# What does end stage in neuromuscular diseases mean? Key approach-based transitions

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**Purpose of review**
To revise the definition of end stage in the setting of neuromuscular disease (NMD), to understand the implications for the patient, family and healthcare team, and to address the obstacles involved in the lack of definition.

**Recent findings**
Unlike several conditions such as cancer, kidney or liver disease, the literature reveals no clear definition or categorization for NMD. Many articles mention end stage without defining it. Many years ago an expert consensus panel defined it based on functional criteria (forced vital capacity values and hypercapnic events). Only for amyotrophic lateral sclerosis/motoneurone disease has a wider criteria been proposed. As a consequence, the management of this heterogeneous group of disorders is often fragmented compared with the well organized palliative care program for cancer patients.

**Summary**
Better end-stage NMD definitions should help to identify the goals of care, but a broad range in time and intensity of deterioration make a valid definition difficult for end-stage NMD. Respiratory care, life-prolonging therapies, and structured care planning should be seen as complementary rather than dichotomous. This article emphasized the relevance of an integrated approach through the whole trajectories of NMD patients considering key transitions.

**Keywords**
ALS/MND, end of life, end stage, neuromuscular diseases, palliative approach

## INTRODUCTION

A progressive neuromuscular disorder (NMD) and diseases encompass a broad range of illness that impairs the function of the muscles and/or nerves. This is a very heterogeneous group of diseases that have in common the development of respiratory muscle weakness and hypercapnic respiratory failure. However, the time between symptoms onset to impaired lung function and respiratory clinic manifestations is extremely variable: intervals ranging from months to decades and significant clinical and functional stability may occur. Acute respiratory failure is a common cause of morbidity and mortality in several NMDs [1▪]. The prognosis for many patients afflicted by these NMD has dramatically improved over time mainly because of the advancements in medical technology especially noninvasive ventilation (NIV) and other treatments that increase the quality of life (QoL) and extend life expectancy [2▪,3▪,4▪]. The key to care of the respiratory issues in NMD is a proactive and preventive approach. Providing information about treatment options for patients and their families is essential. Special attention must be paid to the regulatory laws in each country.

In several progressive NMDs such as Duchenne muscular dystrophy (DMD) and amyotrophic lateral sclerosis/motoneurone disease (ALS/MND), cure is not expected [5▪▪]. However, much can be done to mitigate the sickness burden of patients and families at the end stage of these illnesses through the use of palliative interventions [6▪▪]. But, what does end stage mean in this heterogeneous group of diseases? Unlike several conditions such as cancer, kidney or liver disease a review of the literature reveals that no

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## KEY POINTS

- The key to care of the respiratory issues in NMD is a proactive and preventive approach.
- For unclear reasons, communications between physicians and patients about the use of NIV to palliate dyspnea frequently occurs late in the course of progressive illnesses.
- By definition, much of the care provided to ALS/MND patients is palliative in nature, which involves helping patients and families cope with the symptoms of the disease, improving QoL and functional status, and helping them make decisions about goals of care.
- Respiratory care, life-prolonging therapies, and structured care planning should be seen as complementary rather than dichotomous.
- It must be emphasized the relevance of an integrated approach through the whole trajectories of NMD patients considering key transitions.

clear definition or categorization is available for end stage of NMD.

The purpose of this review is to revise end-stage disease in the setting of NMD, to understand the implications for the patient, family and healthcare team, and to address the obstacles involved in the lack of definition. We propose to implement the concept of first and second transitions in these patients. Palliative care (PC) must be introduced earlier in the trajectory of illness, often in parallel with disease-modifying treatment [7▪▪,8▪].

## END STAGE IN NEUROMUSCULAR DISEASE: LACK OF DEFINITION

The variety of terms, definitions, and a lack of conceptual clarity, complicate the potentially implementation of life-prolonging therapies, such as artificial feeding, NIV or tracheal mechanical ventilation (TMV). These inaccuracies make it harder to focus on symptom relief, patient education about the chronic conditions, and to involve patients and families in decision making as well.

Respiratory physicians are often asked to advise on end-stage management of patients with NMD affecting the respiratory muscles, as the vast majority dies of respiratory complications. For unclear reasons, communications between physicians and patients about the use of NIV to palliate dyspnea frequently occurs late in the course of progressive illnesses [9▪]. PC physicians, whose research is focused on end-of-life decision-making and the management of symptoms like pain and dyspnea in other patient populations, have a common agenda with the neurologists, pulmonologists, and other members of the core NMD team [10].

