

Palliative care and advance care planning in Myotonic dystrophy type 1

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BACKGROUND

The creation of an 'emergency care plan' is recommended for all patients with complex neuromuscular disorders. These provide concise, relevant and rapidly accessible clinical recommendations for emergency use, including 'escalation plans', and ceilings of care whilst considering and engaging the patient's views and wishes. In contrast, 'advance care plans' (ACPs) focus more specifically on symptom control and 'end of life' decisions and are usually produced with involvement of the palliative care team. Such discussions may be more difficult to have with patients due to their complexity.

A 'traffic light' system relevant to patients with neuromuscular disease, the 'Oswestry model', has been created and subsequently adapted for DM1. This categorises patients as red, amber, or green, according to the likely palliative care needs at each stage of disease. Use of this tool has been piloted in four geographically varied neuromuscular centres in the UK, highlighting the need for identifying patients for discussion around ACP and 'symptom control' clinic. Those categorised as 'amber' in the traffic light system requires consideration for discussion' and those who were 'red' prompt referral to a palliative care clinic.

We report on the last five years of deaths from four neuromuscular centres in the UK.

AIMS/OBJECTIVES

To understand whether the 'Oswestry Traffic Light model (DM1) adapted' successfully highlights patients with DM1 nearing the end of their life, who may then benefit from palliative care and advance care planning.

To see whether this method of categorisation leads to more palliative care input and advance care planning for patients flagged as 'amber' or 'red'.

METHODOLOGY

Table 1 Oswestry traffic light model (DM1) adapted

	Blue	Green	Events	Amber	Events	Red	Events
Respiratory	No respiratory support or indication for respiratory support	Indication for CPAP or first significant chest infection requiring antibiotics and early indication for NIV	Starting overnight CPAP or stable early NIV including BiPAP	Overnight NIV (tolerated or not), recurrent chest infections requiring escalation of NIV use	Prophylactic antibiotics, home supply of antibiotics or escalation of NIV use	Daytime NIV difficulty in recording peak flow. Serious chest infection that may not be able to wean from assisted ventilation	Starting NIV during day
Cardiac	Normal or mild arrhythmia such as first-degree heart block	Insertion of ICD/PPM. Mildly impaired cardiac function	Insertion of PPM/ICD and/ or initiation of early-phase cardiac failure medication	Moderately impaired cardiac function.	Stroke associated with AF. Cardiac failure medication	Severe cardiomegaly.	Mobilisation limited by cardiac failure
Locomotor	Ambulant with no falls	Ambulant with or without one stick with occasional falls.	Use of one stick and/or occasional falls	New onset of AF due to DM1	Rollator user, difficulties in transfer, occasional wheelchair user. Requires assistance to rise from the floor	Unable to self-feed, dependent for all care, unable to ambulate and may have difficulty using wheelchair	Unable to independently ambulate
GI	No symptoms of dysphagia	Occasional feeding and infrequent dysphagia	Occasional coughing and spluttering with food or liquids with modification of diet	Oral feeding with significant moderation of food consistency and frequent problems with swallowing. Indication for gastrostomy	Insertion of gastrostomy	Severe dysphagia with an indication for gastrostomy. Defunctioning gastrostomy. Consistent incontinence of bowels and bladder	Full dependence for nutrition via gastrostomy
Acute hospital admissions	No acute admissions related to DM1	Occasional admission only with early discharge	DM1-related complication requiring hospital admission	Increasing frequency of admissions with falls and/or chest infections	Possible ICU admission without prolonged weaning	Admissions for life threatening events	ICU admission with difficulty weaning
Prognosis	Condition not expected to be life-limiting in the near future	Condition expected to be life-limiting. Expected to have a period of stability, not expected to die within the next few years				You would not be surprised if this patient died in the next 12 months. Indication for significant palliative care comorbidity	

To help prioritise patients requiring advance care planning, all patients with a palliative or potentially palliative diagnosis will be categorised as red, amber, green or blue. Those in the red category should be the first to be offered discussions about care plans. The category allocated will consider the following factors, but it also depends on the overall condition of the patient. 'Events' are changes in the patient's function that may prompt discussions about care planning.

AF, atrial fibrillation; BiPAP, bilevel positive airway pressure; CPAP, continuous positive airway pressure; DM1, myotonic dystrophy type 1; ICD, implantable cardioverter-defibrillator; ICU, intensive care unit; NIV, non-invasive ventilation; PPM, permanent pacemaker.

