The Heart Is A Muscle Too!

The Cardiomyopathy In Duchenne Muscular Dystrophy



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Why should patients with Duchenne and Becker muscular dystrophy care about cardiac care?



The Heart is a Muscle TOO!!!

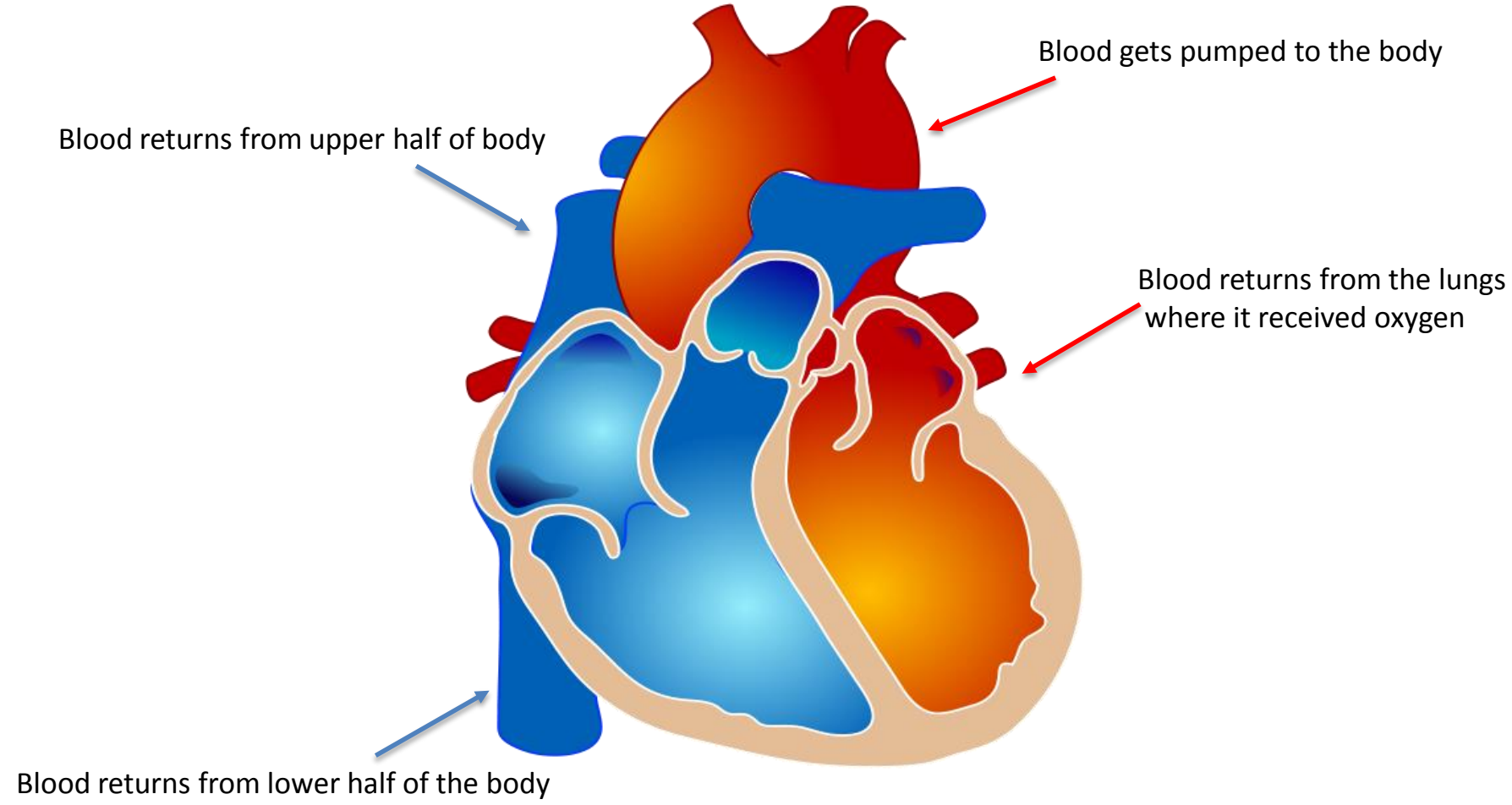


What do I need to know about the heart?



