Myocarditis and Cardiomyopathies

SHAHNAWAZ AMDANI M.D., F.A.C.C.

Pediatric Heart Failure and Transplant Cardiologist

Assistant Professor of Pediatrics, CCLCM

Cleveland Clinic Children's

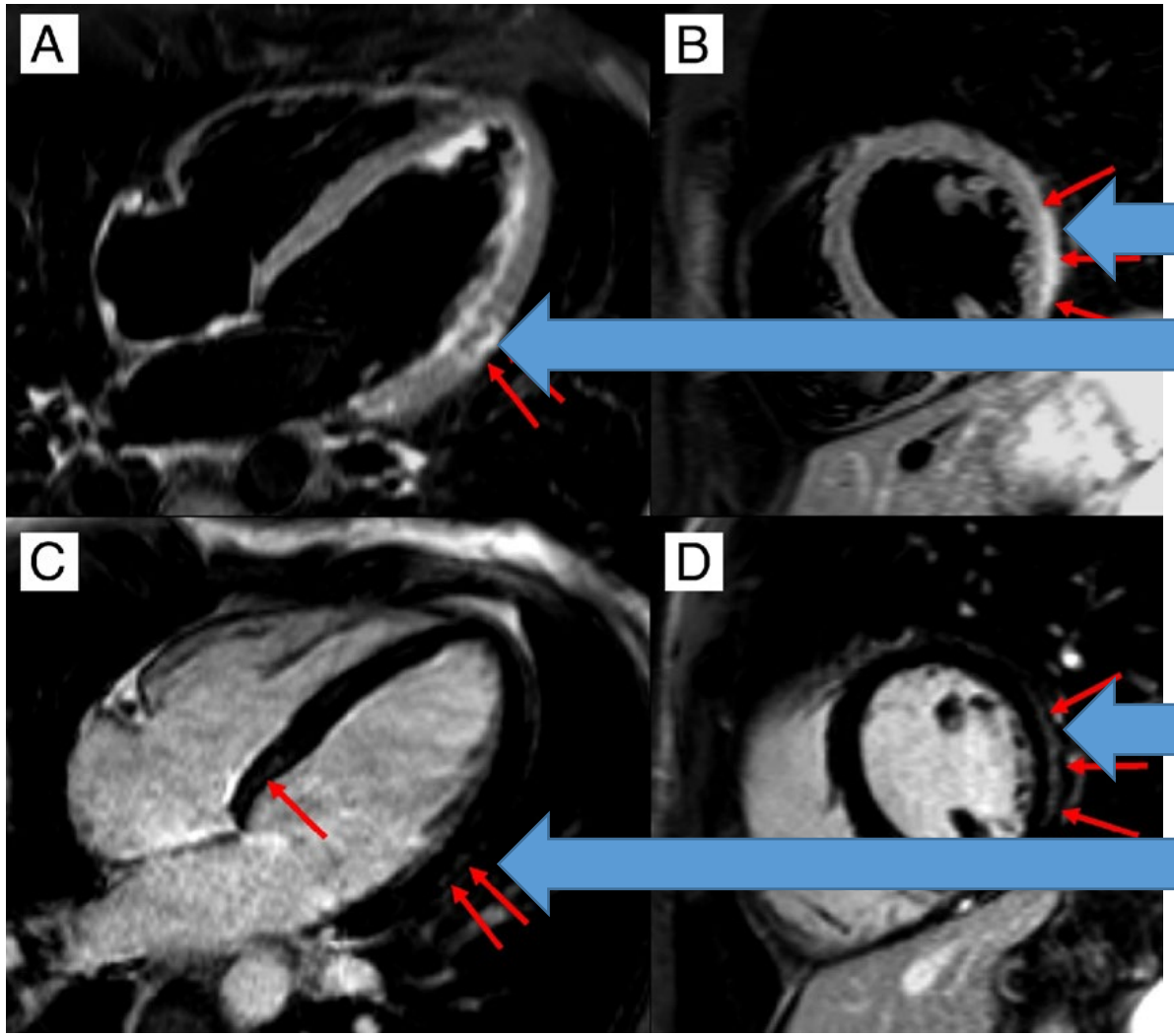
OBJECTIVES

- Discuss about rates of sudden cardiac death in myocarditis and various cardiomyopathies.
- Discuss risk factors for sudden cardiac death in pediatric patients with myocarditis and cardiomyopathies.

Myocarditis

- Inflammatory disease of the myocardium – cause can be infectious (PVB19 and HHV6) or non-infectious (celiac disease, Kawasaki, Crohn's disease, drugs)

