American Heart Association | American Stroke Association

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Top Ten Things to Know
Management of Cardiac Involvement Associated With Neuromuscular Diseases

1. Neuromuscular diseases (NMDs) encompass a broad spectrum of diagnoses with overlapping but distinct phenotypes; cardiac involvement is common to many NMDs.
2. The prevalence of NMDs ranges in the population from 1:3,600-9,300 male births for Duchenne Muscular Dystrophy (DMD) to 1:140-670,000 births for Barth Syndrome (BTHS).
3. The Statement includes a comprehensive overview of the major categories of NMDs with cardiac involvement, including a brief background on the gene defect(s), common clinical manifestations, current therapies, knowledge gaps, and clinical treatment suggestions for clinicians to tailor their treatment approaches to prevent or treat heart failure.
4. The NMDs discussed in this statement include: DMD, BTHS, Becker Muscular Dystrophy (BMD), Limb Girdle Muscular Dystrophy (LGMD), Emery Dreifuss Muscular Dystrophy (EDMD), Myofibrillar Myopathy (MFM), , Friedreich Ataxia (FA), Myotonic Dystrophy (DM), and Congenital Myopathy (CM).
5. Clinically relevant cardiac involvement in NMDs most commonly falls into one of two major categories: 1) cardiomyopathy and 2) conduction defects with arrhythmias; and the severity and onset of cardiac complications varies significantly across classes of NMDs.
6. Most forms of cardiac involvement are detected from childhood to the second decade of life (e.g. DMD, DM, FA, EDMD, BTHS), but others may remain asymptomatic until later in life (BMD, some forms of congenital myopathy (CM abd MFM).
7. Some NMDs increase the risk of cardiomyopathy and heart failure (e.g. DMD, BMD, FA), others elevate the risk of arrhythmia and sudden death (e.g. EDMD, LGMD1B, and DM1), others increase risk of both (e.g. BTHS, MFM), and yet others do not involve the heart (e.g. LGMD1D, oculopharyngeal muscular dystrophy).
8. Considerations for the following therapies are discussed for NMDs with cardiac involvement: Angiotensin Converting Enzyme (ACE) inhibitors and Angiotensin II Receptor Blockers (ARBs); β-adrenergic blockers; mineralocorticoid antagonists; glucocorticoids; diuretics; anticoagulation; antiarrhythmic drugs in NMD; exercise, physical therapy, and weights; assisted ventilation; and cardioverter-defibrillator and resynchronization therapy.
9. The statement includes discussions of end-stage heart failure, transition of care, and supportive and palliative care.
10. Over the last several decades, there has been continued improvement in survival and quality of life for individuals with NMD. However, cardiac impairment represents a major obstacle to further improvements. The cardiac community has an opportunity to devote resources to enhancing cardiac outcomes in this population.

Feingold B, Mahle WT, Auerbach S, Clemens P, Domenighetti AA, Jefferies JL, Judge DP, Lal AK, Markham LW, Parks WJ, Tsuda T, Wang PJ, Yoo S-J; on behalf of the American Heart Association Pediatric Heart Failure Committee of the Council on Cardiovascular Disease in the Young; Council on Clinical Cardiology; Council on Cardiovascular Radiology and Intervention; Council on Functional Genomics and Translational Biology; and Stroke Council. Management of cardiac involvement associated with neuromuscular diseases: a scientific statement from the American Heart Association [published online ahead of print August 24, 2017]. *Circulation*. doi: 10.1161/CIR.0000000000000526.