Important heart vocabulary

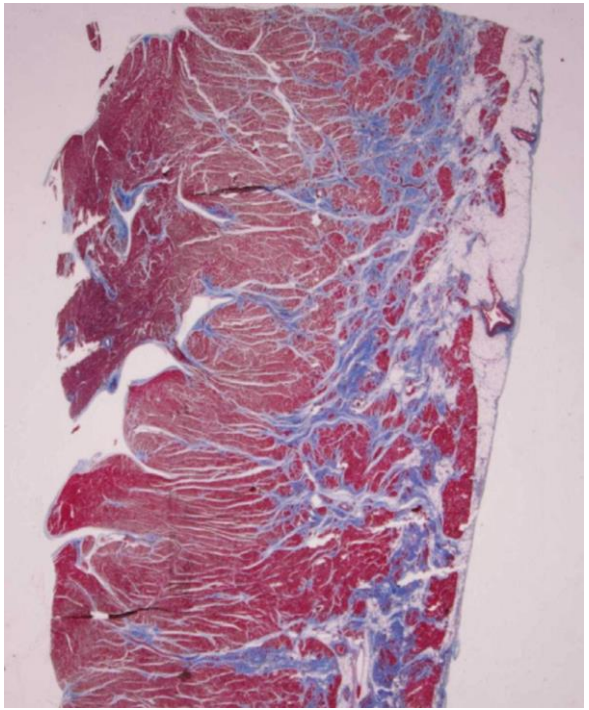
- **Cardiologist-** a heart doctor
- **Atria-** the chambers that receive blood in the heart
  - There are two atria (left and the right)
- **Ventricles-** are pumping chambers of the heart
  - there are two ventricles (left and right)
- **Cardiomyopathy-** disease of the heart muscle
- **Heart Failure-** when the heart is no longer able to meet the body's demands
- **Ejection fraction-** is a measurement of the percentage of blood leaving the heart each time it contracts.
- **Fibrosis-** scar tissue


Blood gets pumped to the body
Blood returns from upper half of body
Blood returns from the lungs where it received oxygen
Blood returns from lower half of the body

A key concept in understanding DMD cardiomyopathy

Cardiomyopathy in Duchenne muscular dystrophy is characterized by extensive sub-epicardial fibrosis





Fibrosis can be seen on cardiac MRI

- Sub-epicardial fibrosis is easily identified easily on CMR
  - Fibrosis starts in the sub-epicardium of the basolateral or inferolateral wall and progresses to apex and then septum
  - Importantly, it is present
    - Prior to decline of LV function
    - Early in the disease
- Of note, sub-epicardial enhancement is seen in myocarditis
  - Represents inflammatory infiltrate
- Begs the question:
  - Are similar pathophysiological mechanisms at work in DMD and myocarditis?

