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Clinicians' statement regarding the use of Mexiletine in patients with Non-dystrophic Myotonia

In the NHS England nationally commissioned Highly Specialised Service for Muscle Channelopathies at the Queen Square Centre for Neuromuscular Diseases, National Hospital for Neurology and Neurosurgery we currently use mexiletine as our first line agent for the treatment of symptomatic myotonia in patients with non-dystrophic myotonia (NDM). This has been our established practice for the last 20 years and it is an effective treatment if carefully titrated. Our average dosing for patients is approximately 300-400mg per day and this is usually sufficient to improve quality of life to normal but this is reviewed on a case by case basis. In a smaller group of severely affected patients we may use up to 600-800mg per day but this is less common and can vary by season, with higher doses often only being needed in colder months.

In specific circumstances where mexiletine cannot be used we use other agents including lamotrigine but we consider it our second line treatment as its precise place in the management of NDM is not fully established. Anecdotally we have found lamotrigine can be effective and there is now published RCT evidence supporting its use. At present we are unable to compare mexiletine with lamotrigine and a head to head study needs to be done. We currently use lamotrigine when patients have failed mexiletine, cannot try it due to cardiac arrhythmias, or in women who are trying to conceive. We have found that titrating lamotrigine to a sufficient dose takes significantly longer than with mexiletine and, unlike with mexiletine, it is rarely associated with severe complications or death, making us more cautious in its use. Carbamazepine and flecainide are additional alternatives but seem to have less efficacy than mexiletine in our experience.

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