ACTIVITY RESTRICTIONS IN PEDIATRIC CARDIOVASCULAR DISEASE

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Children's Hospital of Philadelphia

DISCLOSURES

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- Consultant: Abbott Technologies, AbioMed Incorporated

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GOALS

- Where do things stand?
- What do the guidelines say?
- How do we get where we need to go?

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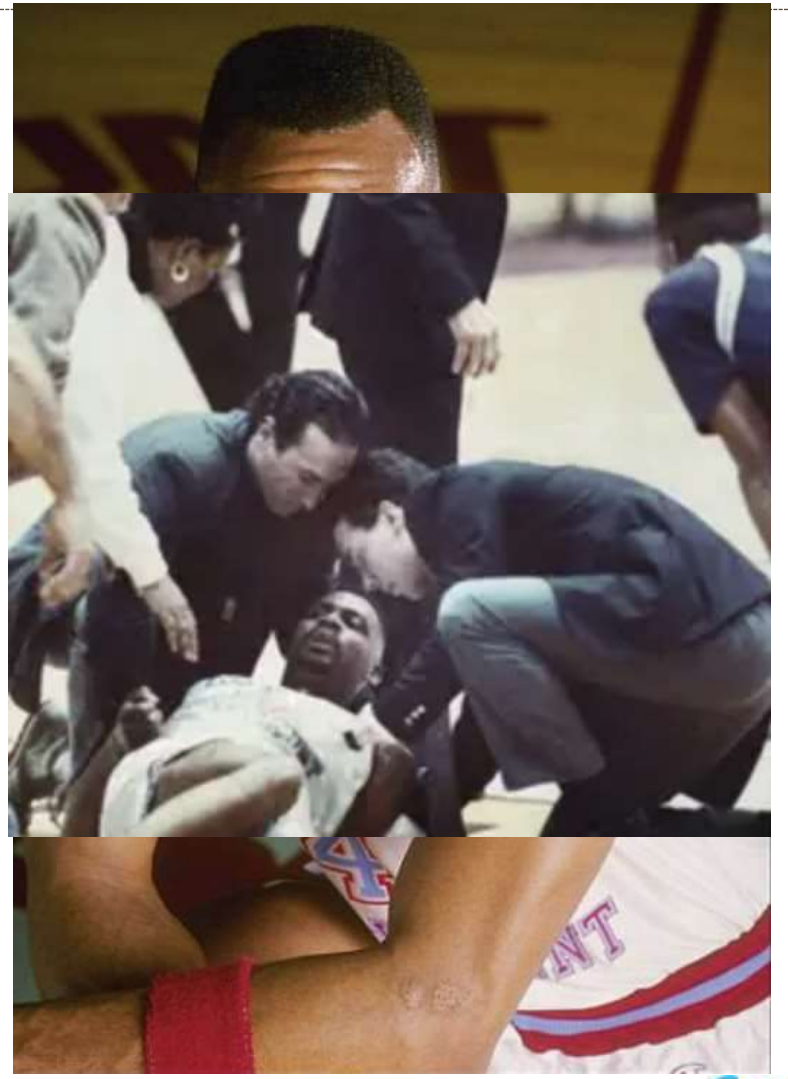
HYPERTROPHIC CARDIOMYOPATHY

Exercise: a 4 letter word

- Risk of sudden Death
  - LVOTO→ Coronary Malperfusion
  - Catecholamine Surge→ Primary Arrythmia
- Worsening of Fibrosis, promotion of LVH?