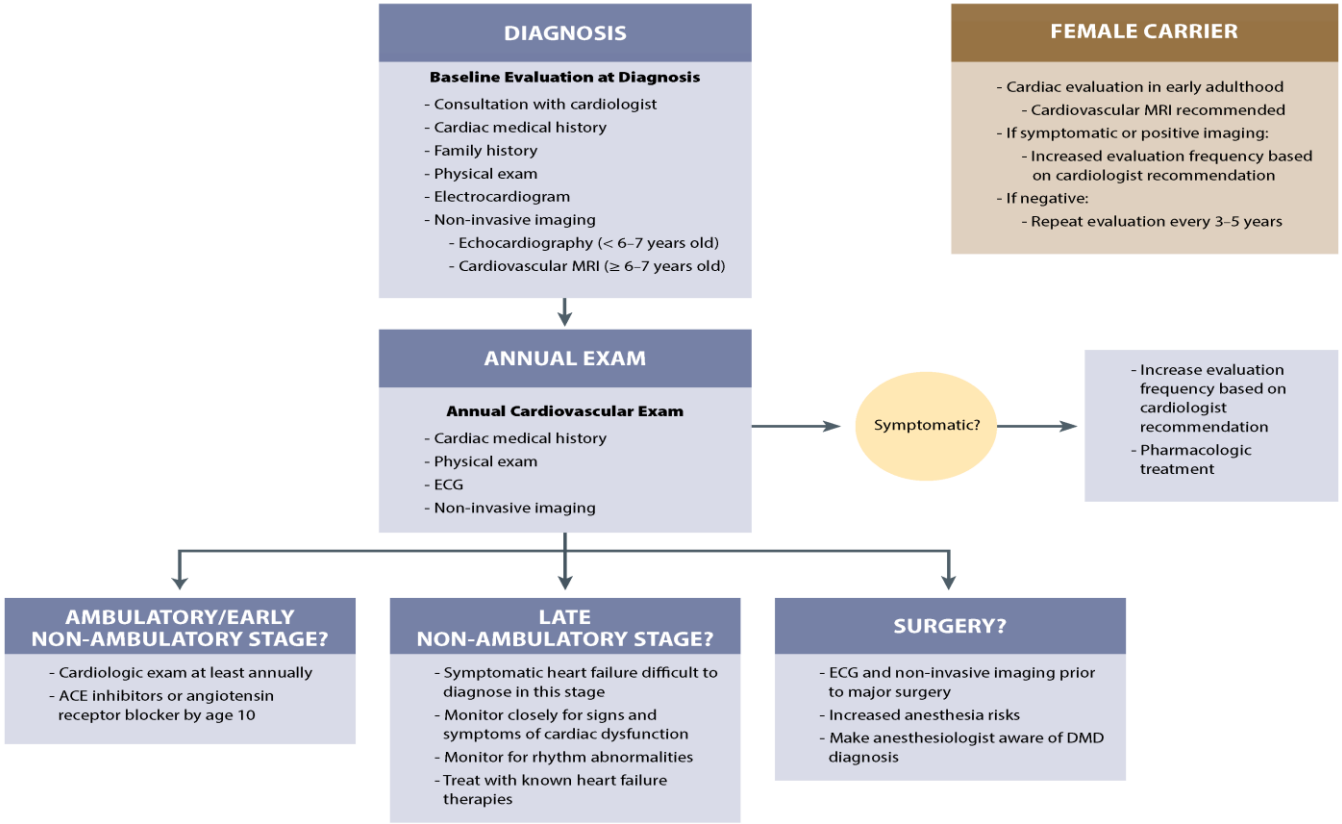


How should I care for my son’s heart?

Diagnosis and management of Duchenne muscular dystrophy, part 2: respiratory, cardiac, bone health, and orthopaedic management

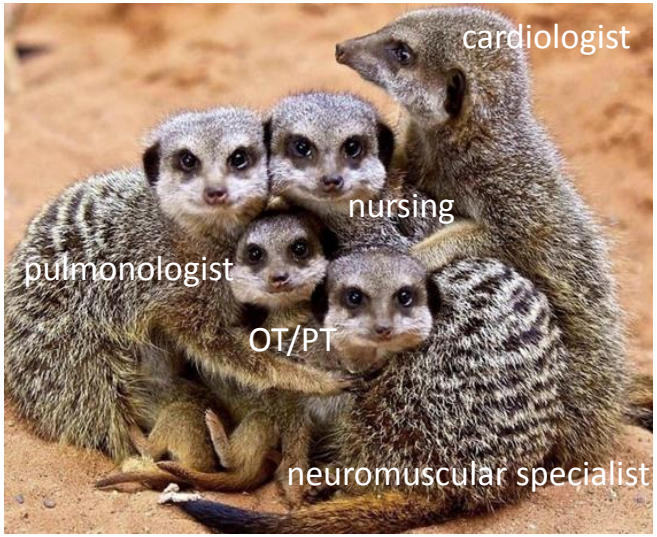
*David J Birnkrant, Katharine Bushby, Carla M Bann, Benjamin A Alman, Susan D Apkon, Angela Blackwell, Laura E Case, Linda Cripe, Stasia Hadjiyannakis, Aaron K Olson, Daniel W Sheehan, Julie Bolen, David R Weber, Leanne M Ward, for the DMD Care Considerations Working Group**

Cardiac Monitoring, Diagnosis, and Treatment Algorithm for Patients with DMD



Chest pain in DMD/BMD

- FREQUENT and often blamed on chest wall pain
- Often dismissed in a pediatric ER
- Formal cardiac evaluation rarely undertaken
  - ECG is sometimes obtained
  - Troponin I (cTnI) rarely obtained
    - cTnI is a marker of cardiac muscle tissue injury
    - cTnI is normal or minimally elevated at baseline in DMD
- We hypothesize that DMD cardiomyopathy results from episodic heart muscle injury rather than continuous ongoing injury
  - Silent and recurrent events lead to this injury
  - Process similar to that which occurs in skeletal muscle
  - Suggests a step wise model of disease progression as opposed to a linear one
- Unknown if there could there be external triggers (viral infection, stress, other illnesses)







We can achieve better outcomes with breakdown of clinical silos….
and giving each other a BIG GROUP HUG

THANK YOU



