

Title: Recommendations of an expert group for cardiac assessment of non-dystrophic myotonic adult patients treated with mexiletine

Authors: S. Vicart¹, K. Wahbi², J. Duchateau³, JM. Sellal⁴, JC. Deharo⁵, G. Bassez⁶, E. Salort-Campana⁷, F. Labombarda⁸.

Affiliations:

¹Assistance Publique-Hôpitaux de Paris, Sorbonne Université, INSERM, Service of Neuro-Myology, Muscle Channelopathies Reference Center and UMR 974, Institute of Myology, University Hospital Pitié-Salpêtrière, Paris, France

²Centre de référence constitutif des maladies neuromusculaires, département de cardiologie, Ap-Hp Cochin, Paris, France

³Service de cardiologie, électrophysiologie et stimulation cardiaque, Hôpital Haut Lévêque, CHU de Bordeaux, France

⁴Département de cardiologie, CHU de Nancy, France

⁵Département de cardiologie, Hôpital de la Timone, CHU de Marseille, France

⁶Centre de référence constitutif des maladies neuromusculaires, service de neuro-myologie, Ap-Hp Pitié-Salpêtrière – Paris, France

⁷Centre de référence neuromusculaire coordonnateur PACA Réunion Rhône alpes, service du Pr Attarian, Hôpital de la Timone, CHU de Marseille, France

⁸Département de cardiologie, CHU de Caen, France

Abstract topic: Muscle and neuromuscular junction disorder

Introduction

Mexiletine (NaMuscla™) is indicated for the symptomatic treatment of myotonia in adults with non-dystrophic myotonia (NDM). A cardiac assessment is required as mexiletine may have a pro-arrhythmic effect. Long-term safety data supporting use of mexiletine in patients with NDM combined with the extensive clinical experience of an expert group resulted in creation of an algorithm for cardiac monitoring of NDM patients treated with Mexiletine.

Methods

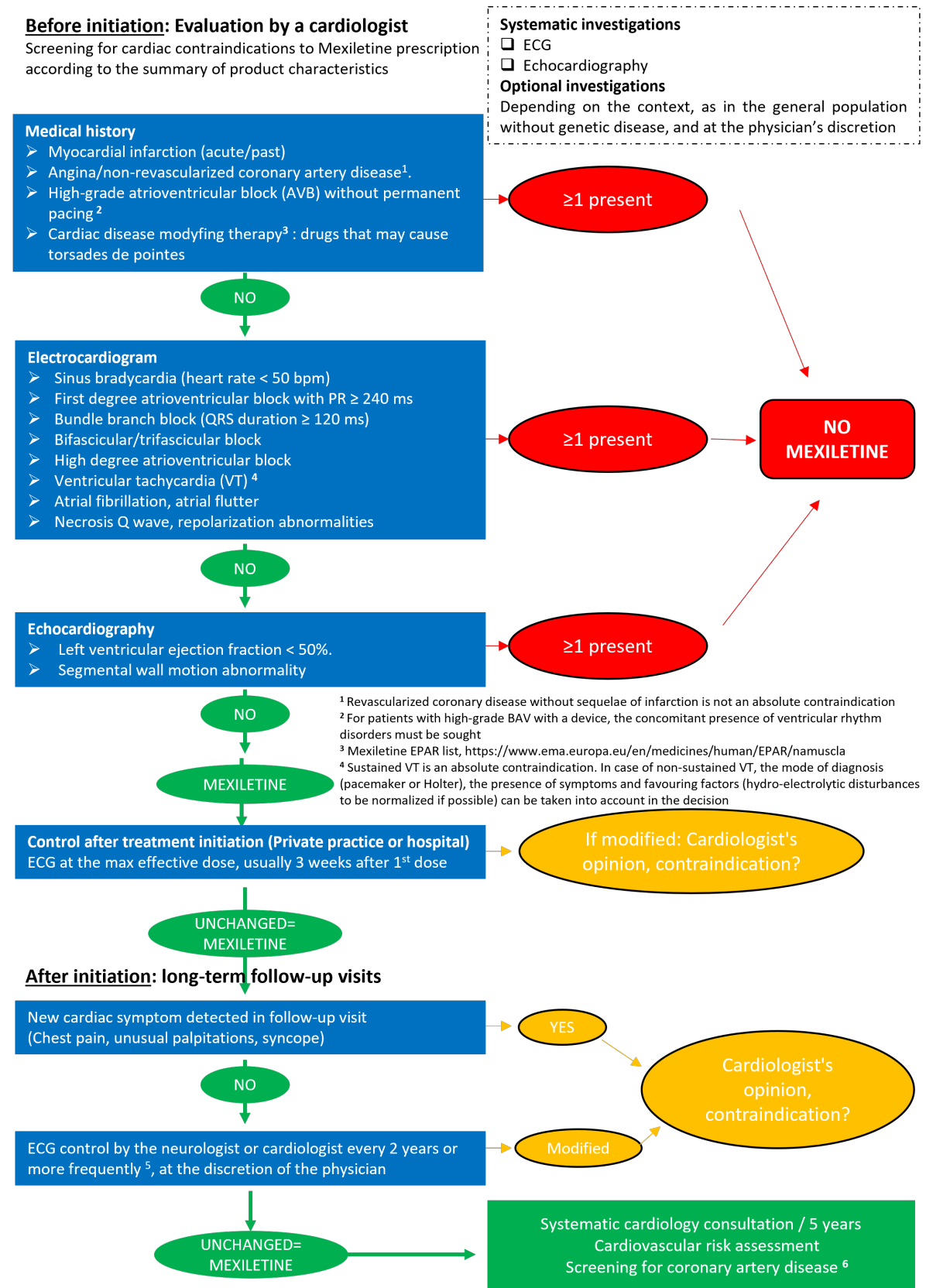
To define the treatment algorithm, several workshops with experts including 3 neurologists and 5 cardiologists from different French neuromuscular reference centres were set up. These workshops aimed to define the screening and surveillance tools required to avoid cardiac events in mexiletine-treated patients.

The recommendations are based on the summary of product characteristics (SmPC), a review of the literature on the safety of mexiletine-treated NDM patients and on the expertise of the authors.

Results

The expert group concluded that the cardiac safety profile of mexiletine in NDM patients appears similar to that of the general population. Therefore, NDM patients treated with Mexiletine should be monitored as any patient treated with a class 1b anti-arrhythmic. Cardiac assessment should be performed before initiation of mexiletine and at least every 2 years under treatment (Figure 1).

Figure 1: Mexiletine prescription algorithm in patients with NDM



¹ Revascularized coronary disease without sequelae of infarction is not an absolute contraindication
² For patients with high-grade BAV with a device, the concomitant presence of ventricular rhythm disorders must be sought
³ Mexiletine EPAR list, <https://www.ema.europa.eu/en/medicines/human/EPAR/namuscla>
⁴ Sustained VT is an absolute contraindication. In case of non-sustained VT, the mode of diagnosis (pacemaker or Holter), the presence of symptoms and favouring factors (hydro-electrolytic disturbances to be normalized if possible) can be taken into account in the decision

⁵ Mexiletine multi-disciplinary team meeting (MDT) recommends ECG + Echocardiography every year in case of known cardiac abnormality or more if deemed necessary; ⁶ according to the recommendations of the European Society of Cardiology

Conclusion

An algorithm for cardiac safety monitoring in patients with NDM treated with mexiletine has been developed to assist the neurologists and cardiologists managing these patients.

Disclosures:

All authors declare consulting fees from Lupin.