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HYPERTROPHIC CARDIOMYOPATHY

Low Intensity Activities

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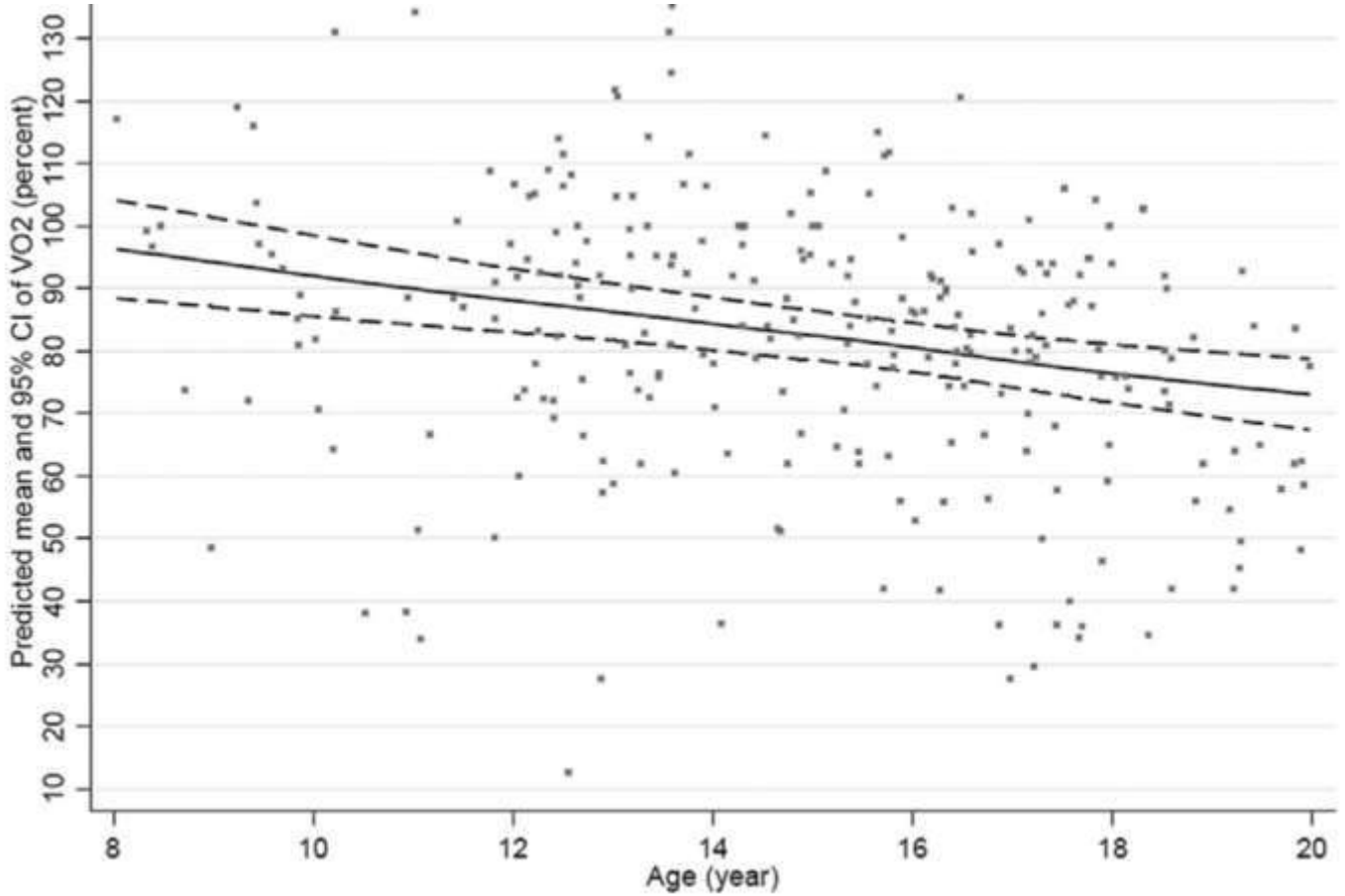
RESTRICTION COMES WITH A COST: IMPAIRED FITNESS

140 patients with HCM (<21 yo)

Low levels of cardiorespiratory fitness→**81% predicted**

<u>Fitness continues to get worse as patients get older</u>



Edelson et al. JACC: Advances 2022

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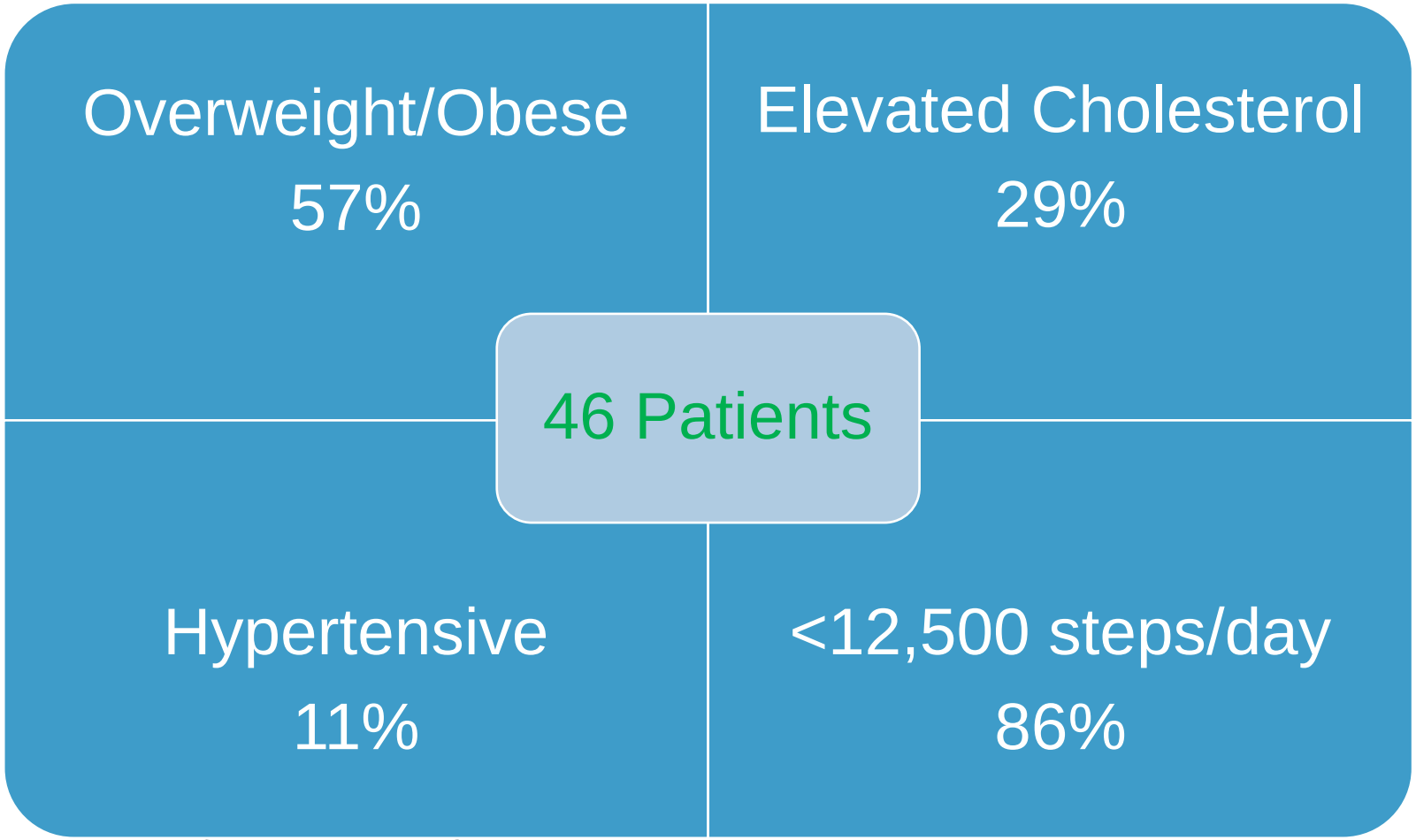
RESTRICTION COMES WITH A COST: INCREASED CV RISK