It is interesting to observe the definition and evolution of the concept of end-stage disease and related terms in the MEDLINE/PubMed article database. The tool Medical Subject Headings (MeSH) was introduced in 1960. It is a comprehensive controlled vocabulary for the purpose of indexing journal articles and books in the life sciences; it serves as a thesaurus that facilitates searching. Table 1 shows various concepts related to end stage, definitions, and the year they were incorporated.

It should be noted that when searching for *end stage*, it returns only related to *liver* and *kidney diseases*. There is not MeSH terms *EoL* and *dying*. Furthermore, *end-stage MND* is not in MeSH. Also, The Palliative Care Handbook Advice on clinical Management, at miscellaneous problems describes end-stage conditions: end-stage heart failure, end-stage kidney failure, and end-stage chronic obstructive pulmonary disorder (COPD). Moreover, the last American Thoracic Society (ATS) Consensus Statement, point out goals of EoL care for Duchenne Muscular Dystrophy patients without any mention about end-stage disease. [11,12].

**Table 1.** MeSH terms related to end-stage disease

| MeSH term | Definition | Year introduced |
|---|---|---|
| Kidney failure, chronic | It is characterized by the severe irreversible kidney damage (as measured by the level of proteinuria) and the reduction in glomerular filtration rate to less than 15 ml/min (Kidney Foundation: Kidney Disease Outcome Quality Initiative, 2002). These patients generally require hemodialysis or kidney transplantation | 1966 |
| Terminal care | Medical and nursing care of patients in the terminal stage of an illness | 1968 |
| Terminally ill | Persons with an incurable or irreversible illness at the end stage that will result in death within a short time. (From O'Leary *et al.*, Lexikon: Dictionary of Healthcare Terms, Organizations, and Acronyms for the Era of Reform, 1994,p. 780) | 1997 |
| End-stage liver disease | Final stage of a liver disease when the liver failure is irreversible and liver transplantation is needed | 2011 |

By definition, much of the care provided to ALS/MND patients is *palliative* in nature, which involves helping patients and families cope with the symptoms of the disease, improving QoL and functional status, and helping them make decisions about goals of care [10]. However, many patients continue to suffer from uncontrolled symptoms and struggle to make decisions about life-prolonging therapies. Although research has provided new options for treatment of some symptoms, others, such as ALS-related pain, remain poorly understood.

Conversely, Bach *et al.* claim that many articles continue to be published on PC for ALS/MND, with none thus far referring to prolonging life by continuous NIV. Because about 40% of patients with ALS/MND can survive using continuous NIV for a mean of [11] months, PC precepts are inappropriate for properly equipped and trained patients with adequate personal care before they meet the criteria for tracheostomy. For most non-ALS/NMD diagnoses, PC interventions are distinctly inappropriate because oxygen therapy and narcotics hasten death. Thus, palliative respiratory care for patients with NMD perpetuates the misconception that NIV is only for symptom relief and not for continuous NIV to prolong survival [6▪▪].

It must be emphasized that nowadays it is anachronistic to ignore the beneficial effects of NIV on survival. This misconception may arise from the fact that there is a dichotomous thinking of palliative interventions and prolong survival devices. Even more, many patients with NMD can live many years with NIV support, even under full setting NIV [13]. It is intuitively difficult to accept in these conditions the label of end-stage. Logically, the patient is not terminal, but the disease is [14].

Little information has been published about the use of NIV in the care of ALS/MND patients with end-stage respiratory muscle failure. There continue to be great variations by country, economics, physician's interest and experience, local concepts of palliation, hospice requirements, and resources available for home care. However, authors demonstrated the feasibility of NIV management to prolong survival, optimize wellness and the chance to die peacefully and even at home [13]. With patients' proper selection and follow-up by trained professionals, most of ALS/MND patients should be successfully treated with NIV for years. As relevant as the proper use of NIV is the airway clearance techniques, including both manual and mechanical-assisted cough, and secretions mobilizations techniques. These procedures should always be included in the treatment of NMD patients [14,6▪▪,15].

