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A review of the symptom control clinic, advance care planning and mortality in patients with Myotonic Dystrophy Type 1 in a UK neuromuscular centre.

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BACKGROUND

Myotonic Dystrophy type 1 (DM1)
DM1 is the commonest muscular dystrophy seen in adults. The trinucleotide repeat disorder is caused by an expansion at the end of the DMPK gene (chromosome 19), coding for myotonic dystrophy protein kinase, predominantly expressed in skeletal muscle[1]. This multisystem disease causes cardiac and respiratory compromise, endocrine and gastrointestinal problems, cognitive difficulties, cataracts and progressive skeletal muscle weakness, wasting and myotonia. Over 75% of early mortality is caused by cardiorespiratory problems, including cardiac arrythmias and less commonly dilated cardiomyopathy. The mean age of death is still approximately 58 years.

Palliative care provision for neuromuscular patients
Palliative care in neuromuscular (NM) disorders has traditionally been centred around children's hospice care for Duchenne Muscular Dystrophy and adults with Motor Neurone Disease. During recent round table discussions to review national guidelines for myotonic dystrophy, the palliative care group reviewed the availability and access to palliative care and advance care planning. They identified palliative care services are underutilised for neuromuscular patients, particularly in DM1. There is growing awareness of the value of palliative care support for DM1 patients for improved symptom control and quality of life, amongst strong evidence for advance care planning for neuromuscular patients[5,6,7,8,9].

Neuromuscular at Oswestry (NeMO)
NeMO is a centre of excellence for neuromuscular care covering a population of approximately 6 million. We here describe our novel symptom control clinic covering part of the West Midlands and North Wales area. We published 'the Oswestry model; DM1 adapted'[2,4], following identification of a need for more guidance at the national round table guideline review for myotonic dystrophy, to enable teams to identify those patients more at risk of dying and in need of palliative care support and an advance care plan (ACP)/emergency care plan (ECP).

AIM
This work aims to describe our symptom control clinic for patients with DM1, assess if our use of the Oswestry traffic light screening tool is identifying appropriate patients, and evaluate patient outcomes of this clinic. Our ultimate objective to aim to standardise and improve quality of care for patients with DM1.

Consensus-based care recommendations for DM1 [3]

Aim to standardise and improve quality of care.

Life Threatening Symptoms- ***poor recognition of the value of palliative care.***

Risks of surgery related to potential severe adverse reactions to anaesthesia and analgesia including opioids.

Respiratory symptoms remain leading cause of mortality, weak cough, respiratory insufficiency.

Cardiac symptoms are second cause of mortality- arrthymias (sinus bradycardia, atrial flutter, ventricular tachycardia and heart block).

Key points from Ashizawa, Tetsuo et al. "Consensus-based care recommendations for adults with myotonic dystrophy type 1." *Neurology. Clinical practice* vol. 8,6 (2018)

The Oswestry Symptom Control Clinic Model

Model
- Quarterly reviews of neuromuscular patients, including DM1, with a Palliative Care Consultant, Neuromuscular (NM) Specialist Nurse and other appropriate professionals as required.
- This can be face to face, virtual or by telephone as wished by the patient.

Outcomes
- Symptom control and focus on quality of life.
- Facilitates engagement with palliative care services and access to the hospice when appropriate.
- The completion of Emergency Care Plans and Advance Care Plans.

ECP
- ECPs (Emergency Care Plans) should be considered in all patients with complex medical conditions, including DM1.
- Often the first of the two plans to be written
- Personalised, concise, relevant and rapidly accessible clinical recommendations for emergency use, including 'escalation plans', particularly for respiratory care. Not just limited to cardiopulmonary resuscitation status.

ACP
- ACPS (Advance care plans) may be prompted by disease progression or a specific situation, or are complimentary to ECPS and can be considered at the same time.
- This plan is often more detailed and may be completed by the patients and families themselves.
- They focus on ceilings of care, 'end of life' decisions and specific patient wishes.

Aspiring to deliver world class patient care

The Oswestry Model Traffic Light Tool

Patients who would benefit from the symptom control clinic are identified at NM clinics and MDT with the aid of the 'traffic light' tool (see Box 1)[2,4]. Identification is based on scoring "Red" or "Amber' on the tool, stage of illness and symptom severity. For example; identifying patients with frequent ITU admissions or a tracheostomy, to identify those who should be 'considered for discussion', and prioritising those who were symptomatic or "Red" in the traffic light system, for symptom control clinic and to discuss ECP/ACP.

Box 1: The Oswestry model; DM1 adapted'[4]

	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 oral antibiotics and early indication for NIV	Starting overnight CPAP or stable early NIV including BiPAP	Overnight NIV (tolerated or not), recurrent chest infections requiring antibiotics	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 HB	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 New onset of AF due to DM1	Stroke associated with AF. Cardiac failure medication	Severe cardiomyopathy.	Mobilisation limited by cardiac failure
Locomotor	Ambulant with no falls	Ambulant with or without 1 stick with occasional falls.	Use of 1 stick and/or occasional falls	Rollator user, difficulties in transfer, occasional wheelchair user. Requires assistance to rise from floor.	Frequent falls if mobilises	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	Orally 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 indication for gastrostomy Defunctioning ileostomy. