



JOHNS HOPKINS  
M E D I C I N E

# Cardiomyopathy and Sports:

## Who can play?

5 February, 2017

Jane Crosson, MD

**I.**

I have received (a)  
research grant(s) /  
in kind support

**A**

... from current  
sponsor(s)

**YES**
☐
**NO**
☒
**B**

... from any institution

**YES**
☐
**NO**
☒
**II.**

I have been a speaker  
or participant in  
accredited CME/CPD ...

**A**

... from current  
sponsor(s)

**YES**
☐
**NO**
☒
**B**

... from any institution

**YES**
☒
**NO**
☐
**III.**

I have been a  
consultant / strategic  
advisor etc. ...

**A**

... for current  
sponsor(s)

**YES**
☐
**NO**
☒
**B**

... for any institution

**YES**
☐
**NO**
☒
**IV.**

I am a holder of  
(a) patent / shares /  
stocks or ownership...

**A**

... related to  
presentation

**YES**
☐
**NO**
☒
**B**

... not related to  
presentation

**YES**
☐
**NO**
☒

# SCORE: 1

# The diseases and the problem

- **Hypertrophic cardiomyopathy (HCM)**
  - most prevalent cause of sudden death (SCD) in athletes (US registry 36%, lower in Italian)
  - Risk of SCD  $\sim 0.5\%$ /year overall
- **Arrhythmogenic right ventricular cardiomyopathy (ARVC)**
  - Risk of SCD  $\sim 1\%$ /year overall
- **Dilated CM and non-compaction**
  - Limited data, no further discussion

# Current published guidelines

- **Both American Heart Association (2015) and European Society of Cardiology (2005) advocate that cardiomyopathy patients be restricted from almost all competitive sports**
- **Differ in treatment of genotype positive, phenotype negative patients**
- **Consensus based, little hard data**

# Ultimate Goal: Avoiding this...





# While allowing this in as many children as possible



# What is our goal?

- **Goal:** prevent every sports related death in patients
  - Solution: Draconian restriction of all patients
- **Goal:** maximize the lives of our patients
  - Solution: Assess individual risk, work with patient and family to customize best plan

# Benefits of exercise

- Widely known to decrease all cause mortality over a lifetime
- Reduces obesity and Type 2 diabetes
- Improves functional capacity in heart failure patients
- Mouse HCM model suggests physical activity decreases myocyte disarray



# Hypertrophic cardiomyopathy

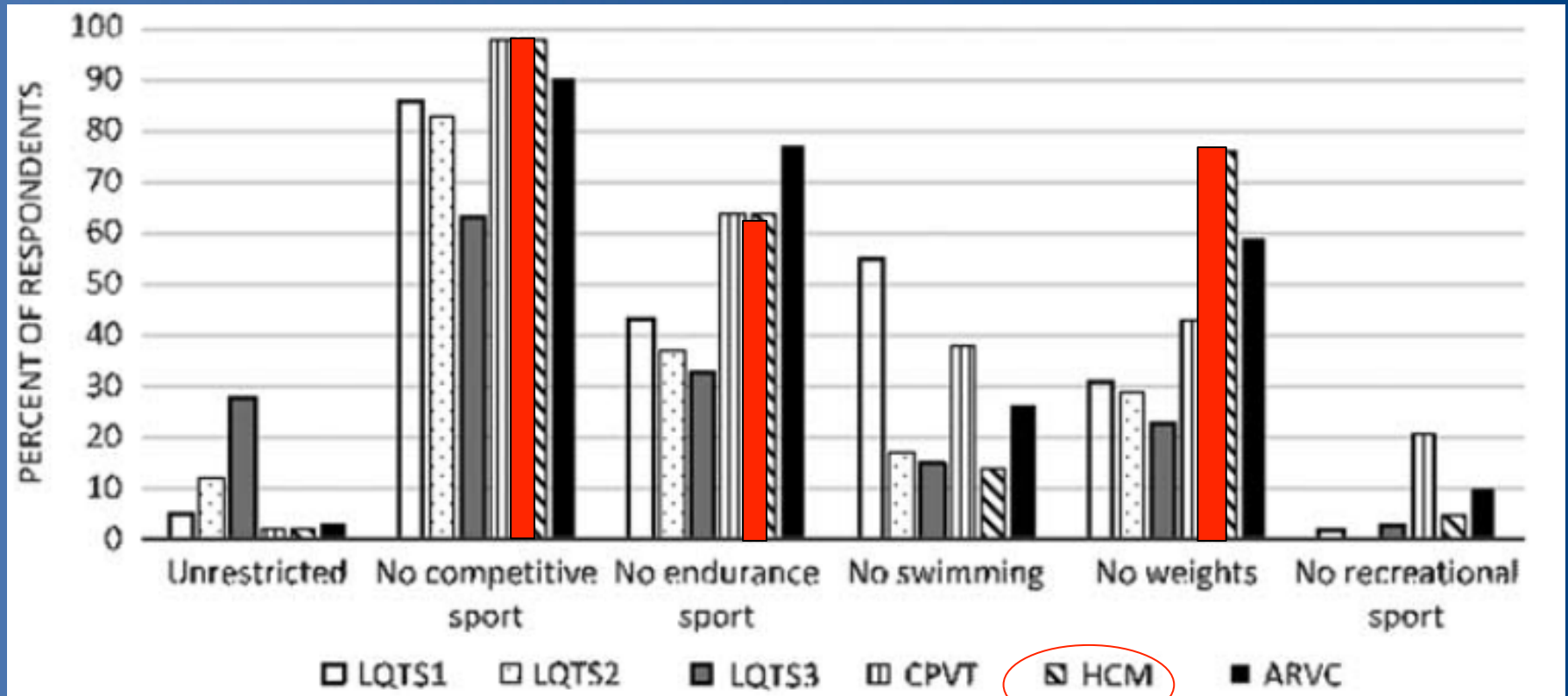
- Very heterogeneous group:
  - many will never develop symptoms, have very low risk of SCD
- SCD risk increased by:
  - family hx of SCD, septum >30 mm, unexplained syncope, NSVT, apical aneurysm, delayed enhancement
- Theoretical evidence of increased risk of disease progression with sports, other data that it actually improves outcome

