of Neuromuscular & Electrodiagnostic Medicine (AANEM), the American College of Cardiology (ACC), the American Heart Association (AHA), the American Society of Anesthesiologists (ASA), the Asia Pacific Heart Rhythm Society (APHRS), the Child Neurology Society (CNS), the European Heart Rhythm Association (EHRA), the Heart Failure Society of America (HFSA), the Japanese Heart Rhythm Society (JHRS), the Latin American Heart Rhythm Society (LAHRS), the Pediatric and Congenital Electrophysiology Society (PACES), and the Sociedade Brasileira de Arritmias Cardíacas (SOBRAC), is intended to guide electrophysiologists, cardiologists, neurologists, and other clinicians in caring for patients with arrhythmic complications of NMDs.

The document presents an overview of arrhythmias in NMDs followed by detailed sections on specific disorders with an emphasis on arrhythmic cardiac manifestations. Conditions with similar clinical presentations and demonstrated cardiac involvement are grouped into sections: Duchenne muscular dystrophy (DMD), Becker muscular dystrophy (BMD), and limb-girdle muscular dystrophy (LGMD) type 2 (LGMD2) (see Section 3.1, Table 4 for additional nomenclature); myotonic dystrophy (DM) type 1 (DM1) and type 2 (DM2); Emery-Dreifuss muscular dystrophy (EDMD) and LGMD type 1B (LGMD1B); facioscapulohumeral muscular dystrophy (FSHD); and mitochondrial myopathies, including Friedreich ataxia (FA) and Kearns-Sayre syndrome. It is noted that the 229th European Neuromuscular Centre (ENMC) workshop[1] has suggested a reclassification and revised nomenclature for LGMD, in which autosomal dominant type 1 LGMD is classified as specific myopathies or as LGMD D (dominant) variants (D1–D4) and autosomal recessive LGMD2 is renamed LGMD R1–R24 in addition to descriptive names for some of the recessive variants. In this revised nomenclature, *LMNA*-associated myopathies (LGMD1B, Emery-Dreifuss muscular dystrophy type 2 [EDMD2], and Emery-Dreifuss muscular dystrophy type 3 [EDMD3]) are named EDMD. In this document, we have retained the LGMD1B and EDMD naming convention. Each section covers general concepts specific to that disorder followed by condition-specific recommendations. These sections are further categorized into diagnostic testing and risk stratification, bradycardias/conduction disease and the use of pacing and cardiac resynchronization therapy (CRT), atrial arrhythmias, and ventricular arrhythmias/sudden cardiac death and the use of implantable cardioverter-defibrillators (ICDs). A framework and recommendations for end-of-life management of arrhythmias in patients with NMDs are also covered.

Where possible, recommendations put forth in this document are based on published evidence with the understanding that NMDs are rare and the referenced evidence base is largely observational. Studies using small sample sizes and larger randomized controlled trials (RCTs) marked by underrepresentation of patients with NMDs were frequently encountered throughout this document's development. Although summaries of clinical conditions are presented, this document is not a comprehensive review; rather, it serves to provide practical and actionable clinical information and management recommendations, with the goal of improving overall patient care. As with many guideline statements, this document is designed to help guide shared decision-making with the individual patient and is not intended to dictate management.

Throughout this document, the term "clinical status" is used to refer to a patient's overall level of functioning and may include objective and subjective measures of ambulatory capability, level of respiratory impairment, and degree of frailty, as examples.[2] This is a particularly germane concept, as the noncardiac consequences of NMDs may dominate a patient's clinical picture and prognosis, influencing clinical decision-making. Clinical status also encompasses a patient's age; children, in particular, require special consideration. Many recommendations, particularly those involving ICD implantation and anticoagulation for atrial arrhythmias, are based on research conducted in adult individuals. Although applying these studies in the care of children may be reasonable in some circumstances, extrapolating such evidence in all children is not appropriate and calls for clinician discretion and careful discussion. The recommendations should therefore be tempered by the clinician's judgment regarding the expected impact and appropriateness of a particular intervention while taking into account mitigating factors that may affect its benefit.

This consensus document provides recommendations for care of these complex patients based on current evidence for best practice in the assessment and management of arrhythmia risk in patients with NMDs. When evidence was lacking or contradictory, a consensus expert opinion was developed. For both adult and pediatric patients, the health benefits, side effects, and risks were comprehensively considered in formulating the recommendations. The document is intended to provide practical guidance and advice for management and is expected to improve the quality of care. Adherence to recommendations can be enhanced by patient engagement and a shared decision-making process. Recommendations are not a replacement for clinical judgment and are not intended to dictate management.

1.2. Organization of the writing committee

The writing committee consisted of internationally recognized experts from 12 countries in the fields of cardiac electrophysiology, cardiology, pediatric cardiology, neurology, physical medicine and rehabilitation, congestive heart failure, and anesthesiology, representing AANEM, AAPM&R, ACC, AHA, APHRS, ASA, CNS, EHRA, HFSA, HRS, JHRS, LAHRS, PACES, and SOBRAC, and selected according to each society's procedures. The HRS strives to ensure diversity in formation of the writing group. Disclosure of any relationships with industry and other entities (RWIs) was required from the writing committee members (Appendix 1) and from the peer reviewers (Appendix 2), in accordance with the HRS policies; of the 38 committee members, 20 (53%) had no relevant RWIs. Sections with