How can we help patients with ALS/MND and their families make decisions about life-prolonging interventions? NIV has been shown in many studies to improve life expectancy and QoL in patients with a forced vital capacity (FVC) of less than 50%. Patients who use NIV live an average of several months longer than those who do not. In addition, they may have less dyspnea, daytime fatigue, and insomnia. These benefits may persist even as the disease continues to progress. Despite benefits in both quality and quantity of life, many patients do not use NIV. One study indicated that only about 9% of patients with predicted FVC of less than 40% were using NIV support. Although these numbers increased in the years after the publication of the ALS practice parameter in 1999 (from 9 to 21%), use of NIV remains far below recommended levels. The reasons for this are unclear; however, in the ALS CARE study group report, about half of patients with an FVC of less than 40% who were not using NIV were offered this intervention but refused or did not tolerate it [10].

In ALS/MND patients, the use of NIV was described as beneficial and was not perceived by carers or most professionals to have adversely impacted patient's EoL experience [16]. A study highlights variation in patient wishes regarding usage toward the EoL, uncertainty regarding appropriate management among professionals, and the importance of disseminating EoL wishes. There is little information in the literature about enabling ALS/MND patients to make choices about their EoL care, particularly relating to the preferred place of death. EoL care should be discussed early and throughout the disease trajectory as an integral part of holistic care [17▪].

## AVAILABLE END-STAGE DEFINITIONS FOR NEUROMUSCULAR DISEASE

The European Respiratory Society (ERS) Task Force on ethics and decision-making in end-stage lung disease has defined the end-stage 'restrictive' respiratory patient (Table 2) [18].

Using these definitions, PC includes EoL care, but is broader and also includes care focused on improving QoL and minimizing symptoms before the EoL period [19].

The ERS survey has clearly shown that the large majority of patients with end-stage chronic respiratory disorders is treated by pulmonologists in those specialized areas [18]. NIV can be considered, in part, a palliative treatment, as it reduces the respiratory symptomatology associated with NMD and improves QoL. Although NIV is a widely accepted therapeutic option, there is controversy with respect

**Table 2.** Definition of end -stage restrictive respiratory patient according to the European Respiratory Society Task Force [18]

| |
|---|
| Main criteria |
| A patient with FVC less than 0.6 l and/or at least one admission for hypercapnic respiratory failure |
| Additional criteria |
| The need for assistance with at least one instrumental activity of daily living (e.g., housework or shopping), to improve the prognostication with respect to life expectancy |

to TMV, which improves survival but does not change the progression of the disease. This ambivalence, especially in the case of rapidly progressive diseases, is because of the possibility that undesirable situations will arise, such as 'locked in' syndrome, not to mention the huge burden for both the family and healthcare system [20]. Discussing in advance a treatment plan should be a standard of care in these patients, in particular those diagnosed with type I spinal muscular atrophy and ALS/MND, who are the most fragile subjects [19].

It is important to stress that hypercapnic ventilatory failure is not necessarily a terminal event in patients with some forms of NMD. Patients with DMD or Becker disease could have a FVC below 0.61 and remain normocapnics without using NIV for years (although probably they will require cough assistance). We believe that these examples in the context of slow trajectories do not usually seem to be end stage. Furthermore, using cough assistance, nocturnal NIV or full setting NIV, these patients and their families have needs of palliative interventions avoiding dichotomous thinking. Actually, besides the criteria proposed in Table 2, there is another approach for end-stage diseases. Table 3 shows the Medicare Hospice Entry Criteria for patients with ALS/MND. Patients must meet at least one of the following criteria [21]. Note that this approach includes many other domains rather than a value of FVC and hypercapnic event [21].

