Cardiology Standards of Care

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Cardiac Care of the Boys
And their moms

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The Heart has dystrophin in it Too!!!
What does “cardiomyopathy” mean?

Disease of heart muscle

Normal heart

Dilated heart (systolic heart failure)
What does “Heart Failure” mean?

• Complicated

• **DOES NOT** mean the heart has stopped working

• **DOES** mean that the heart is not meeting the body’s needs

• Heart failure occurs when heart function is poor
  • can occur with good function and increased demand

• Body’s response is initially helpful but eventually results in harm

• People can live for a long time with heart failure if treated
Who should care for my son’s heart?

• **Cardiologist** is a “heart doctor”
  - Not all cardiologists are the same
  - Pediatric **cardiologists**
    - Train in pediatrics and cardiology
  - Adult **cardiologists**
    - Train in adult medicine and cardiology

• Some **cardiologists** have special interests
  - Heart failure/transplantation
  - Neuromuscular disorders
  - Talk to your son’s doctor about finding an expert who is comfortable treating “heart failure” or Duchenne
When should cardiac care begin?

Care Should:

• Begin at diagnosis
• Repeat investigation:
  • At least annually until age 10
    • Or with the onset of cardiac signs and symptoms
  • Biannually after the age of 10
    • Or more frequently based on cardiac signs and symptoms
  • Prior to any major surgery
Diagnostic Testing of the conduction (electrical) system

- Electrocardiogram (ECG)
  - Evaluates heart rate and rhythm
  - Creates an electrical “picture” of the heart

- Holter monitor
  - Allows you to monitor the heart rhythm over 24 hours

- Event Monitor
  - Allows you to monitor the heart during times of concern
Diagnostic Testing of the contraction (muscle) system

- Imaging of the heart evaluates structure, size, and function
Diagnostic Testing of the contraction (muscle) system

• **Echocardiogram**—ultrasound of the heart
  • Evaluate anatomy and function
  • Advantages:
    • Readily available/portable
    • Quick (scan typically takes less than 30 minutes)
  • Disadvantages:
    • Image quality affected in the older / bigger DMD patient)
    • Scoliosis and positional needs impact this as well

• **Cardiac MRI**
  • Advantages
    • No radiation exposure
    • Detailed and accurate cardiac information is obtained
      • Additional information regarding fibrosis (LGE)
  • Disadvantages
    • IV placement
    • Claustrophobia
    • Cost
    • Sedation may be required, esp in younger or claustrophobic boys
Signs and Symptoms of a heart muscle problem

Symptoms may be subtle and non-specific

- Weight gain or loss
- Swelling/edema of feet, abdomen
- Palpitations (sense of fast or irregular heart rate)
- Syncope (fainting)
- Chest pain
  - musculoskeletal
  - coronary arterial
  - myocarditis (infection or inflammation)
- Nausea, vomiting, diarrhea, abdominal pain
What treatments are available?

Heart drug therapies:
- **ACE inhibitors**
  - enalapril, lisinopril, perindopril
- **Angiotensin- receptor blockers**
  - Losartan
- **β-blockers**
  - metoprolol, carvedilol
- **Diuretics**
  - furosemide, thiazide
- **Aldosterone receptor antagonists**
  - Spironolactone, eplerenone
- **Anti-coagulation**
  - Coumadin, Aspirin

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Starting Medications

• Most DMD patients will develop cardiac dysfunction at some time during their lives

• Families exert significant pressure to do something!!

• Begin ACE inhibitors
  • Left ventricular enlargement
  • Ventricular pump dysfunction
  • Myocardial fibrosis
  • Tachycardia (?)
  • Age 10 or over
DMD Carriers

- Cardiac disease may be the only manifestation
- Cardiomyopathy risk increases with age
  - Approximately 350 DMD/BMD carriers
    - age < 16 yrs: all normal
    - age 16-30 yrs: 6%; 31-50 yrs: 9%; > 50 yrs: 16% DCM
- Current recommendations
  - Baseline evaluation as young adult
    - Frequency of follow-up unclear (? Every 3 to 5 years)
    - Be aware of symptoms
    - Take care of yourself
      - minimize other CV risks
        - smoking, HTN, cholesterol
Conclusions

• Cardiac evaluation should begin at diagnosis
• Follow-up is important
• Pharmacotherapy is likely of benefit
• Care of the carrier is important
• Transition is important