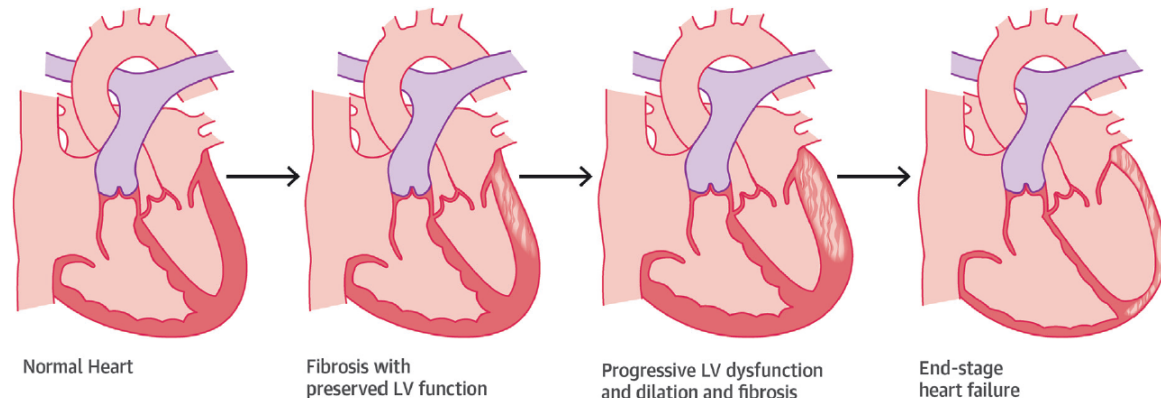


Cardiac management of Duchenne muscular dystrophy: the 2018 DMD care considerations

Implications for the Netherlands

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No financial disclosures

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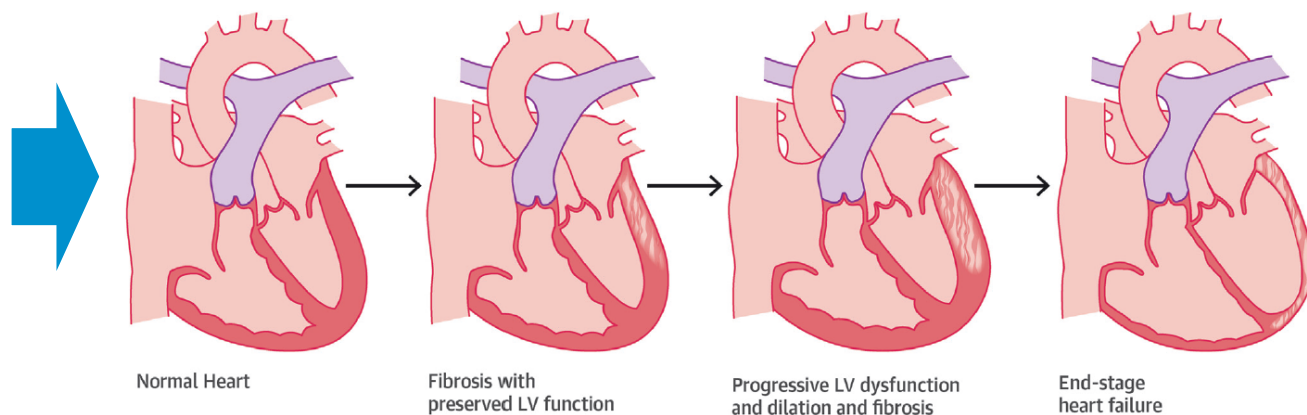
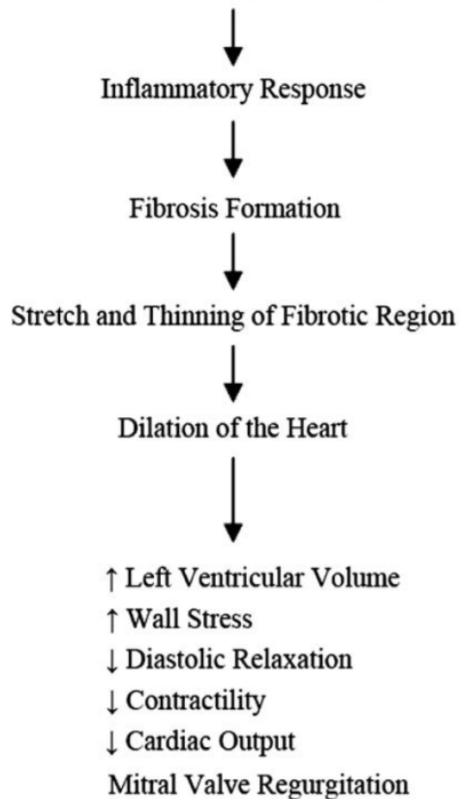
Dr EH Niks, pediatric neurologist, LUMC