- Also wide variability in penetrance of disease
- 6-fold higher incidence of events with exercise than at rest
- Strong data for disease progression linked to high endurance/intensity exercise

Are physicians following the  
guidelines?

Survey of PACES members:

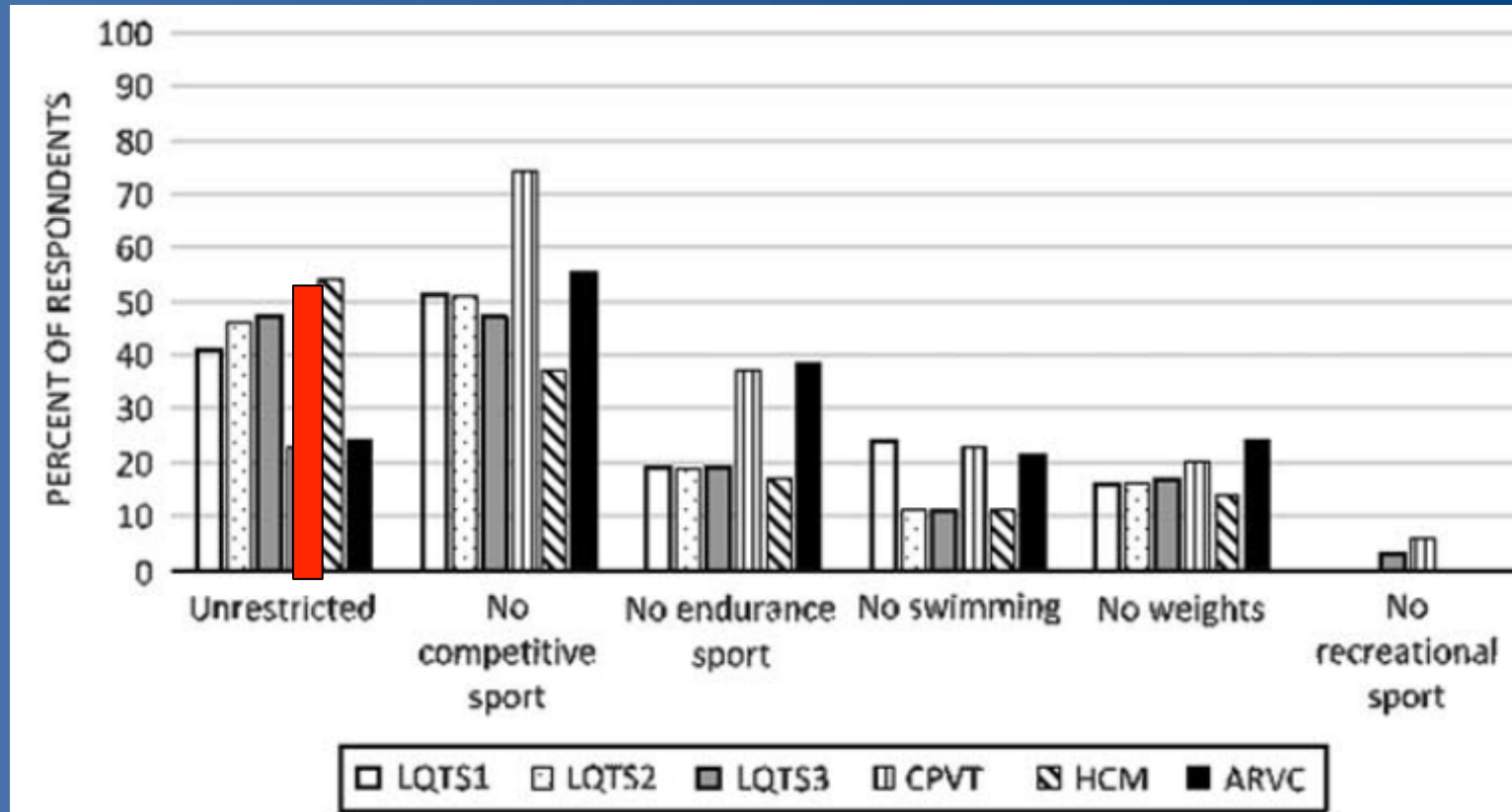
# What pediatric EPs are currently recommending for HCM patients



