
CHA2DS2-VASc-Score !!!**



HYPERTROPHE KARDIOMYOPATHIE

MEDIKAMENTÖSE BEHANDLUNG DER LVOT-OBSTRUKTION

NEGATIV CHRONOTROP NEGATIV INOTROP

- **BETABLOCKER**
- **DISOPYRAMID**
- **(VERAPAMIL)**