Dystrophinopathy Ambulatory Electrophysiologic Monitoring Harmonization

**BACKGROUND**

Sudden death and aborted sudden death have been described in dystrophinopathy. The exact frequency of arrhythmia death has been difficult to assess given the frequency of unwitnessed mortality and potential contributions of respiratory failure. In addition, heart rate, heart rate variability, and arrhythmias have also been described in dystrophinopathy and are thought to be driven by fibrofatty infiltration of the myocardium.1-3 Based on the risk and substrate, ambulatory monitoring is common, although highly variable in terms of frequency and method of screening.4 This document is not meant to supersede recommendations such as the HRS consensus statement, but to provide specific guidance in relation to dystrophinopathy and to help harmonize clinical practice within the network. In particular consensus was sought regarding periodicity and duration of monitoring which are not specifically addressed in the existing guidelines or consensus statements.

**ACTION REVISED DATE:** 04/08/2024

**OBJECTIVES**

To harmonize clinical practice of ambulatory rhythm monitoring in dystrophinopathy based on expert consensus. There is no sufficient data at this time to specifically recommend a specific method, duration, or indication for initiation.

**PROTOCOL**

**Duchenne muscular dystrophy (DMD)**

Indication for Initiation for Monitoring

* Atrial and ventricular ectopy have been described in DMD. The frequency and severity of each generally correlates with the severity of cardiomyopathy, although individual cases of symptomatic arrhythmia including sudden cardiac death have been reported early in the disease process.1, 2, 5 The substrate is generally thought to be areas of fibrofatty replacement of the myocardium, although the long term risk of arrhythmias in men with DMD and with preserved or relatively preserved systolic function is not well delineated. Using this as the paradigm, we propose the following:
  + Ambulatory rhythm monitoring is not recommended in asymptomatic young patients < 18 years, without evidence of cardiomyopathy (LGE, systolic dysfunction, or left ventricular [LV] dilation), and who do not require night time respiratory support.
    - Yearly ambulatory monitoring is reasonable in adults (>18 years) and those who require night time respiratory support given the minimal data in these subgroups.
  + It is reasonable to obtain yearly ambulatory rhythm monitoring for patients with evidence of cardiomyopathy (LGE, systolic dysfunction, or LV dilation).
  + Yearly ambulatory rhythm monitoring is recommended in patients with systolic dysfunction.

Duration of Monitoring

* Longer duration, continuous, monitoring can increase the frequency of arrhythmia detection in relation to 24 hour Holter or event based monitoring systems.6 This holds potential value in understanding the frequency of ectopy and arrhythmias, disease progression, and outcomes. However, further study is needed to demonstrate this clinical value and to understand the impact of this approach on resource utilization. The value and cost of the longer term monitoring strategies is likely center and insurance specific and these should be investigated accordingly when discussing risks and benefits with patients/families. With these factors in mind, we propose the following:
  + 24 hour Holter monitors are recommended for ambulatory monitoring of arrhythmias in dystrophinopathy patients with evidence of cardiomyopathy
  + Use of longer duration non-implantable monitors (e.g. Zio Patch) are reasonable for ambulatory monitoring of arrhythmias in dystrophinopathy patients with evidence of cardiomyopathy.

Implantable cardiac monitoring

* As noted above, longer duration, continuous, monitoring can increase the frequency of arrhythmia detection.6 There is limited data on the risks or benefits of implantable loop recorders (ILR) in DMD. There is an ongoing study specifically addressing this topic. As we await these data, we propose the following:
  + Use of ILRs is reasonable in symptomatic patients where other methods of monitoring are not feasible or non-diagnostic (e.g. patients with developmental delay unable to tolerate external monitors, skin sensitivity, etc).

Specific considerations in Becker muscular dystrophy (BMD) and carriers

* There is limited data describing the utility of ambulatory monitoring in persons with BMD and carriers (both symptomatic and non-symptomatic). With this in mind, we propose the following;
* Yearly ambulatory rhythm monitoring is recommended in patients with BMD and with evidence of cardiomyopathy (LGE, systolic dysfunction, or LV dilation).
* Yearly ambulatory monitoring is recommended in carriers with evidence of cardiomyopathy (LGE, systolic dysfunction, or LV dilation).
* Yearly ambulatory monitoring is reasonable in adults (>18 years) with BMD and those who require night time respiratory support in the absence of cardiomyopathy given the limited data in these subgroups.
* Yearly ambulatory monitoring is reasonable in symptomatic carriers who require night time respiratory support in the absence of cardiomyopathy given the limited data in this subgroup.

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***Disclaimer:*** *The ACTION network is focused on quality improvement efforts such as harmonizing best practice protocols, disseminating them among institutions, and helping centers to improve care practices at the local level. This protocol was developed as a consensus tool for pediatric VAD programs. The information in the protocols are based on center practices, individual opinions, experiences, and, where available, published literature. Centers may choose to adapt this protocol to include in their center-specific protocols with reference to ACTION with the understanding that these are meant as guidelines and not standard of care. (Revised: 05/24/2021)*