## Key phases at the end of lifr care

Judging prognosis is particularly difficult for non-cancer patients. Identification of people with a life-limiting illness when they are starting to need a change in their goals of care contributes to EoL care planning and can aid communication with patients

**Table 3.** Medicare Hospice Entry Criteria for Patients with ALS/MND [21]

| | |
|---|---|
| Patients must meet at least one of the following criteria: | |
| Criteria 1: Critically impaired breathing capacity as demonstrated by all of the following | Forced vital capacity less than 30% of normal |
| | Dyspnea at rest |
| | Patient declines mechanical ventilation |
| Criteria 2A: Patient should demonstrate both rapid progression of ALS/NMD and critical nutritional impairment | Rapid progression: independent ambulation to wheelchair or bed-bound status |
| | Progression from normal to barely intelligible speech |
| | Progression from normal to pureed diet |
| | Needing major assistance by caretaker in all activities of daily living |
| Criteria 2B: Critical nutritional impairment as demonstrated by all of the following | Oral intake of nutrients and fluids insufficient to maintain life |
| | Continuing weight loss |
| | Dehydration or hypovolemia |
| | Absence of artificial feeding methods sufficient to sustain life but not for relieving hunger |
| Criteria 3: Patient should demonstrate both rapid progression of ALS/NMD and at least 1 life-threatening complication | Recurrent aspiration pneumonia |
| | Decubitus ulcers |
| | Recurrent fever after antibiotics |
| | Inability to maintain sufficient fluid and caloric intake with 10% weight loss during past 6 months or serum albumin lower than 2.5 g/dl |

ALS/NMD, amyotrophic lateral sclerosis/motoneurone disease.

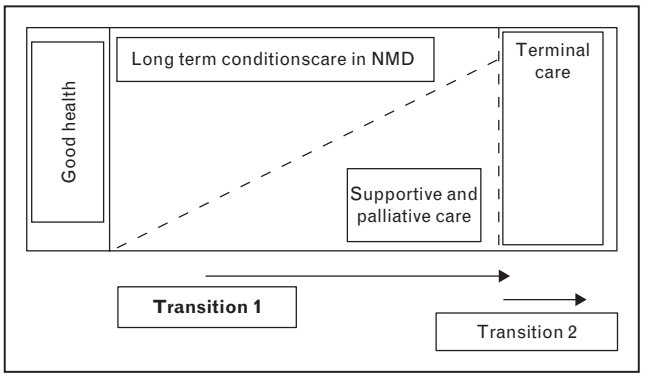


**FIGURE 1.** Phases of illness introducing first and second transitions and structured care planning (modified from [7▪▪]).

and families. It depends on clinical judgment and weighing up a complex mix of disease, clinical findings, therapeutic response, comorbidities, psychosocial factors, and rate of decline. Prognostic paralysis may delay a change in gear for too long. Being alert to the possibility that a patient might benefit from supportive and PC is central to delivering better EoL care [7▪▪].

EoL care encompasses three overlapping phases of illness (Fig. 1). Boyd and Murray offer guidance about recognizing EoL transitions. They also consider the challenge of changing the goals of care in patients with slowly progressive or fluctuating long-term conditions [7▪▪,22]. The first steps to design and implement a program to improve PC for patients with chronic conditions with a public health and population-based approach are to identify these patients and to assess their prevalence in the healthcare system.

## Transition 1: would my patient benefit from supportive and palliative care?

Managing the transition to supportive and PC is arguably more of a challenge than identifying people who are in the last days of life. Doing so earlier can affect how, and potentially where, people die, but what constitutes 'EoL care' is not uniformly understood and opinions vary as to who is a 'palliative care' patient.

Disease-specific prognostic tools use statistical models to predict the risks of individuals dying from conditions such as heart failure, COPD, or liver disease. Prognostic models were not found to be specific or sensitive enough when used to estimate survival of 6 months or less in older people with a range of noncancerous illnesses. Such survival data have limited meaning for individual patients who are 'sick enough to die' [7▪▪].

An alternative to prognostic tools is the use of criteria based on the clinical features of different advanced illnesses. Gómez-Batiste *et al.* [23] describes conceptual innovations in PC epidemiology and the methods to identify patients in need of PC, in all settings. Concerns about deciding which of these patients should have additional assessment