Timeline:

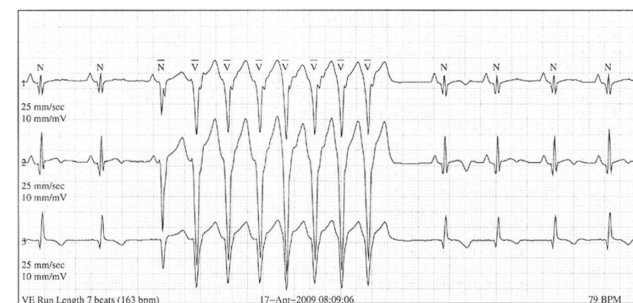
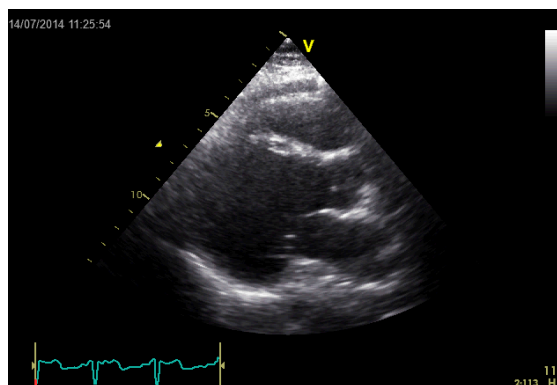
- Based on 2018 DMD care considerations 2 TelComs June and August 2018
- Discussion on the implications of the DMD care consideration on Cardiac management of DMD for DMD management in The Netherlands
- Discussion of results presented at Duchenne Care Conference 7th Sept