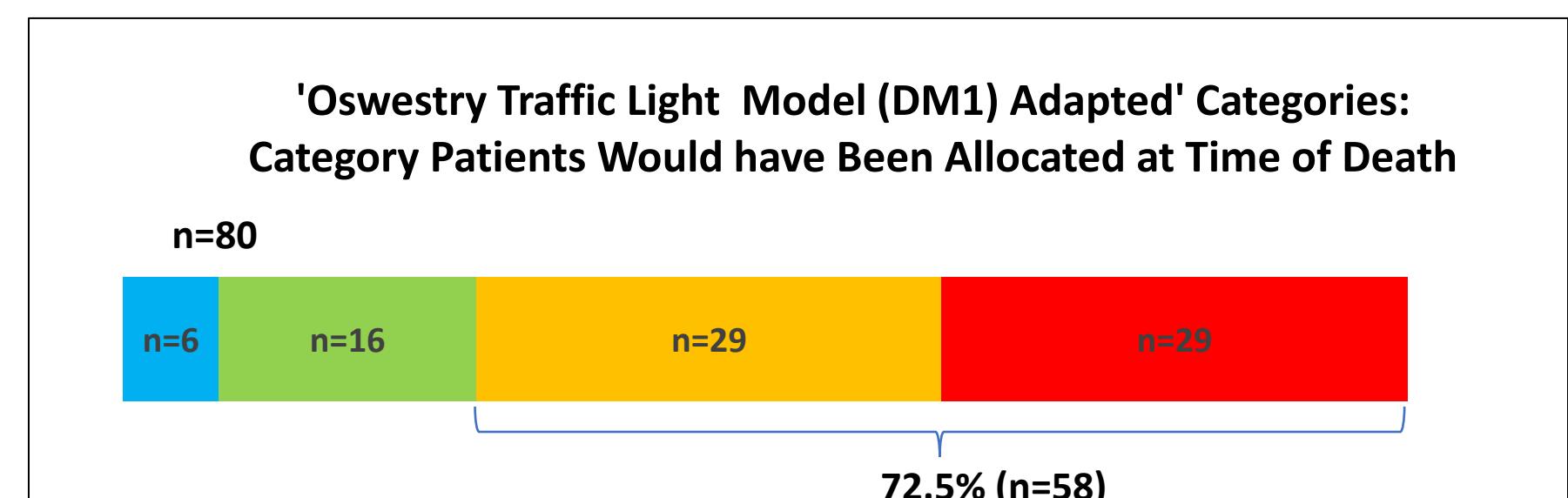
- DM1 Deaths from the last 5 years at four neuromuscular centres have been reviewed.
- Three centres retrospectively applied the 'Oswestry Traffic Light model (DM1) adapted' to patients with DM1 who had died (2020-2025).
- The fourth centre: Robert Jones and Agnes Hunt (RJAH) Orthopaedic Centre, had prospectively used the 'Oswestry Traffic Light model (DM1) adapted' for their patients with DM1 over the last 5 years, allowing insight into whether the tool can successfully prompt action and appropriate support.



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RESULTS

81 deaths occurred across the 4 centres over 5 years, 1 data set was excluded due to lack of information to be able to apply the traffic light model.



The traffic light system successfully identified over 70% of the DM1 patients as approaching the latter stages of their life. Had the model been prospectively applied, 58 patients meeting amber or red categorisation could have been identified, prompting more advance care planning and palliative care referrals allowing better support and patient-centred care towards the end of life.

The Traffic Light System prospectively piloted at RJAH, Oswestry, had allowed allocation of these 11 patients to the traffic light categories prior to death; of these patients; 5 allocated to 'amber' were known to Palliative Care at the time of their death, 2 had documented ACP discussions – one in the 'green' category and one in the 'amber'. Average age of death in 'amber' 59.8 years. None of the patients who died in the Manchester (n=32) or Sheffield (n=4) cohorts had any documented evidence of ACPs or palliative care input – despite some of these patients' deaths being identified as expected. The London cohort (n=33) had evidence of 4 ACPs in place, 3 of these were in the 'red' category and 1 in the 'amber'; it was unknown whether any of this cohort were known to palliative care.

Average Age at death ranged from 56.7- 59.8 years. Data was limited about cause of death. However, it was observed that of the unexpected 'Green group' deaths there were recurrent features of frailty and poor self care with reports of falls, poor nutritional status, evidence of poor personal care and poor compliance. Future work will explore whether this could be another sign of poor prognosis leading to additional key indicators to influence traffic light allocation.

CONCLUSIONS

Palliative care involvement and advanced care planning is an important but often overlooked facet of the management of patients with complex neuromuscular disorders. Our results suggest that tools such as the 'Oswestry model' can be used to structure care, to prompt advance care planning and to facilitate access to appropriate palliative care services.

Further work is needed to understand whether there are adaptations needed to the 'Oswestry Traffic Light model (DM1) adapted' to more accurately stratify patients into categories that better represent their prognosis.

Future work will continue to look at the impact of the prospective use of this model for people living with DM1 to inform the ongoing development of services and support.

REFERENCES

Willis D, Kulshrestha R, Bassie C, Vithlani R, Macfarlane M, Willis T. (2021). Neuromuscular diseases and advance care plans: traffic light system. BMJ Supportive & Palliative Care, 11(1), 116. <https://doi.org/10.1136/bmjspcare-2020-002336>

Willis D, Willis T, Bassie C, Eglon G, Ashley EJ, Turner C. Myotonic dystrophy type 1: palliative care guidelines. BMJ Support Palliat Care. 2024 Jan 22:spcare-2023-004748. doi: 10.1136/spcare-2023-004748. Epub ahead of print. PMID: 38253488.