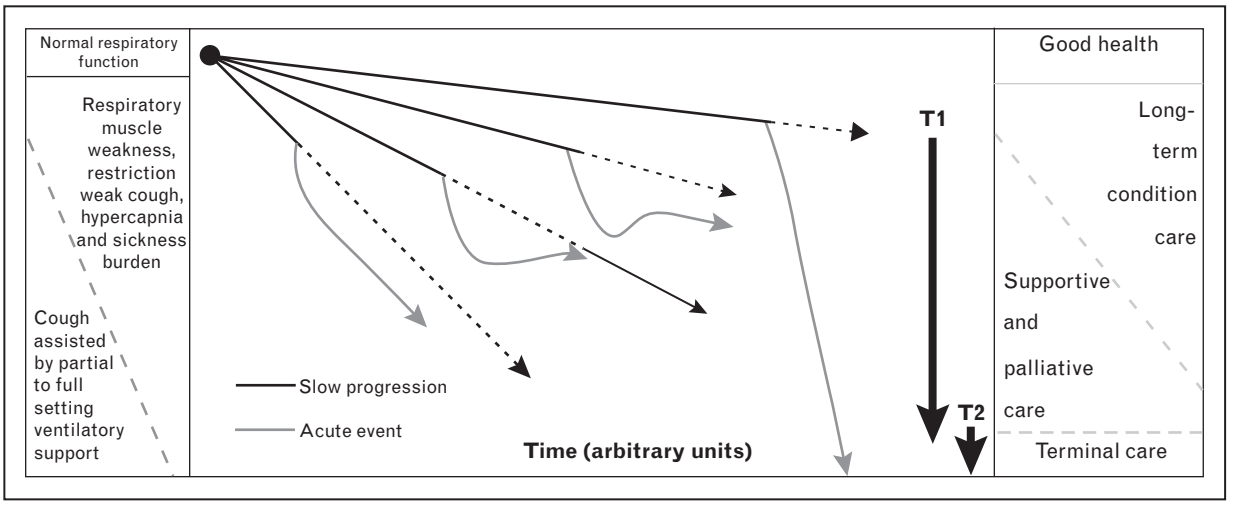


**FIGURE 2.** Schematic representation of trajectories in most of progressive NMD with respiratory impairment framed into the key transitions approach and structured care planning (original from Tripodoro and De Vito). The central panel shows the trajectories to the end-stage neuromuscular disease. Straight lines (black) represent the slow and progressive trajectory to hypercapnic ventilatory failure. During these slow trajectories, acute event (usually precipitated by infections pneumonia, aspiration and surgery) can occur at any time. Once the event overcame, the recovery of respiratory function may be total or partial and prolonged ventilatory assistance may results. The left panel represents some main events that lead to the end-stage respiratory disease and the most relevant respiratory support interventions. The right panel shows the structured care planning. In this connection, the vertical downward arrows represent the first and second transition of care. Patients can benefit by identifying such transitions (see the similarities with the Figure 1).