The problem of the myocardium in DMD pts

CARDIOMYOCYTE DEATH



J Am Coll Cardiol 2016;67:2533-46



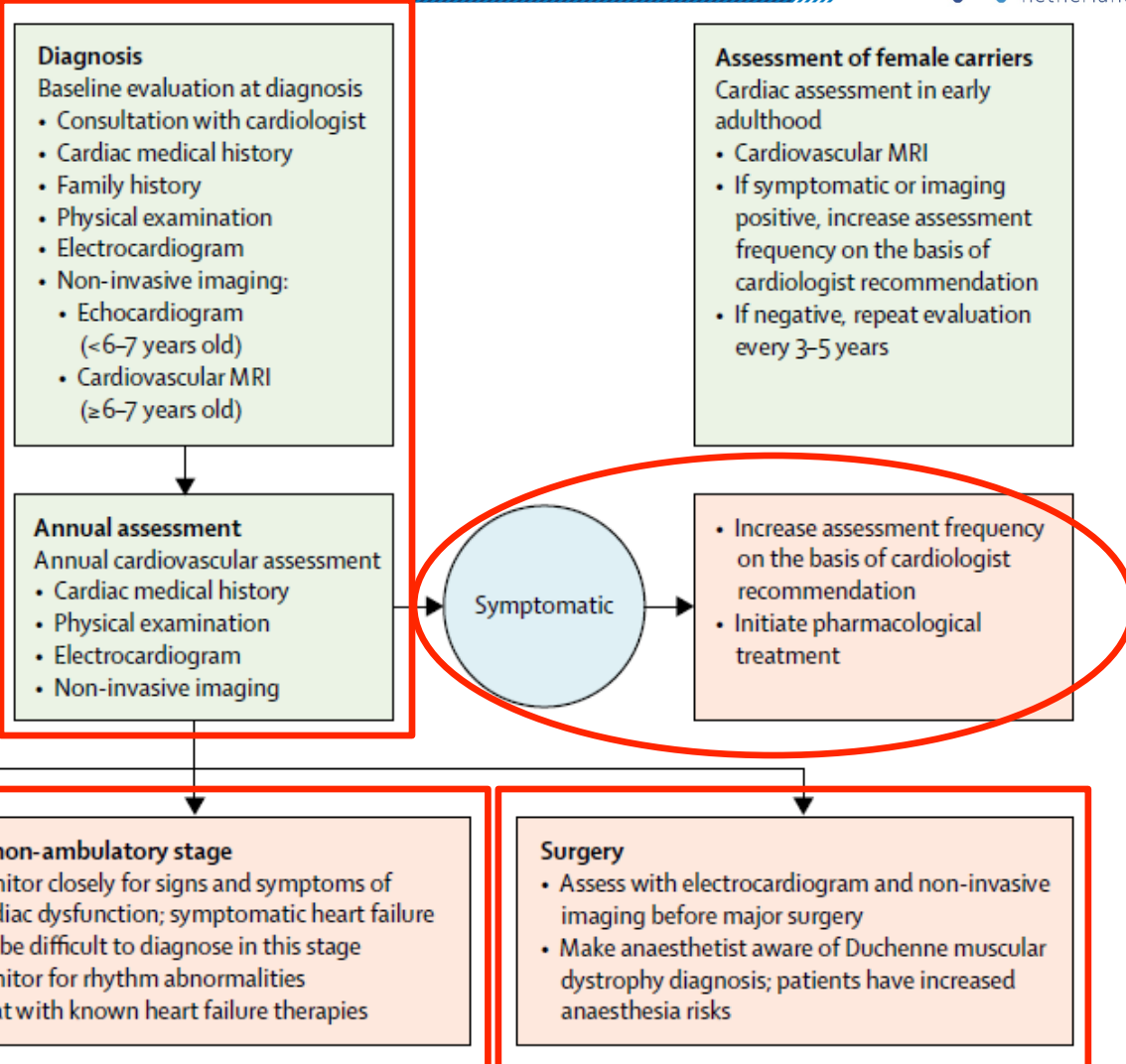
Muscle Nerve 44: 8–19, 2011

Cardiac follow-up and management of DMD pts

Lancet Neurol 2018; 17: 347-61

Most important differences

- cMRI imaging of choice
- Annual cMRI from age 6-7
- Start ACEi at age 10



Cardiac evaluation

- When to start cardiac evaluation in DMD pts
- How to detect cardiac disease in DMD pts

Pharmacological Intervention

- When to treat DMD pts
- How to treat DMD pts

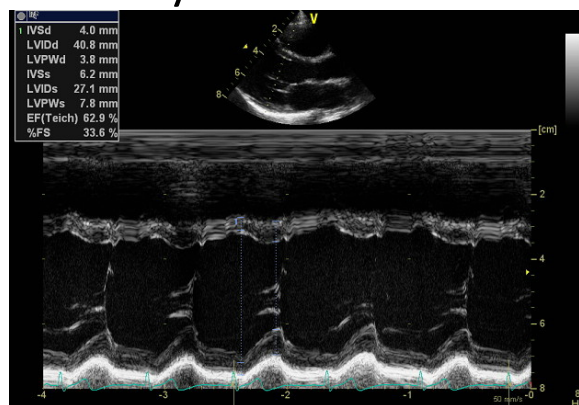
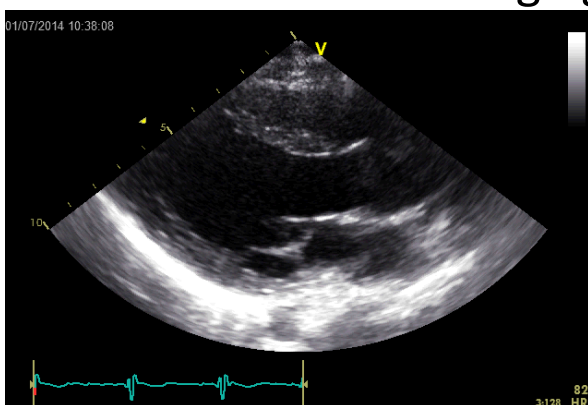
Invasive treatment of cardiac disease

- Mechanical support of end-stage heart failure
- ICD-treatment for primary or secondary prevention of VT/VF

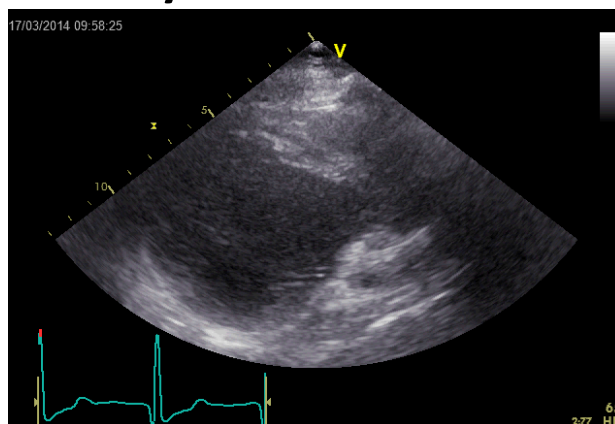
Cardiac Care in DMD carriers

Cardiac evaluation

- Start from diagnosis-even in young pts
- First-line non-invasive imaging modality in children: **echocardiography**



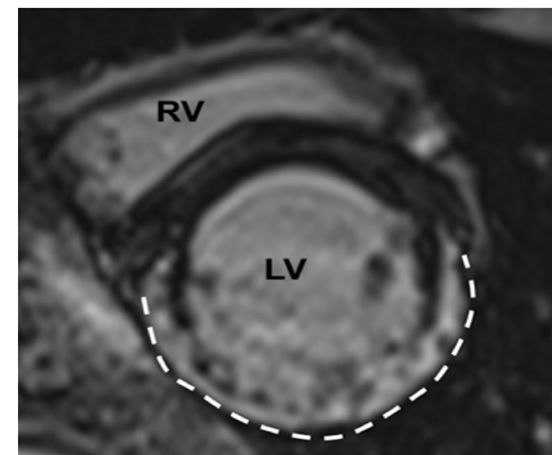
- However with advancing age acoustic window hampered by increase in BMI, scoliosis, immobility.