*Almost all restrict from competitive sports*

Christian, Cardiol Young 2016

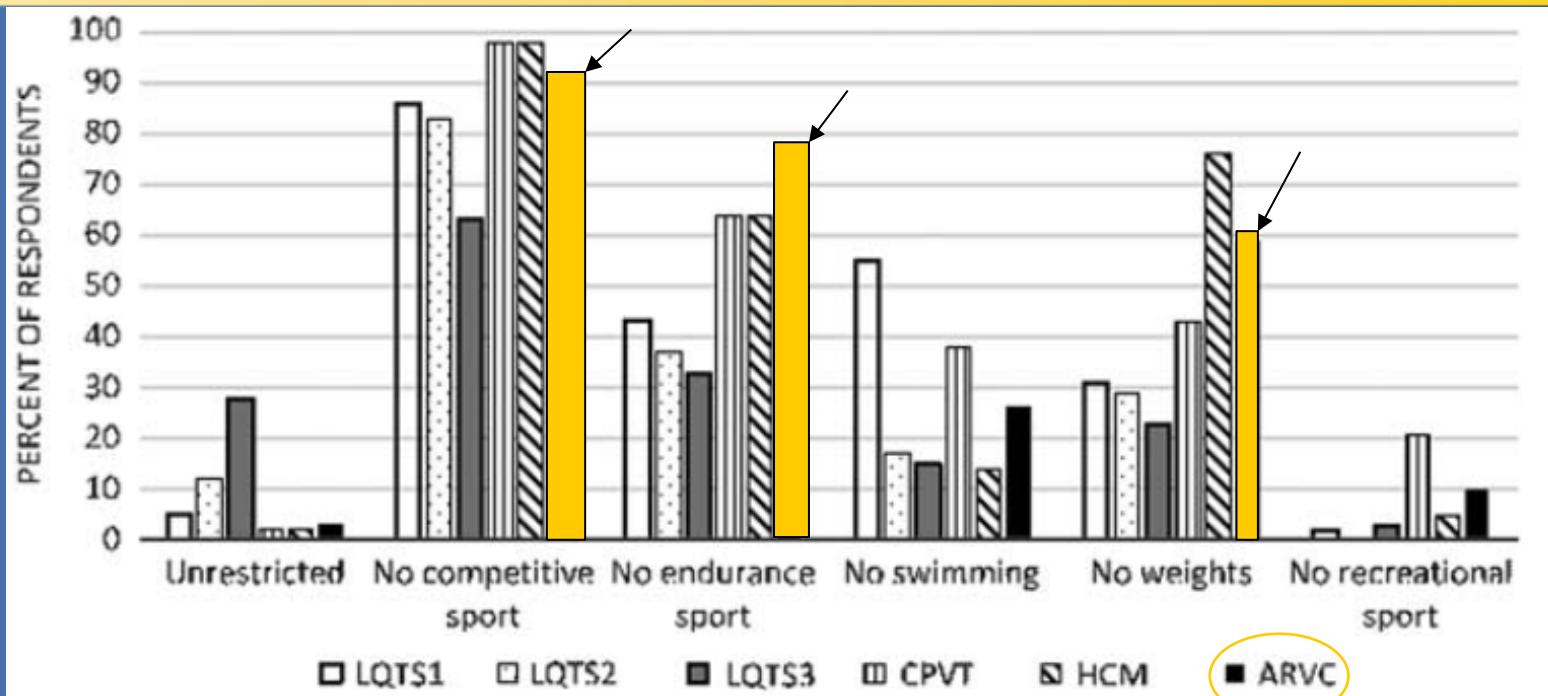
# What about for genotype-positive only HCM patients?