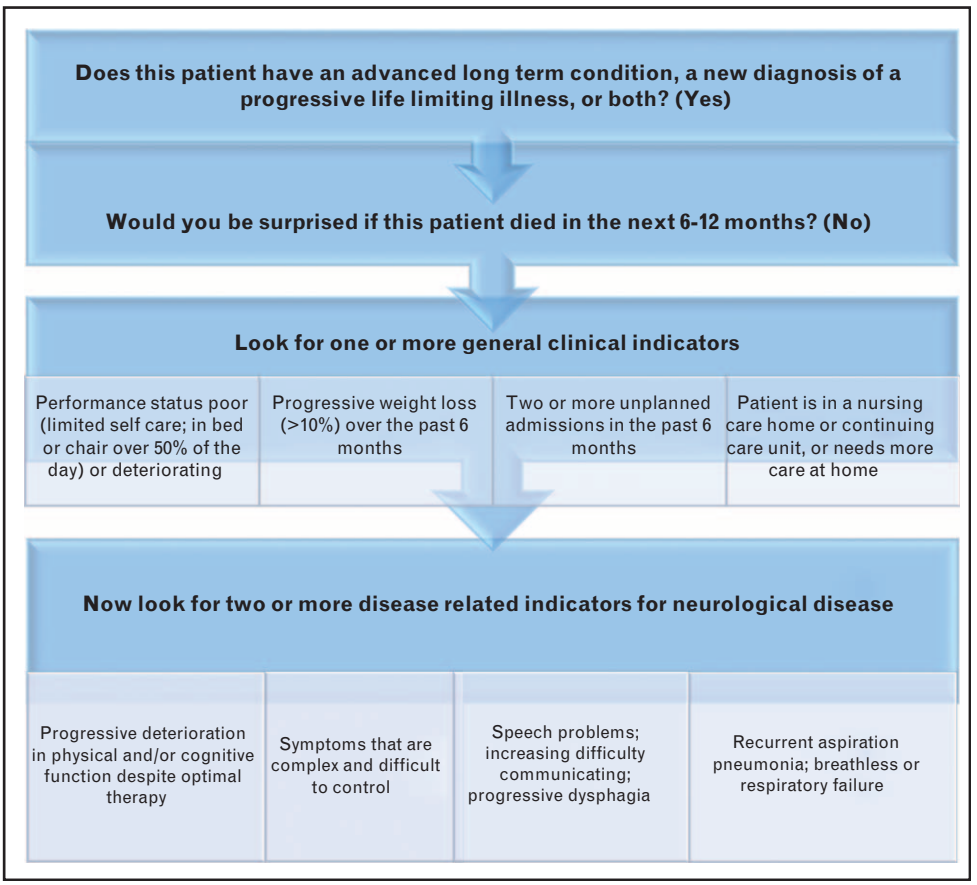


**FIGURE 3.** GRAPHIC 1. Indicators for a supportive and palliative care assessment for neurological diseases (ALS/MND, multiple sclerosis, and Parkinson's disease) [7▪▪].

and structured EoL care planning is common, as are worries about discussing dying 'too soon'. Boyd and Murray suggest that a shortlist of disease-related clinical indicators drawn from prognostic models and existing palliative prognostic guides be used to support clinical decision making [7▪▪]. Figure 2 describes how to identify patients for a supportive and PC assessment since the first transition (ALS/MND, multiple sclerosis, and Parkinson's disease) [7▪▪].

An NMD patient whose illness is associated with acute exacerbations followed by partial recovery may have been receiving health and social care for some time with the emphasis on optimal disease management, personalized care planning, and supported self-management. Variables identified in disease-specific prognostic models are particularly useful as additional indicators in these patients [7▪▪]. The care of patients with ALS/MND is complex with evidence suggesting it is best done by a multidisciplinary team led by a neuromuscular-trained ALS/MND specialist. However, as disease burden increases, access to these specialty clinics may become difficult or impossible for the patient. Extending the current multidisciplinary team paradigm to include collaborations with local providers, including local PC physicians and allied healthcare providers, can assure that the patient's needs are met throughout the disease course, particularly during terminal illness [24▪▪].

## Transition 2: Is my patient reaching the last days of life?

Diagnosis of dying can be problematic for a range of reasons including a lack of continuity of care in the community and in hospitals. In the community, anticipatory care planning should ensure that sufficient care and support are in place to enable most patients who are expected to die soon to remain at home or in their care home. However, any potentially reversible causes of deterioration must be excluded in a patient who might still benefit from appropriate treatment. Such treatment should be started on the basis of clear, agreed goals, including a plan for review [7▪▪]. To improve the transition to terminal care, the care team should ask if a patient's deterioration was expected, find out if the patient or a healthcare proxy wants further interventions, and exclude all potentially reversible causes.

### Neuromuscular disease key transitions-based approach

The so-called natural evolution of respiratory impairment in NMD should be seen as both chronic and acute trajectories. During a slow and progressive evolution to respiratory failure with probable use of partial NIV to full setting ventilatory support, an acute event (usually precipitated by infections pneumonia, aspiration and surgery) can occur at any time. Once the event has been overcome, the recovery of respiratory function may be total or partial but prolonged ventilatory assistance may results (Fig. 3). Respiratory care, life prolonging therapies (left panel in Fig. 3) and structured care planning (right panel Fig. 3) should be seen as complementary rather than dichotomous.

## CONCLUSION

Many articles mentioned end stage without defining it [18]. Many years ago, an expert consensus panel defined it based on functional criteria (FVC values and hypercapnic events). Only for ALS/MND, wider criteria have been proposed.

A broad range in time and intensity of deterioration makes difficult a valid definition for end-stage NMD. Beside these difficulties, patients may benefit by the proper identification of key transitions of illnesses, multidisciplinary team work, communication, advanced care planning, symptom relief, and life-prolonging therapies.

This article emphasized the relevance of an integrated approach through the whole trajectories of NMD patients considering key transitions. Respiratory care, life-prolonging therapies, and structured care planning should be seen as complementary rather than dichotomous.

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### Conflicts of interest

*There are no conflicts of interest.*

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- ■■ of outstanding interest

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