Myocardial findings as seen on CMR

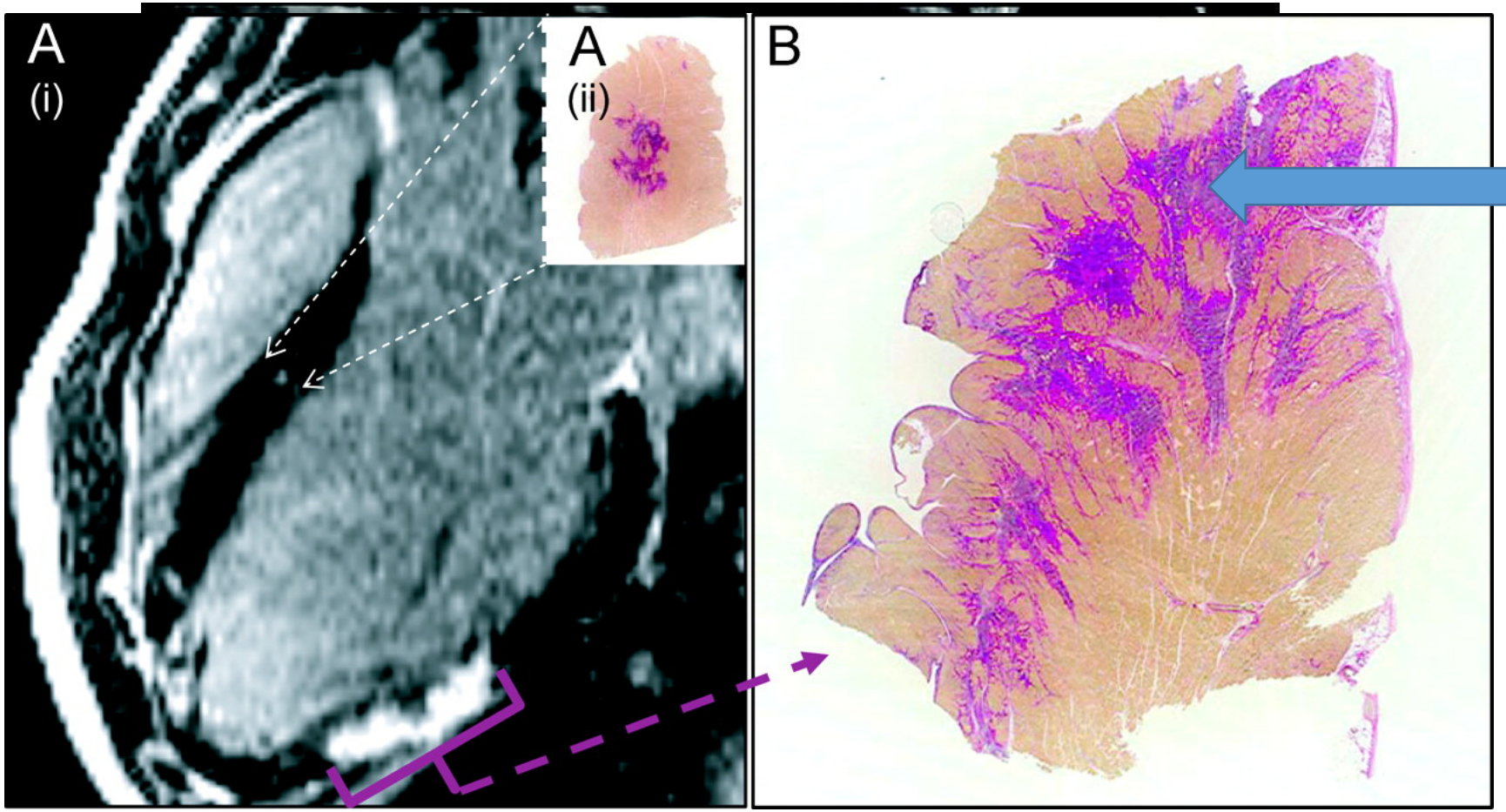


Myocardial edema

Myocardial fibrosis/ scarring

Kindermann I et al. Update on myocarditis. Journal of the American College of Cardiology. 2012 Feb 28;59(9):779-92.

Why is scarring important to identify?



Collagen – a.k.a Scar

Babu-Narayan SV et al. Myocarditis and sudden cardiac death in the young: extensive fibrosis suggested by cardiovascular magnetic resonance in vivo and confirmed post mortem. Circulation. 2007 Aug 7;116(6):e122-5.

Myocarditis increases risk for arrhythmias

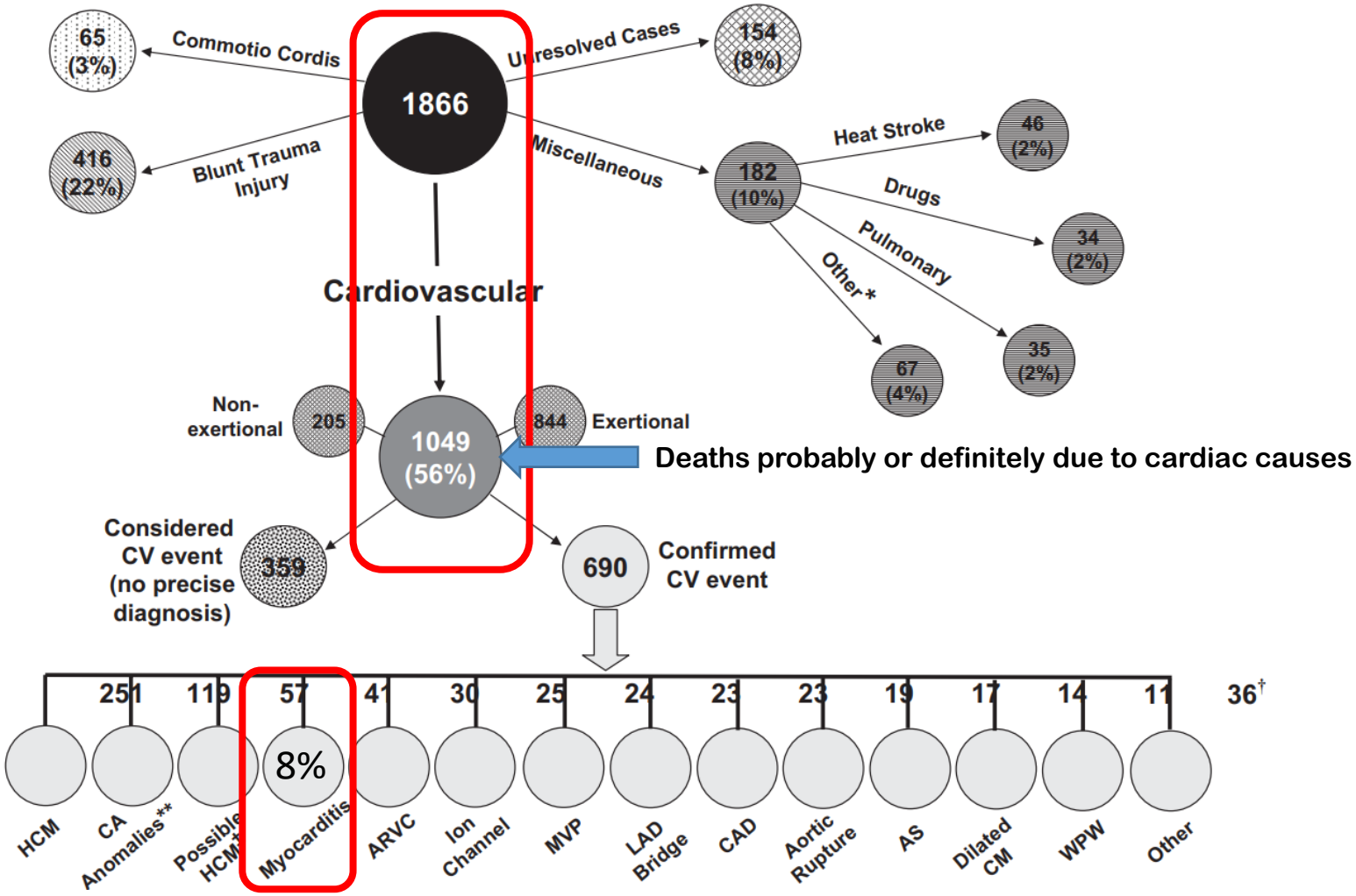
- Short-term : myocardial inflammation can lead to fluctuations in membrane potential and hence enhance arrhythmogenesis.
- Long-term: areas of scarring/fibrosis [areas of atrophy] and areas of secondary hypertrophy produces an arrhythmia substrate.

Maron BJ, et al.Sudden death in young competitive athletes: clinical, demographic, and pathological profiles. Jama. 1996 Jul 17;276(3):199-204.