Cardiac evaluation with cardiac MRI

- Advantages of **cardiac MRI**:
 - Gold standard of cardiac dimensions and function
 - Information on global and focal fibrosis
- Disadvantages of **cardiac MRI**:
 - Inability to perform cMRI in young children <8yoa or older pts
 - Artifacts from scoliosis repair
 - Time consuming and challenging for pts in non-ambulatory stage, esp pts with contractures or on mechanical ventilation

FIGURE 7 CMR Demonstrating Dilated Cardiomyopathy and Fibrosis in a Patient With DMD J Am Coll Cardiol 2016;67:2533-46



- **2018 DMD Care consideration:**
 - Annual non-invasive imaging: cMRI imaging modality of choice.
- **Considerations Dutch working group**
 - From diagnosis on: annual echocardiographic examination
 - First cMRI between the age of 8-10 yrs
 - LV dimensions and function
 - Presence and extent of fibrosis
 - Follow-up cMRI
 - For guidance of management/treatment
 - **Synchronize cMRI-protocols in the Netherlands**

- **2018 DMD Care consideration:**
 - Annual non-invasive imaging: cMRI imaging modality of choice.
- **Considerations Dutch working group**
 - **Needed on an annual base?**
 - Discussion: **what does cMRI add** when a patient is already on ACEi with good normal/stable cardiac dysfunction on echocardiography?
 - With good acoustic window and normal function: suggest cMRI 2-3 years
 - If acoustic window deteriorates: increase frequency of cMRI
 - If symptoms of heart failure: indication for cMRI and increase in frequency of cardiac evaluation
 - If cardiac function is seriously depressed and on maximal therapy-
additional value of cMRI?

ECG and 24h-Holter monitoring

- Optimum frequency of monitoring not established



Muscle Nerve **44**: 8–19, 2011

2018 DMD Care consideration:

- Initiate annual 24h-Holter monitoring with onset of signs of cardiac involvement

Considerations Dutch working group

- Based on recommendation: proposal for cross-sectional evaluation of all DMD-pts with 24h-Holter monitoring and evaluate yield after 2 years
- Further (more specific) recommendation after evaluation of results

When and what cardiac medication to start in DMD pts?

2018 DMD Care consideration:

- ACEi or ARB: first-line treatment, additional β -blocker
- Start ACEi or ARB at the age of 10 years
- With sign/symptoms cardiac failure
- LV dilatation and dysfunction
- Cardiac fibrosis

Considerations Dutch working group

- To follow the 2018 DMD Care consideration
- Preferably with consensus on what ACEi and β -blocker to use
- Current proposal: ACEi: perindopril and β -blocker: carvedilol

- **Maximise pharmacological treatment**
- **Prevent thrombo-embolism**
 - No recommendation what agent to use: adult AF-heart failure guidelines
 - At the discretion of the cardiologist
- **Invasive treatment of cardiac failure**
 - With failure of maximal pharmacological therapy
 - Mechanical circulatory support-heart transplant
 - Inherent high risk
 - Only case reports available
 - To be discussed within multi disciplinary team on a case-by-case basis

Dutch working group to follow 2018 DMD Care consideration

- In LUMC limited experience with CRT-D in DMD pts with cardiac failure

- **No recommendation in 2018 DMD Care consideration**
- Implantable cardioverter defibrillator (ICD) can be considered as:
 - Primary prevention indication based on adult heart failure guidelines
 - Secondary prevention

Dutch working group

- To be discussed within multi disciplinary team on a case-by-case basis

At risk for cardiomyopathy

- Baseline cardiac assessment in early adulthood
- Follow-up cardiac evaluation every 3-5 years
- More frequent evaluation and treatment if signs of cardiac disease develop

Dutch working group to follow 2018 DMD Care consideration

Dutch working group vs 2018 DMD Care consideration: discussion @ Duchenne Care Conference

Writing of Dutch DMD Care considerations based on 2018 Care considerations

Discussion of Dutch DMD Care consideration with **Netherlands Society of Pediatric Cardiology** and **Netherlands Society of Cardiology** for approval

After approval implementation of consideration in clinical care