ASPIRE-HCM

Conway et al. AHA Scientific Sessions 2022

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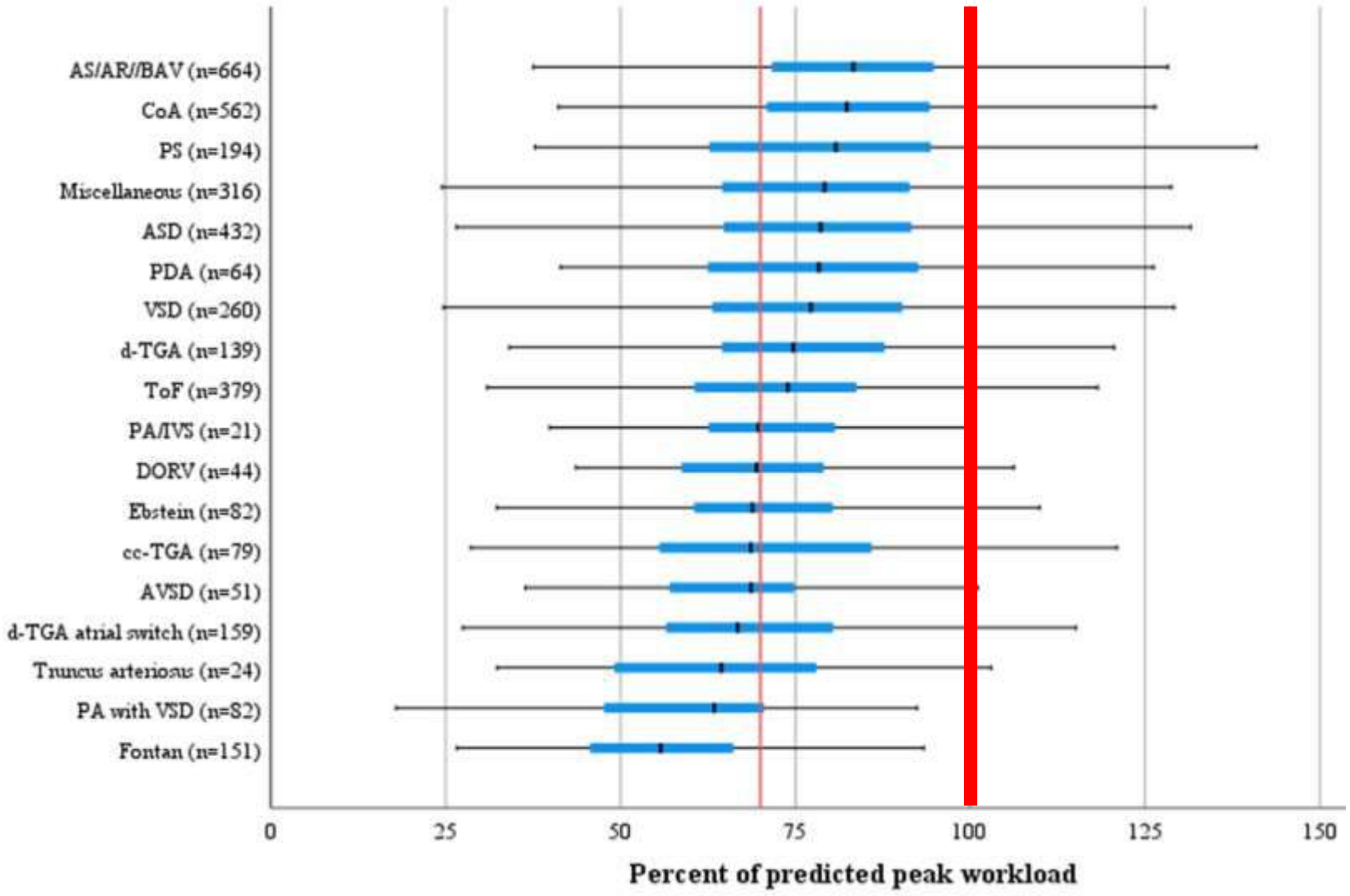
EXERCISE PERFORMANCE IN CONGENITAL HEART DISEASE