Cardiovascular Abnormalities in 134 Young Competitive Athletes With Sudden Death*

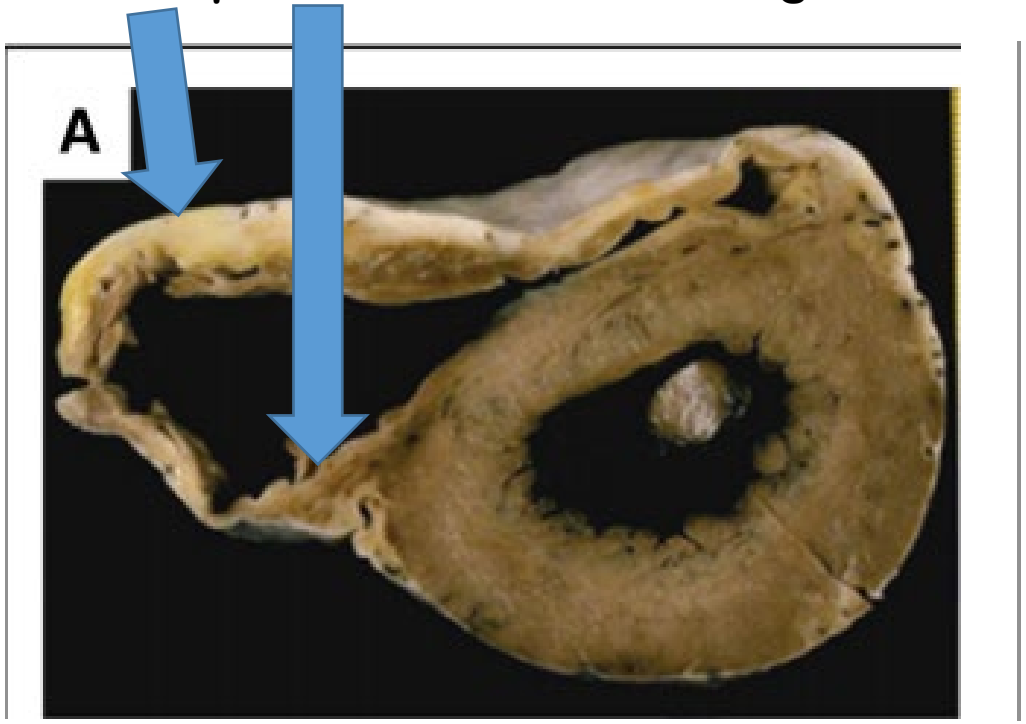
Primary Cardiovascular Lesion	No. (%) of Athletes	Median Age (Range), y
Hypertrophic cardiomyopathy	48 (36.0)	17.0 (13-28)
Unexplained increase in cardiac mass† ("possible hypertrophic cardiomyopathy")	14 (10.0)	17.0 (14-24)
Aberrant coronary arteries‡	17 (13.0)	15.0 (12-23)
Other coronary anomalies	8 (6.0)	17.5 (14-40)
Ruptured aortic aneurysm§	6 (5.0)	17.0 (16-31)
Tunneled LAD coronary artery	6 (5.0)	17.5 (14-20)
Aortic valve stenosis	5 (4.0)	14.0 (14-17)
Lesion consistent with myocarditis	4 (3.0)	15.5 (13-16)
Idiopathic dilated cardiomyopathy	4 (3.0)	18.0 (18-21)
ARVD	4 (3.0)	16.0 (15-17)
Idiopathic myocardial scarring	4 (3.0)	20.0 (14-27)
Mitral valve prolapse§	3 (2.0)	16.0 (15-23)
Atherosclerotic coronary artery disease	3 (2.0)	19.0 (14-28)
Other congenital heart diseases‖	2 (1.5)	13.5 (12-15)
Long QT syndrome¶	1 (0.5)	. . .
Sarcoidosis	1 (0.5)	. . .
Sickle cell trait#	1 (0.5)	. . .
"Normal" heart**	3 (2.0)	18.0 (16-21)

Maron BJ, et al. Sudden deaths in young competitive athletes. Circulation. 2009;119(8):1085-92.

