Commentary

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The article "Cardiac evaluation in patients with neuromuscular diseases" deals with an important issue in specialist cardiology practice. Individuals with neuromuscular diseases are a heterogeneous group of patients with various clinical presentation in terms of both neurological symptoms and potential cardiovascular complications.

While the knowledge on specific neuromuscular disorders is well established among the neurologists, in the cardiology practice these patients may be collectively referred to as "patients with dystrophy", without detailed differentiation between different types. Obviously, this may be sometimes justified and explained by the organization of health care, as the duration of the diagnostic workup from the suspicion of neuromuscular disease to the genetic diagnosis is usually long. The authors briefly present the most important aspects of cardiac investigations and therapeutic possibilities in the most important and most common neuromuscular diseases, including Duchenne and Becker muscular dystrophies, Emery-Dreifuss muscular dystrophy, and myotonic dystrophy type 1 (Steinert disease) and type 2.

Cardiac involvement in myotonic dystrophies deserves a more detailed discussion. Indeed, the most dangerous consequences of cardiac involvement in myotonic dystrophy type 1 include advanced atrioventricular conduction disturbances, with or without prior first degree atrioventricular block present for many years. It should be stressed that sudden cardiac death in patients with myotonic dystrophy type 1 is most commonly associated with advanced or complete atrioventricular block. In contrast, tachyarrhythmias are less frequent in Steinert disease but may also contribute to poor prognosis. Established risk factors for sudden cardiac death in this population should be borne in mine, *i.e.*, non-sinus rhythm (particularly atrial fibrillation), PR interval prolongation > 240 ms, QRS duration > 120 ms, intermittent second- and third-degree atrioventricular block, and corrected QT interval > 450 ms. A history of bradycardia-induced ventricular fibrillation is also considered an indication for prompt cardioverter-defibrillator implantation in some countries, especially in patients with left ventricular systolic dysfunction.

As indicated by the authors, the risk of cardiac involvement in myotonic dystrophy type 2 is lower compared to type 1. Our experience indicates that atrioventricular and interventricular conduction disturbances are much less common, while supraventricular and ventricular arrhythmia is significantly more common in type 2 (except for atrial fibrillation which often accompanies myotonic dystrophy type 1, as also reported by others). For a proper understanding of cardiovascular complications of myotonic dystrophy type 2, it should be remembered that this disorder often manifests later in life. Hence, cardiac involvement related to the underlying neuromuscular disorder may coexist with pathologies resulting from normal aging and concomitant conditions such as diabetes type 2 which is typical for this type of dystrophy, thyroid disorders, or hypertension which is ubiquitous in later years of life.

The value of the article also stems from the discussion of the practical aspects of the proposed cardiac investigation, and a figure that clearly presents suggested cardiac investigations following the diagnosis of neuromuscular disease. It should be noted that a patient with neuromuscular disorder should have a good quality 12-lead electrocardiogram (ECG) recorded during each follow-up visit, with careful evaluation of all necessary parameters including corrected QT interval. Unfortunately, obtaining a good quality tracing is not always easy, and some patients are reluctant to undergo ECG recording due to their limited mobility. Patients with neuromuscular disorders are usually regularly followed-up by neurologists, while cardiac evaluation is usually performed after the genetic diagnosis is established, and only occasionally afterwards. It is thus important, as also highlighted in the commented article, that these patients should also undergo regular cardiological controls, optimally in specialized tertiary care centres cooperating with neurology experts.

References

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