→ *Nearly 1/3 of children with CHD are overweight or obese*

Wikner et al. JACC Adv 2023, O'Byrne et al. WJPCHS 2018

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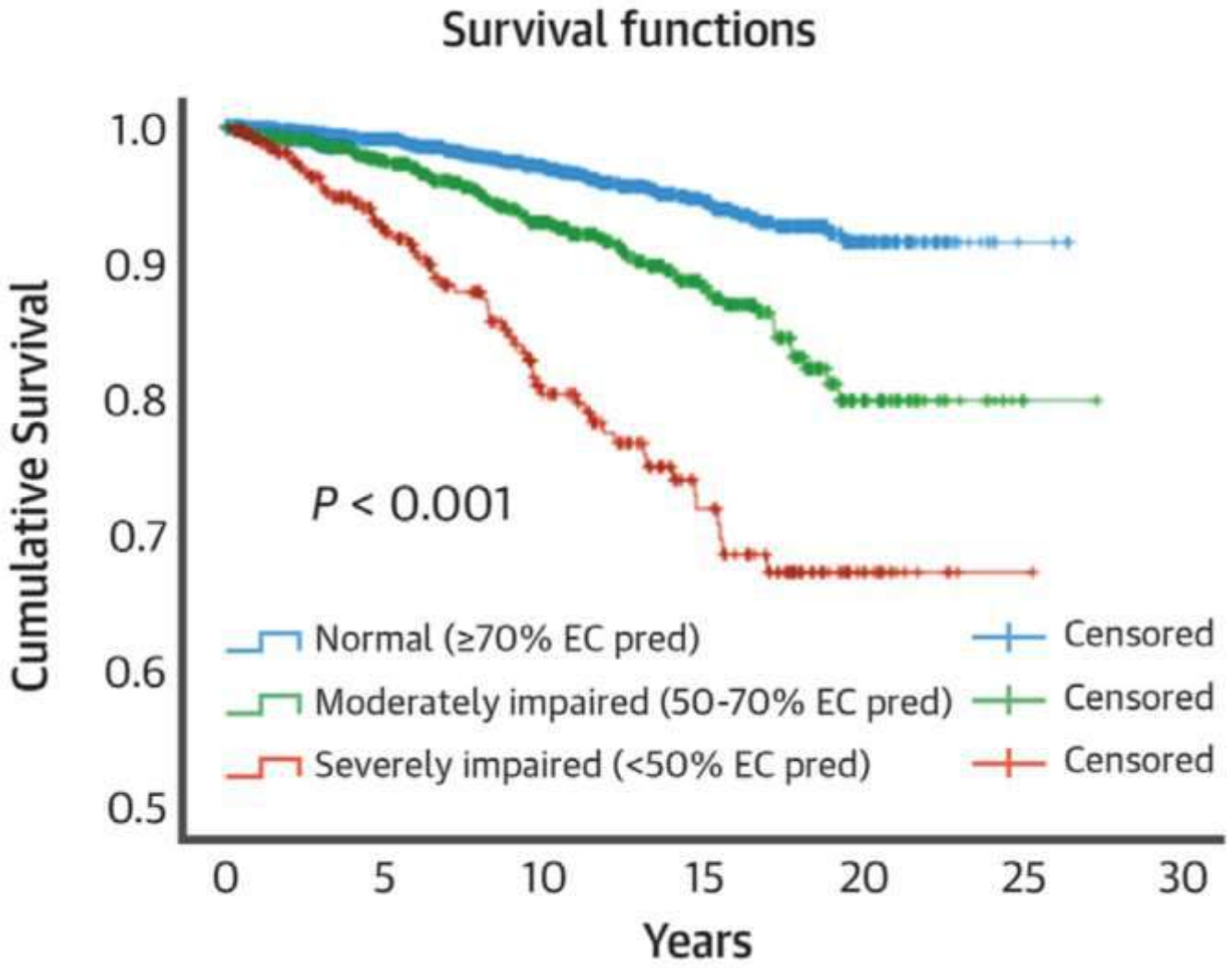
EXERCISE PERFORMANCE IN CONGENITAL HEART DISEASE