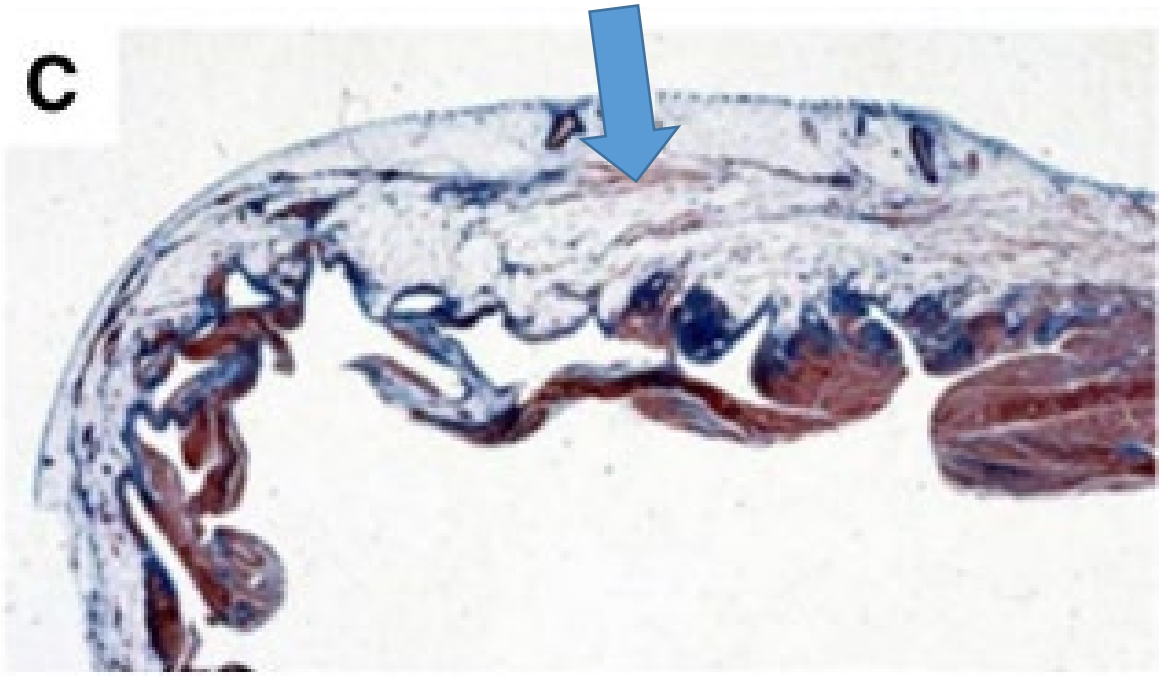


Arrhythmogenic Cardiomyopathy

Anterior and posterior RV wall thinning

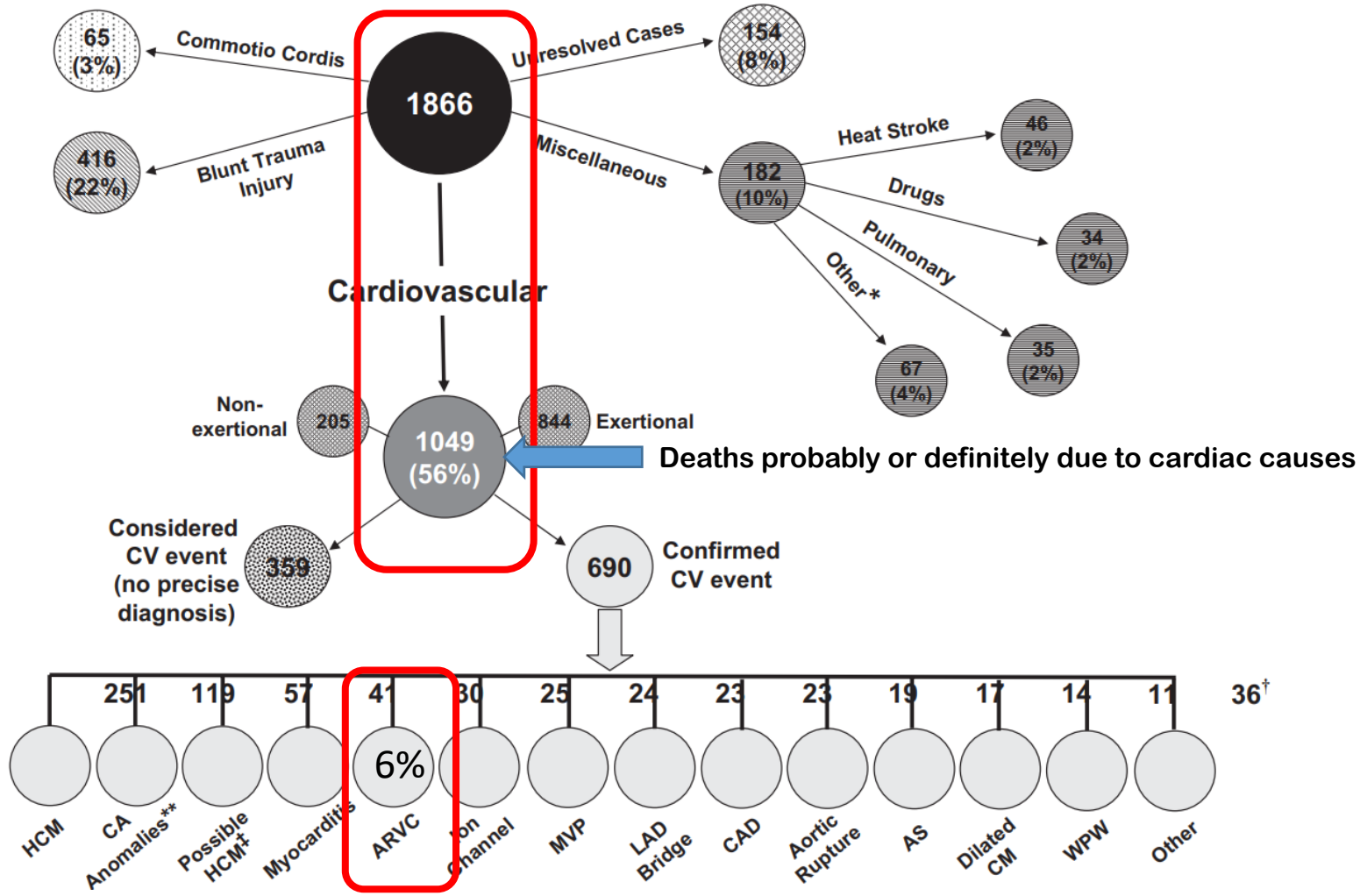


Fibrofatty replacement



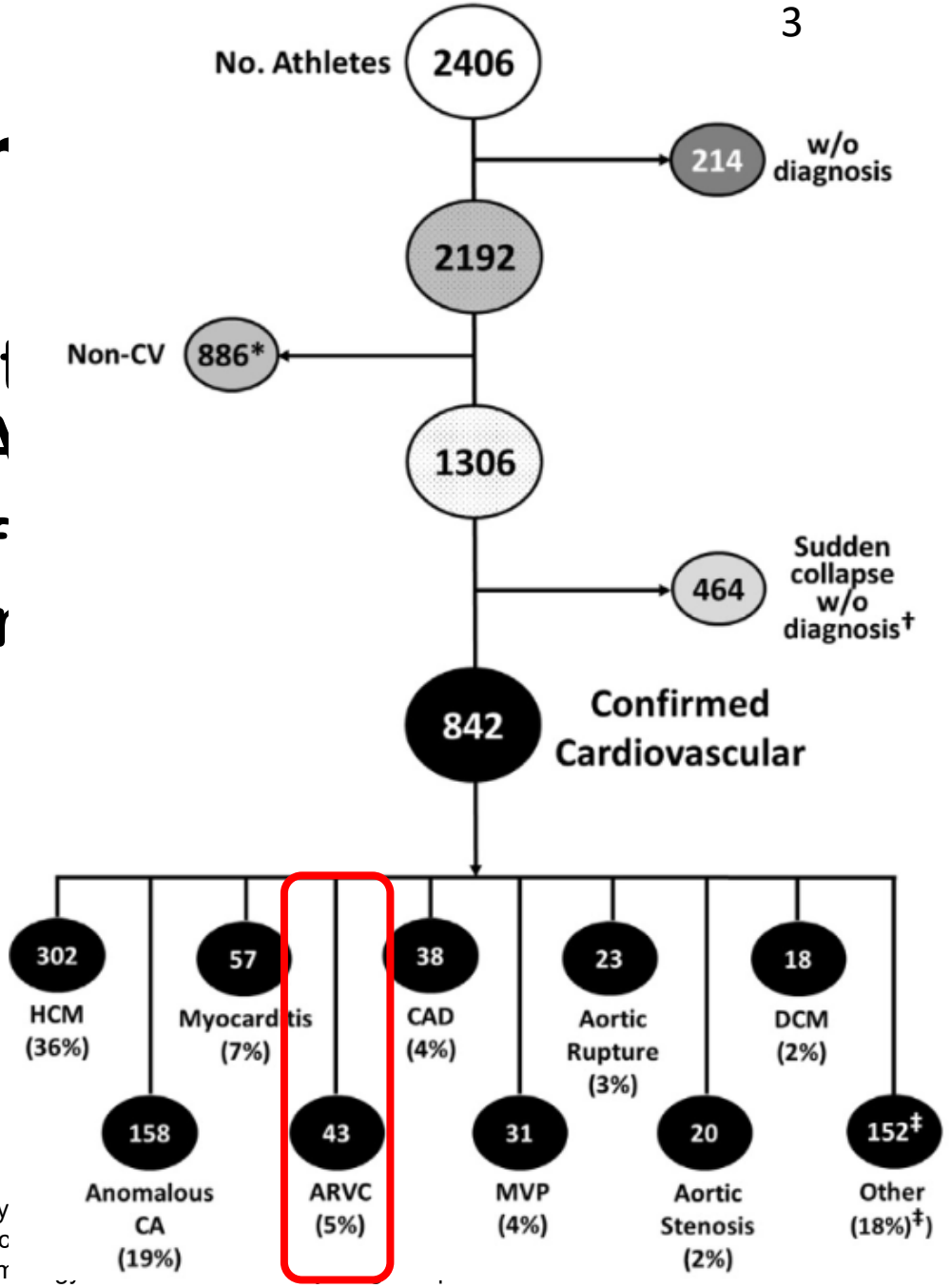
Corrado D, Basso C, Judge DP. Arrhythmogenic cardiomyopathy. Circulation research. 2017 Sep 15;121(7):784-802.