*~50% restrict these patients*

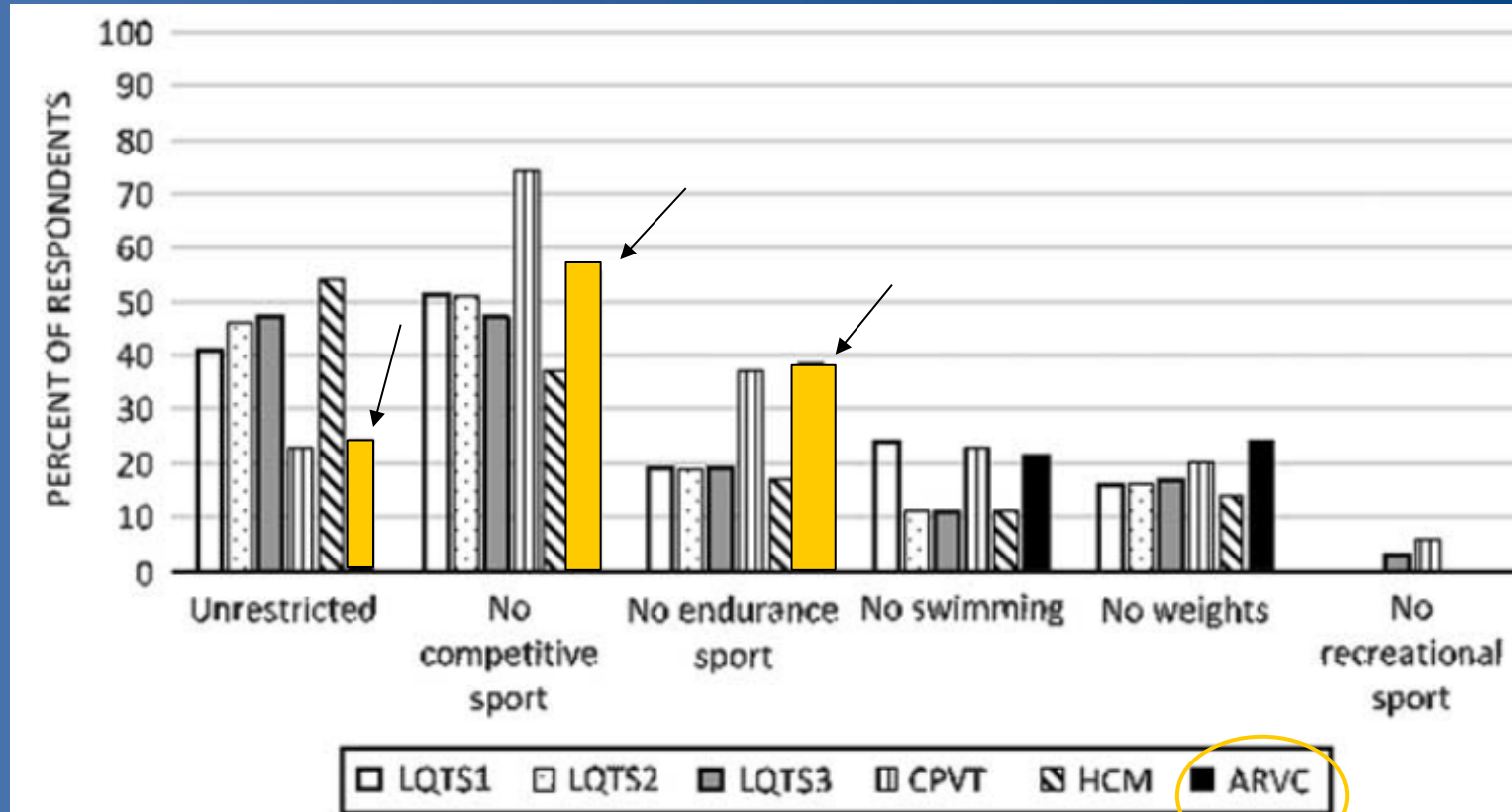


# What are pediatric EPs currently recommending for ARVC patients?



*For phenotype positive pts, 90% recommend restriction from competitive sports, 75% from endurance sports*