Decreased exercise capacity is associated with mortality

Wikner et al. JACC Adv 2023

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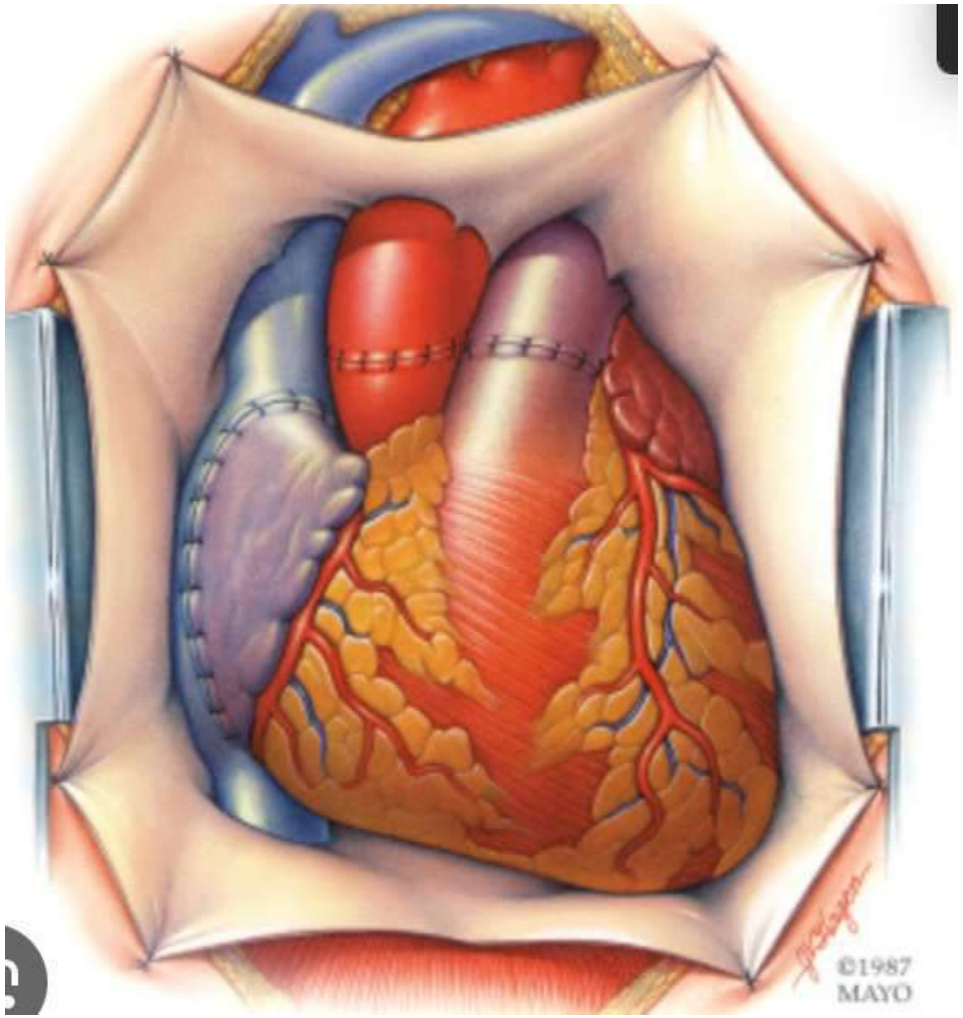
PEDIATRIC HEART TRANSPLANT SURVIVORS

Normal cardiac pump function is typical



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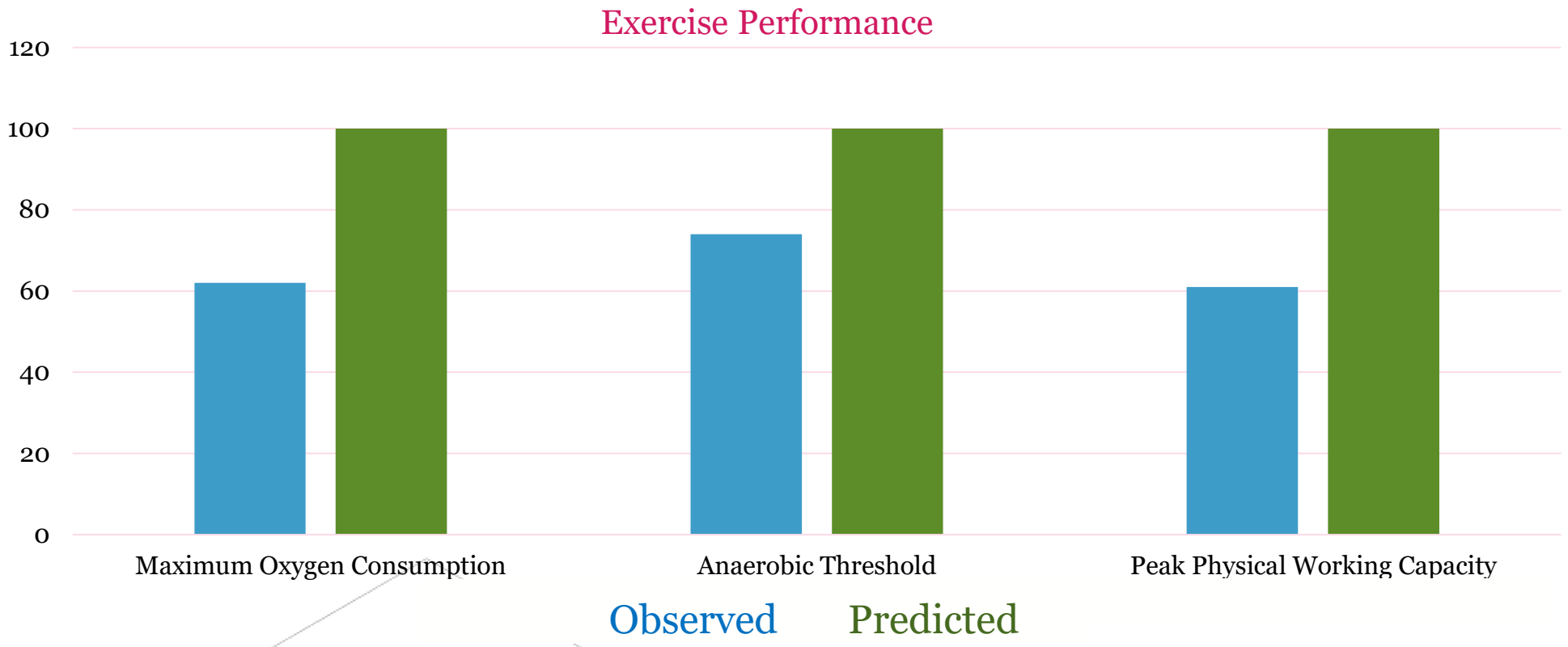
EXERCISE CAPACITY AFTER HEART TRANSPLANT