Maron BJ, et al. Sudden deaths in young competitive athletes. Circulation. 2009;119(8):1085-92.



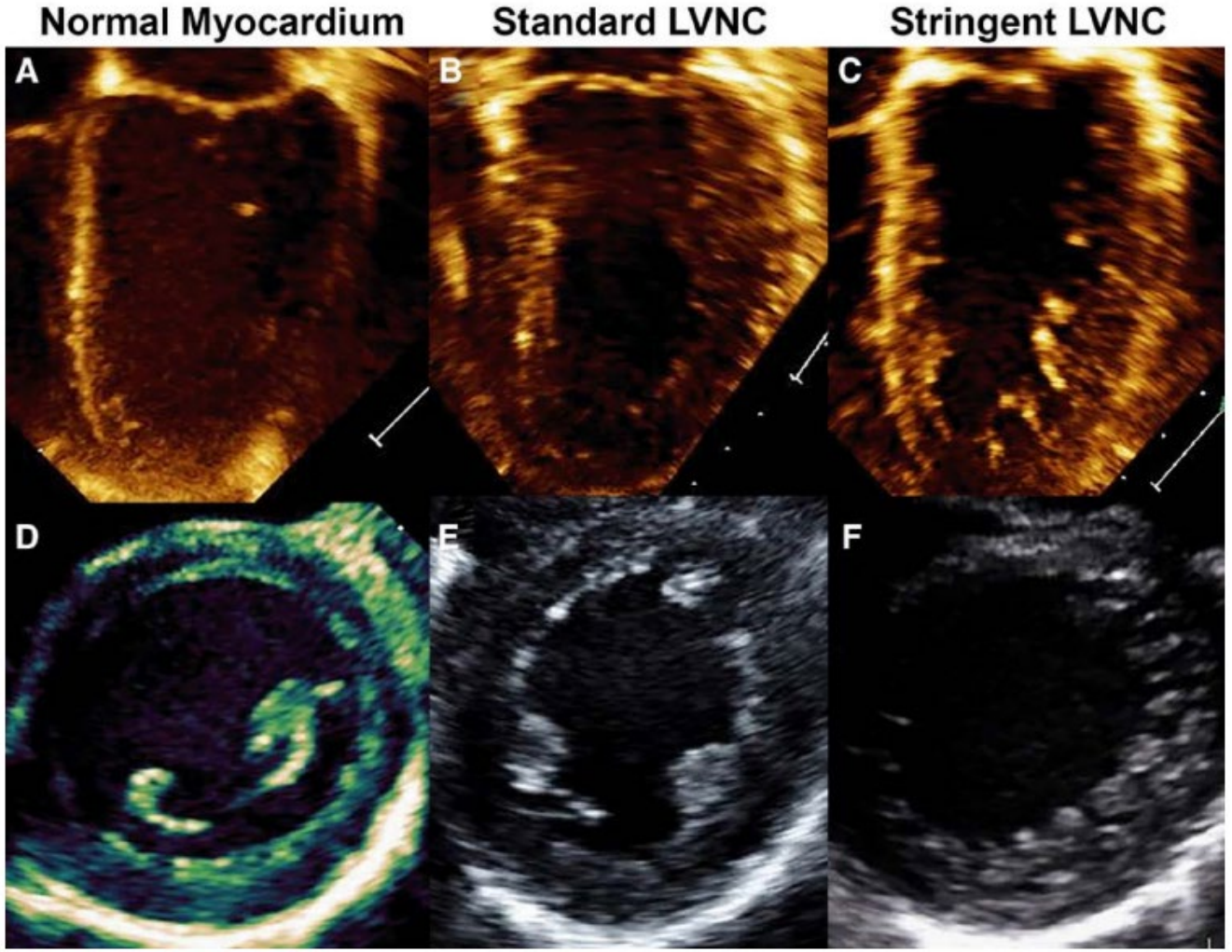
Arrhythr opathy

- In Veneto (I n examinations, 12 (20%) had A
- In the US of %) were diagnosed post-morter



1. Thiene G, et al. Right ventricular cardiomy .1056/NEJM198801213180301.
2. Dalal D, et al. Arrhythmogenic right ventric .1161/CIRCULATIONAHA.105.542266
3. Maron BJ,et al. Demographics and epiden al Registry. Am J Med. 2016;129:1170–1177. doi: 10.1016/j.amjmed.2016.02.031