# Restrictions drop dramatically for phenotype negative ARVC pts



*Over 20% give no restrictions, <40% restrict from endurance sports*

# ARVC:

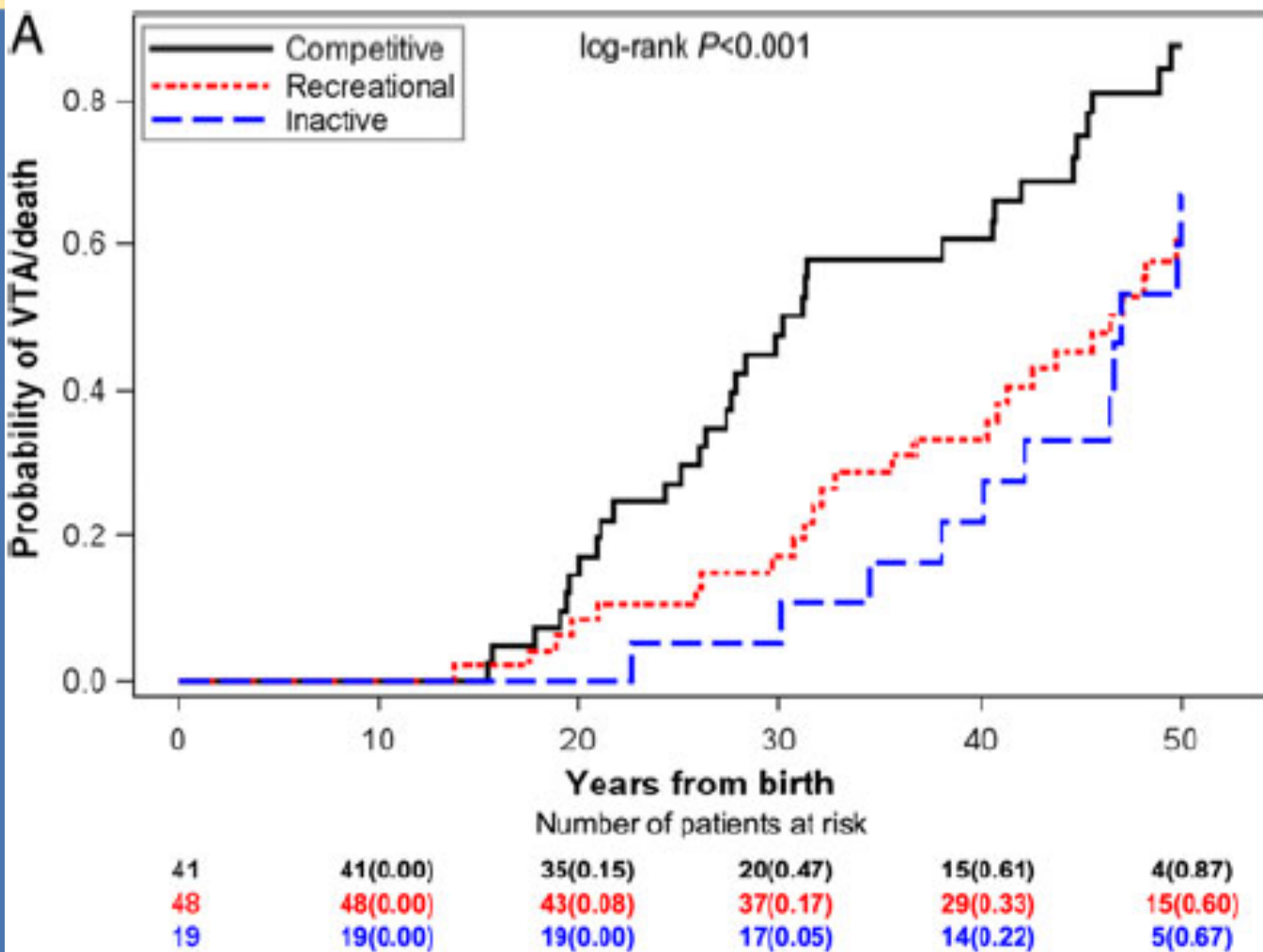
## Why Exercise Restriction is Important