Study of 104 children with heart transplants



Markiewitz et al. JHLT: Open 2024

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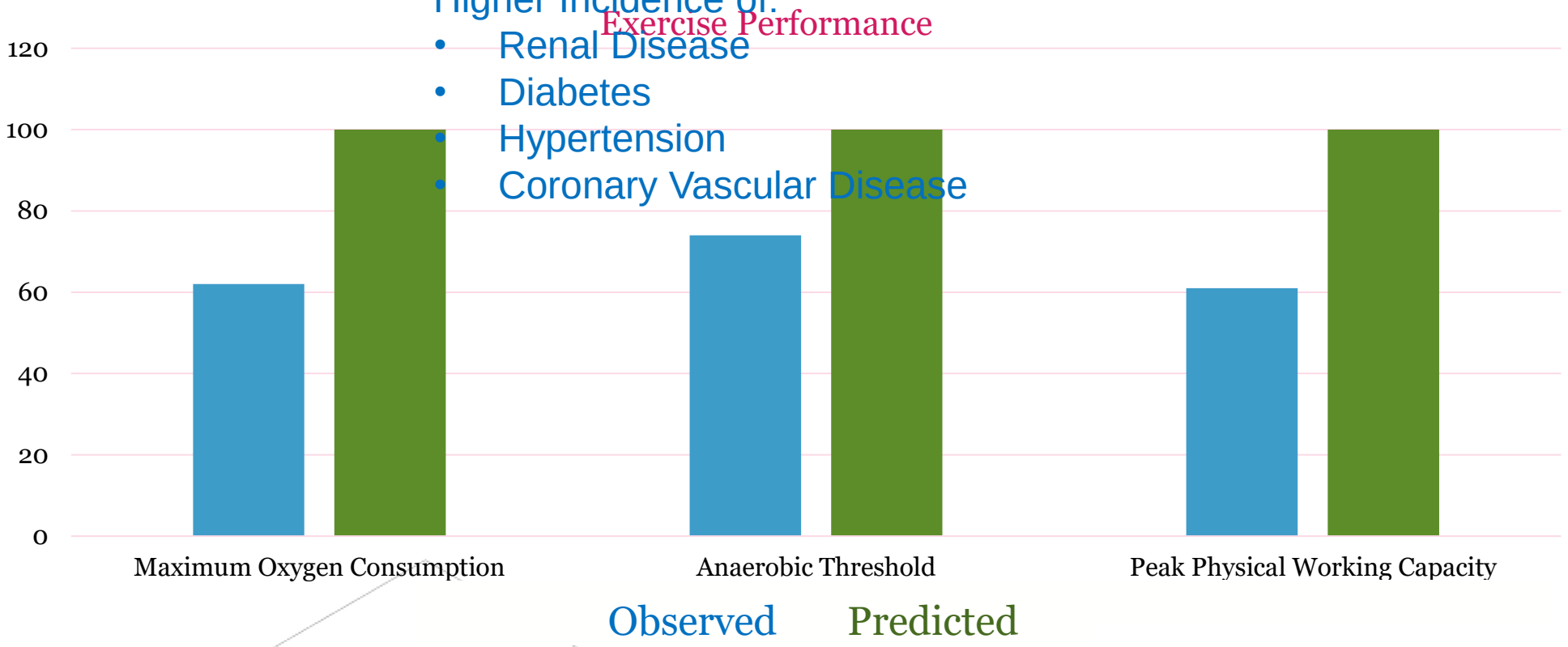
EXERCISE CAPACITY AFTER HEART TRANSPLANT

Study of 104 children with heart transplants

Higher Incidence of:

- Renal Disease
- Diabetes
- Hypertension
- Coronary Vascular Disease



Markiewitz et al. JHLT: Open 2024

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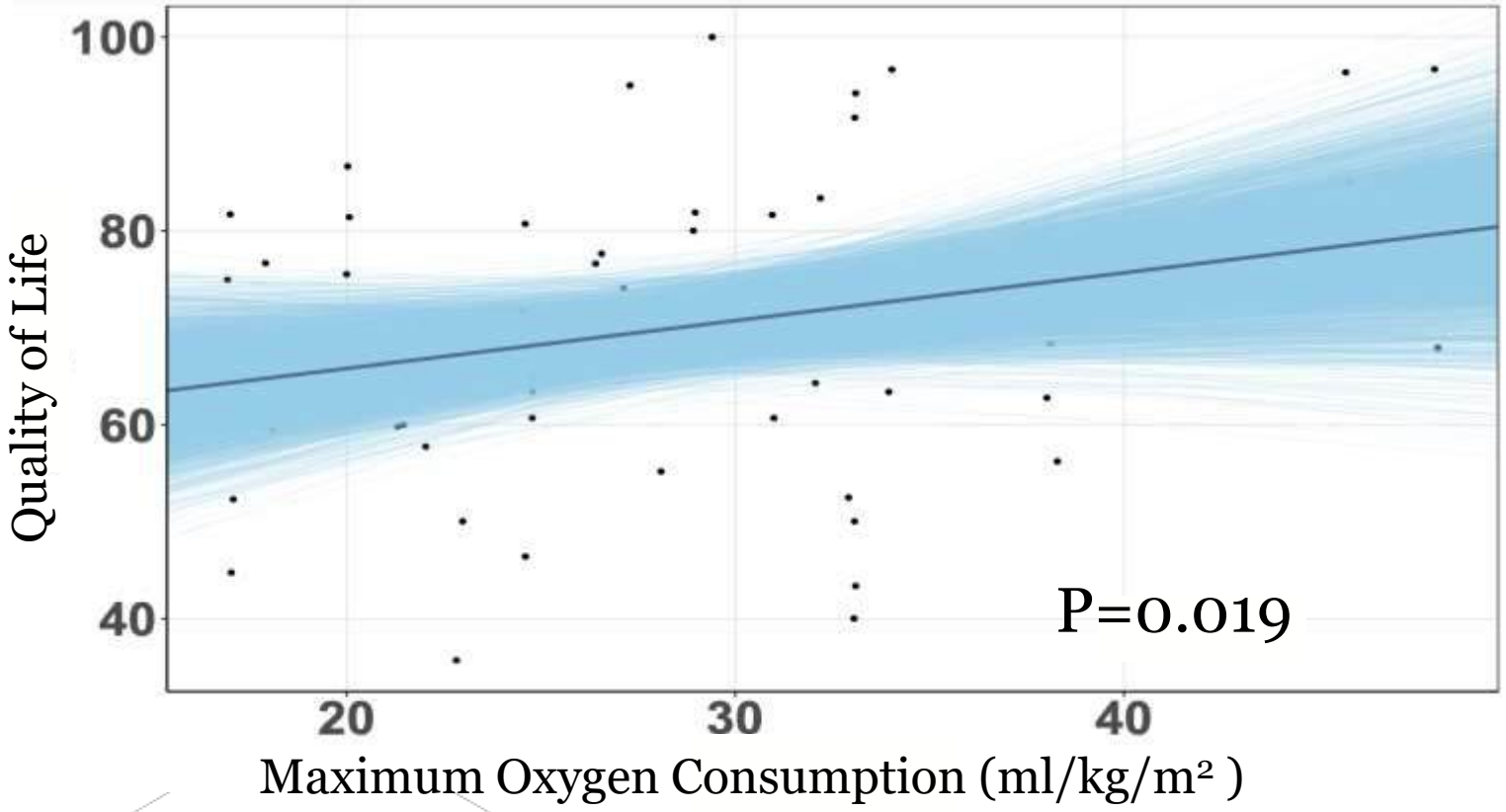
MENTAL HEALTH IN HEART TRANSPLANT SURVIVORS

- **Decreased quality of life in comparison to peers → Association with Exercise Performance**



Edelson et al. Under Review

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A LEGACY OF RESTRICTION: IMPACT

	Exercise Performance	Acquired CV risk Factors	Relationship of Activity with Mental Health
Hypertrophic Cardiomyopathy	↓	-Hypertension -Hyperlipidemia -Obesity	?
Congenital Heart Disease	↓	-Obesity	?
Heart Transplant Survivors	↓	-Diabetes -Hypertension -Coronary Disease	✓

IS THIS LEVEL OF RESTRICTION NECESSARY?

Children with Hypertrophic Cardiomyopathy

- Cardiac events: rest or sleep > activity

- Beneficial Effects of Exercise → Reverse remodeling: reduction in LV wall thickness and myofiber disarray, improved diastolic function

AHA/ACC Guidelines: ***The beneficial effects of exercise on general health can be extended to patients with HCM. Healthy recreational exercise (moderate intensity) has not been associated with increased risk.***

IS THIS LEVEL OF RESTRICTION NECESSARY?

Congenital Heart Disease

- 11,000 children with CHD:
    - 19 sudden deaths
    - none during exercise

Aortic stenosis:

- 403 patients, 45% restricted
- Death/transplant 6.5% restricted group, 3.2% unrestricted

Patients with complex congenital heart disease, including those with ToF and the Fontan procedure, are generally encouraged to participate in sports and/or exercise because improved exercise capacity is correlated with better outcomes

Silka et al. JACC 1998, Jortvirt et al. EHJ 2016, Coller et al. JACC 2024

IS THIS LEVEL OF RESTRICTION NECESSARY?

Heart Transplant Survivors

AHA Guidelines: ***heart transplant recipients have no specific activity restriction***