Left ventricular non-compaction

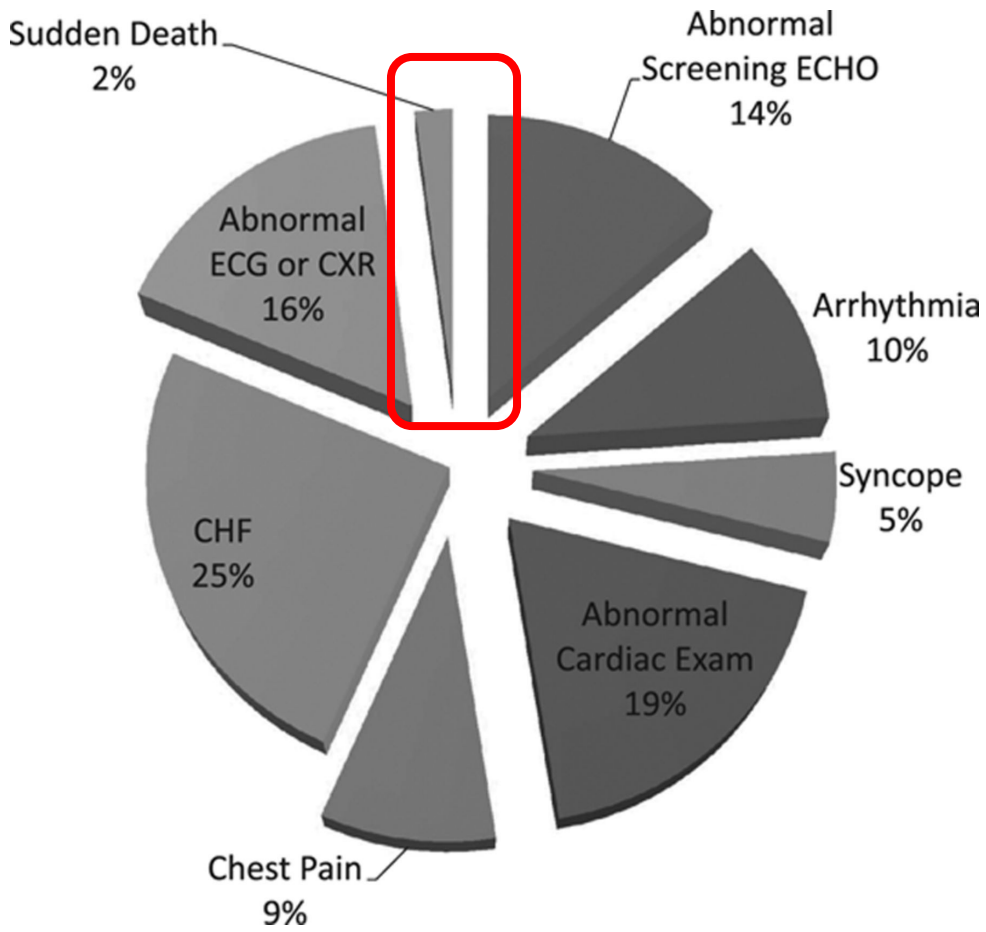


Miller EM, Hinton RB, Czosek R, Lorts A, Parrott A, Shikany AR, Ittenbach RF, Ware SM. Genetic testing in pediatric left ventricular noncompaction. Circulation: Cardiovascular Genetics. 2017 Dec;10(6):e001735.

Brescia ST, et al. Mortality and sudden death in pediatric left ventricular noncompaction in a tertiary referral center. Circulation. 2013 Jun 4;127(22):2202-8.

242 patients with LVNC over 18 years

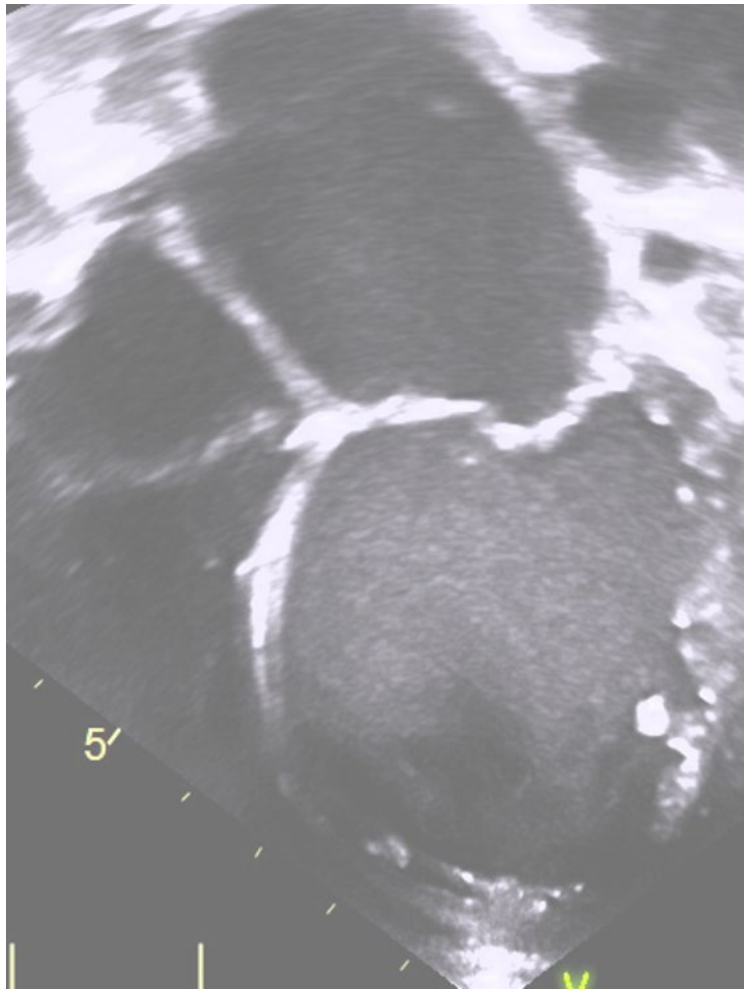
Clinical Characteristics at presentation



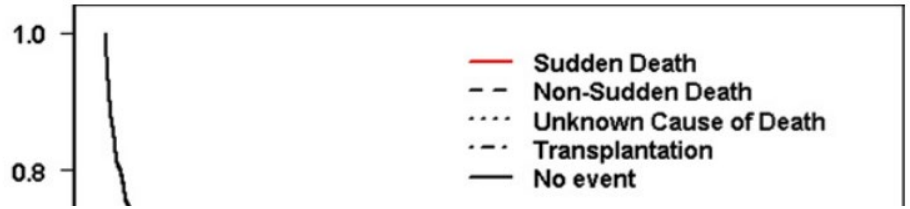
Sudden death in LVNC

- Of 242, 15 (6.2%) patients experienced sudden death over the study period.
- LV dilation, hypertrophy or both were identified in 14/15 patients.
- Systolic dysfunction was identified in 13/15.
- 9/15 had documented arrhythmia. (HR 7.6)
- **No patient with normal cardiac dimensions, function and without preceding arrhythmia died suddenly.**

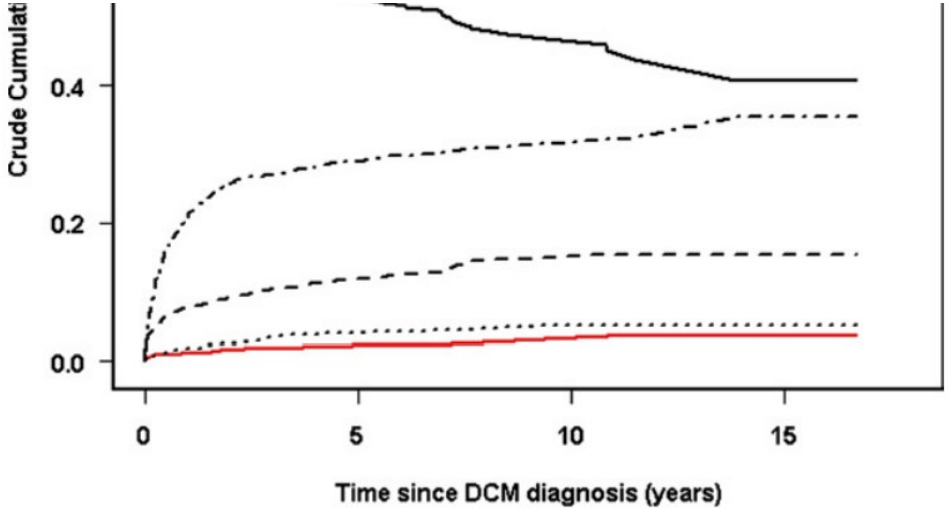
Dilated Cardiomyopathy



Pahl E, et al. Incidence of and risk factors for sudden cardiac death in children with dilated cardiomyopathy: a report from the Pediatric Cardiomyopathy Registry. JACC. 2012 Feb 7;59(6):607-15.