- Progression of disease correlates with high intensity athletics
- Endurance athletes with ARVC have an increased risk of ventricular arrhythmias, heart failure, and MRI abnormalities
- Mouse model shows dramatic progression of RV enlargement with endurance activity



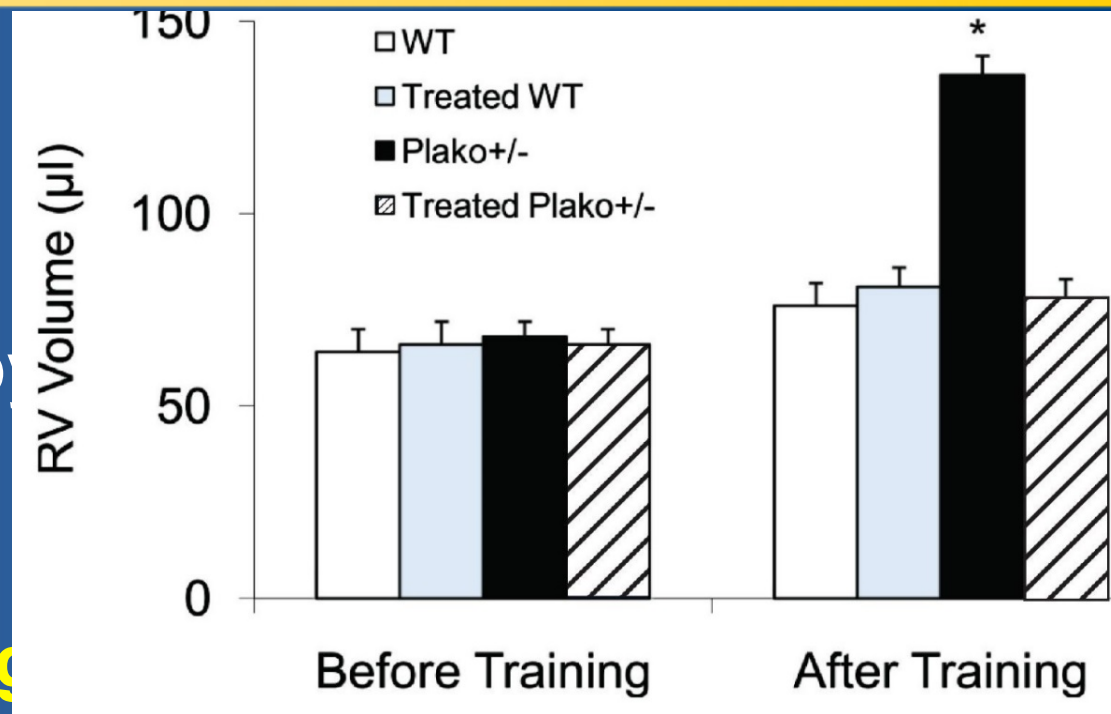
# Impact of competitive sports on outcome in ARVC



In competitive athletes, symptoms developed at an earlier age, risk of VT/sudden death was twice as high

# Load-reducing therapy prevents ARVC expression in plakoglobin-deficient mice

- Heterozygous plakoglobin-deficient mice vs wild type
- Load-reducing therapy
  - Furosemide
  - Nitrates
- **Six weeks swimming = marked increase in RV volume**, blunted by load-reducing





# How safe are sports with an ICD?

- 372 athletes with ICDs, primary and secondary prevention, ages 10-60
  - 10% had shocks during sport, 8% during other physical activity, 6% at rest
  - No ICD complications, resuscitated cardiac arrest or death related to physical activity
  - Freedom from lead malfunction 97% at 5 years
- Larger study of HCM athletes now in progress (Live-HCM)

# What is a reasonable course?

- Customize for each individual patient, using
  - risk factors
  - patient desire to play & type of activity
  - availability of AEDs
  - disease substrate
- No data to support restriction of truly genotype + only HCM, but there is good evidence for restriction of ARVC genotype +

*Summary:*

## Recommendations for Exercise Restrictions

- The pendulum has swung to somewhat decreased restrictions for children with cardiomyopathy
- Positive move for some HCM patients
- But for genotype positive ARVC patients:
  - **No competitive athletics requiring endurance**
  - Recreational play not restricted
- For phenotype positive patients, may further restrict, but encourage some physical activity