HT patients should engage in 60 minutes of physical activity every day

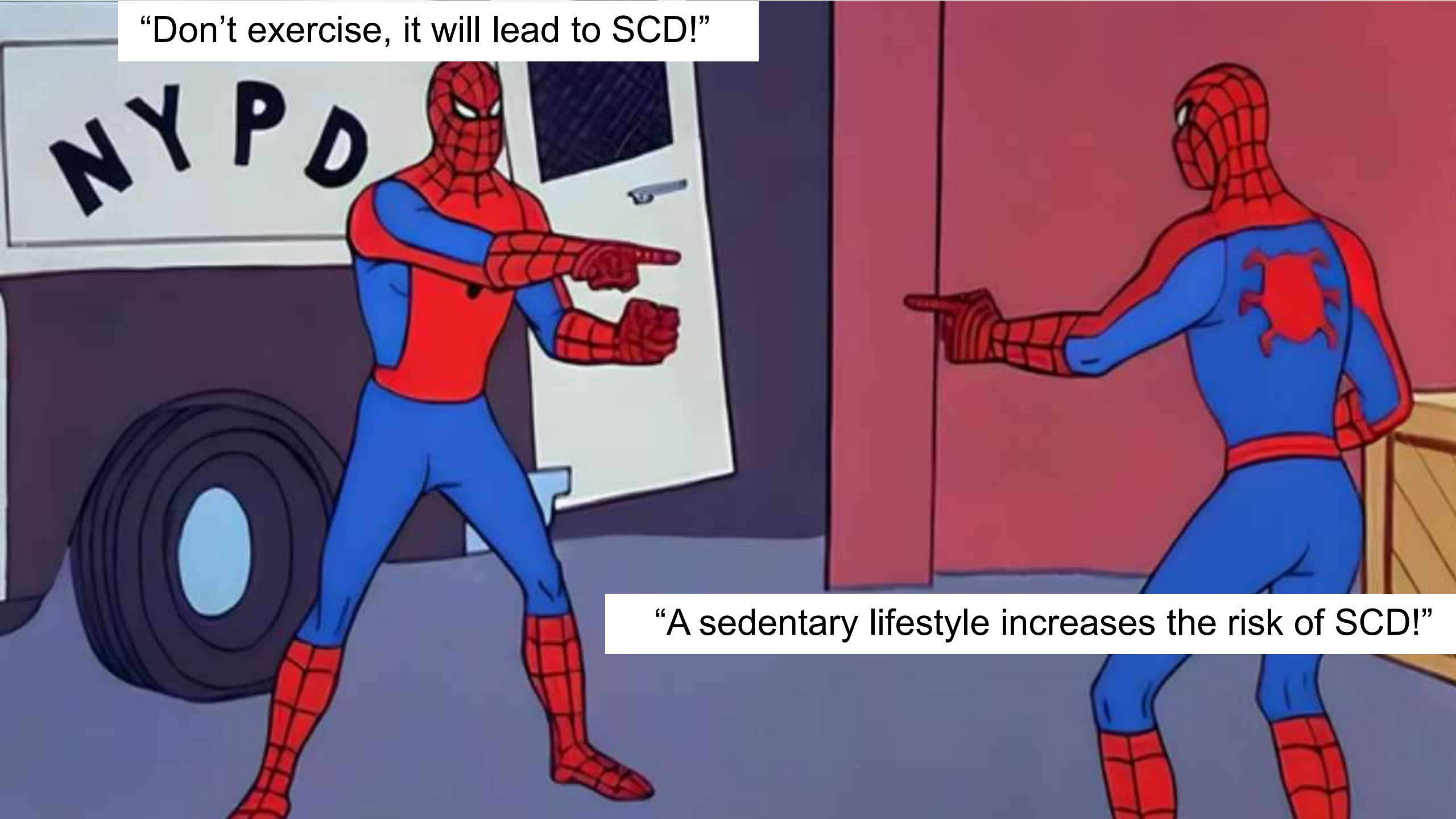
IS ACQUIRED CV DISEASE A PRIMARY DRIVER OF SCD?

Community wide assessment of Sudden Cardiac Death in the young

- 2002-2015
- 5-34 yrs old
- Catchment population = 1 million

High overall prevalence of established cardiovascular risk factors (obesity, diabetes mellitus, hypertension, hyperlipidemia, smoking) with ≥1 risk factors in 58% of SCA cases

Jayaraman et al. Circulation 2017


“Don’t exercise, it will lead to SCD!”
NYPD
“A sedentary lifestyle increases the risk of SCD!”

WHERE WE STAND

Children with cardiac disease have decreased physical activity levels

- This increases their overall cardiovascular risk profile
- The risk of being physically active is *likely* not as high as once believed
- We still don't completely understand risk

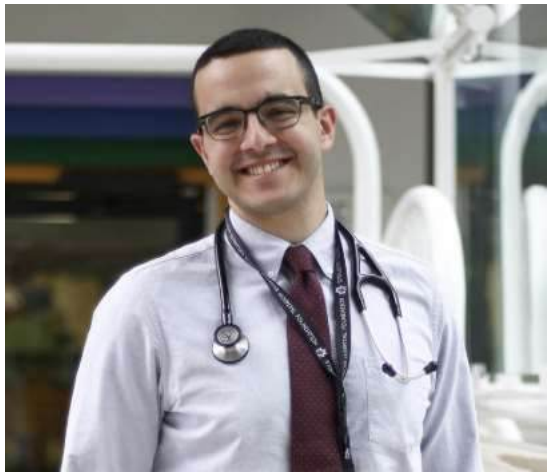
WHAT WE NEED: DISEASE SPECIFIC RISK

Outcomes Registry for Cardiac Conditions in Athletes

Sarcomeric Human Cardiomyopathy Registry

WHAT WE NEED: INVESTIGATORS

Programs aimed at safely and effectively increasing physical activity in children with cardiac disease



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WHAT WE NEED: PHYSICAL ACTIVITY SPECIFIC CARE

- Development of pediatric sports cardiology

- Use of Shared Decision Making Framework
  - Not an abdication of responsibility
  - Includes what we know and what we don't know

- Emergency Action Plans
    - Education to staff and teammates on SCA recognition and resuscitation
    - Practice drills
    - Presence of AED at all events

TAKE HOME POINTS

Children with heart disease are less active than peers, develop acquired CV risk factors

The risk of exercise/mobilization ↓ previously believed

Change is happening!

THANK YOU!