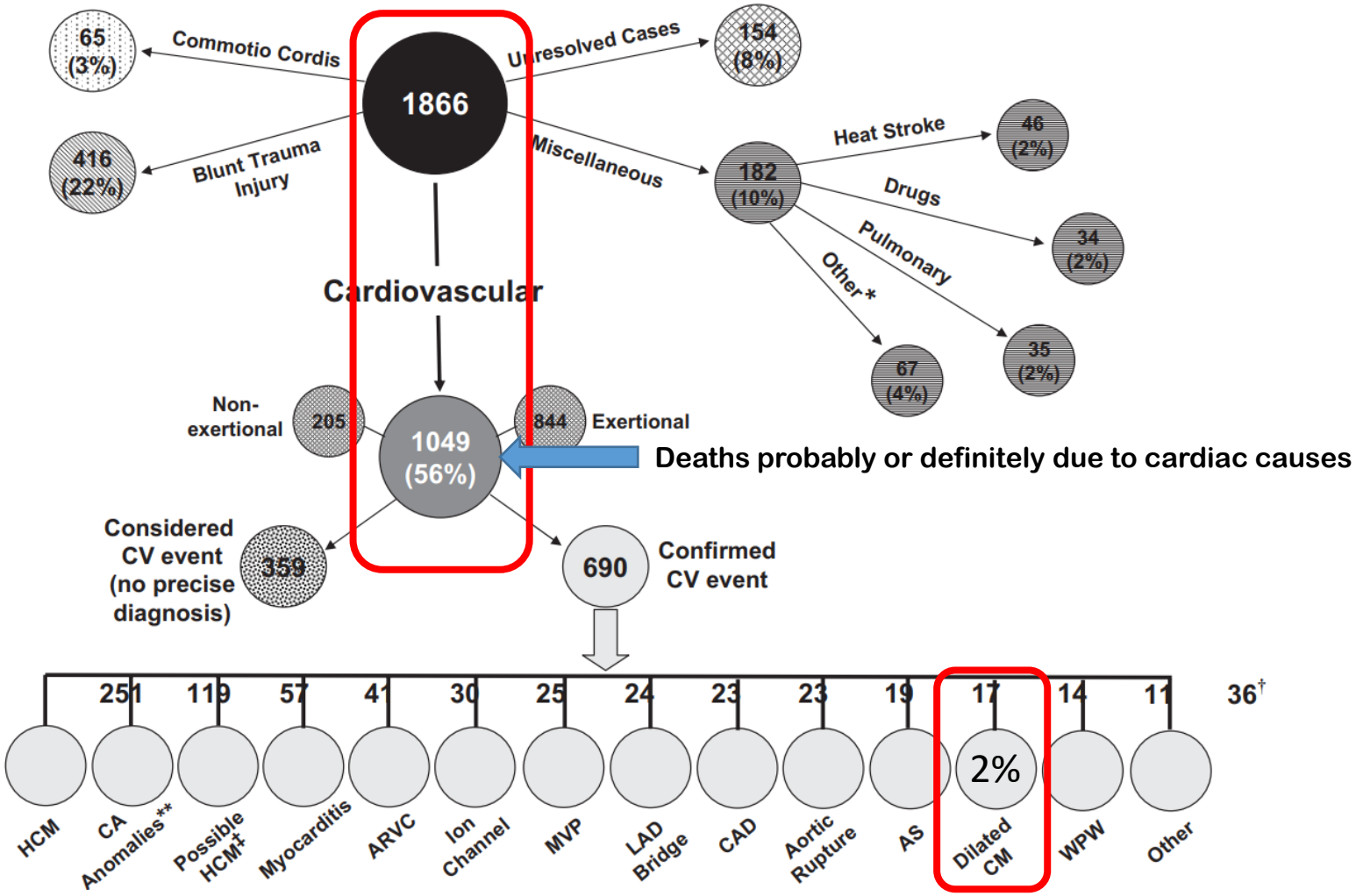
The 3-, 5-, and 10-year cumulative incidence rates (95% confidence interval) of sudden cardiac death are estimated to be **2.0%, 2.4%, and 2.7%** respectively



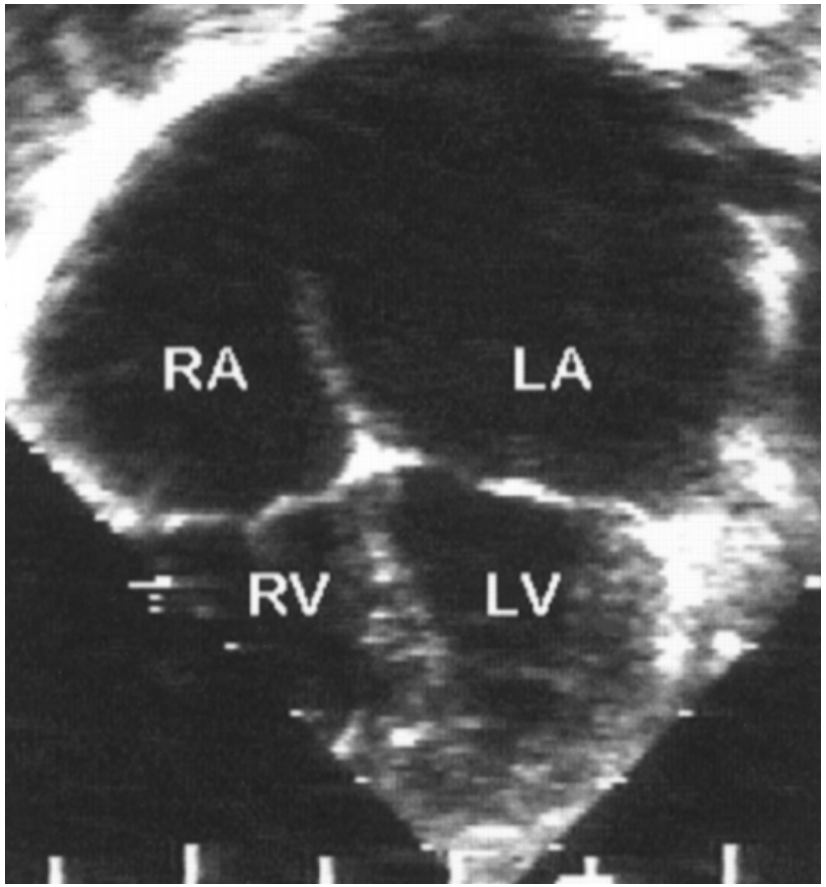
Risk factors for SCD in DCM patients

- CHF at diagnosis (HR 2.8)
- On anti-arrhythmics (HR 3.0)
- Thin LV posterior wall.

Maron BJ, et al. Sudden deaths in young competitive athletes. Circulation. 2009;119(8):1085-92.



Restrictive cardiomyopathies



Rivenes SM, et al. Sudden death and cardiovascular collapse in children with restrictive cardiomyopathy. Circulation. 2000 Aug 22;102(8):876-82.

Patient	Sex	Age at Diagnosis, y	Chief Complaint	Symptoms to Diagnosis, y	SCD	CHF	Outcome
1	F	10.8	Syncope	0	Yes	No	Death
2	F	4.9	Syncope	0	Yes	No	Death
3	F	1.6	Murmur	0	Yes	No	Death
4	F	8.9	Syncope	0	Yes	No	Death
5	F	5.3	CP	0.1	Yes	No	TX
6	M	5.0	CHF	NA	No	Yes	TX
7	F	0.9	CHF	0.8	No	Yes	TX
8	M	2.5	Murmur	0	No	No	TX
9	M	2.5	Family history	0	No	No	TX
10	M	0.9	CHF	0.1	No	Yes	TX
11	M	4.0	CHF	2.6	No	Yes	Death
12	F	1.7	CHF	0.1	No	Yes	Death
13	M	1.6	CHF	0.9	No	Yes	Death
14	F	4.1	CHF	0.9	No	Yes	Death
15	M	2.9	CHF	0.1	No	Yes	Death
16	F	1.1	CHF	0.1	No	Yes	Death
17	M	12.2	Irregular rhythm	0	No	Yes	Death
18*	M	5.0	CHF	NA	NA	NA	NA

28% patients had SCD

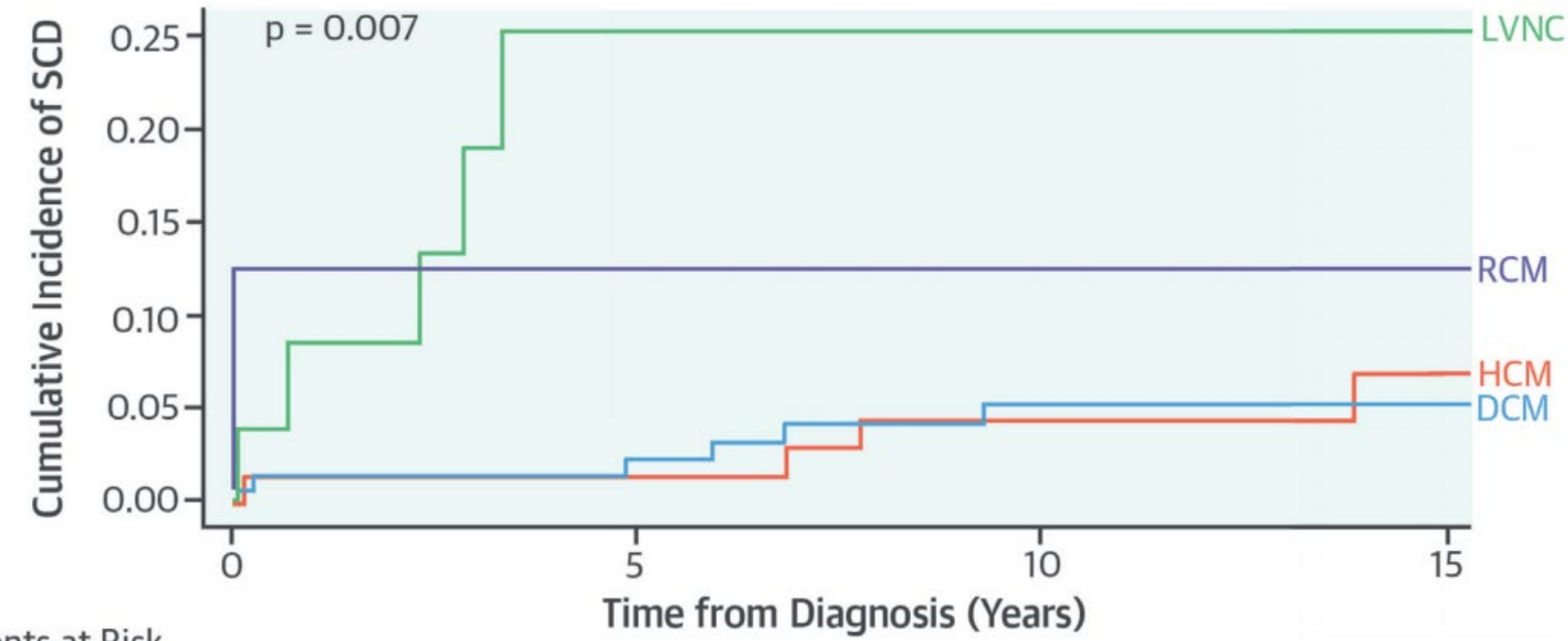
Rivenes SM, et al. Sudden death and cardiovascular collapse in children with restrictive cardiomyopathy. Circulation. 2000 Aug 22;102(8):876-82.

SCD in children with CMP from Australia

TABLE 1 Demographics of Study Population

	DCM (n = 172)	HCM (n = 80)	LVNC (n = 29)	RCM (n = 8)	All Patients (N = 289)
Age at diagnosis, yrs	0.65 (0.16-1.69)	0.45 (0.12-2.48)	0.34 (0.10-1.33)	3.03 (2.59-5.44)	0.56 (0.15-1.91)
Follow-up, yrs	12.5 (0.6-15.9)	14.0 (10.7-17.1)	6.8 (7.0-14.1)	2.8 (1.5-7.2)	11.9 (1.7-15.4)
Follow-up for survivors, yrs*	15.1 (13.0-17.3)	15.7 (12.9-17.7)	14.3 (12.2-14.7)	N/A (n = 1)	14.2 (12.5-17.4)
Positive family history of CM	26 (13.4)	19 (23.8)	9 (3.4)	1 (12.5)	55 (19.0)
Outcome					
SCD	6 **3.5%**	4 **5%**	5 **17.2%**	1 **12.5%**	16
HFD/transplant	62	13	11	6	92
Survivors	104	63	13	1	181

CENTRAL ILLUSTRATION **Sudden Death in Childhood Cardiomyopathy: Cumulative Incidence by Cardiomyopathy Phenotype**



Patients at Risk

LVNC	29	15	13	3
RCM	8	3	1	1
HCM	80	67	63	35
DCM	172	106	91	42

Bharucha, T. et al. J Am Coll Cardiol. 2015; 65(21):2302-10.

Conclusion

- Patients with myocarditis have a 8% risk for SCD. This risk is especially higher in early phase because of myocardial inflammation and is probably higher in the late phase in those with persistent systolic dysfunction and evidence of scarring/fibrosis.
- Children with CMP (other than HCM) have varying rates of SCD.
- It appears the highest risk is for patients with ARVC – LVNC --RCM and then DCM.

Contd.

- CMP patients with systolic dysfunction, evidence of arrhythmia (either on ambulatory monitoring or stress testing), and dilated LVs are at an increased risk for a sudden cardiac event.


THANK
YOU
FOR
YOUR
ATTENTION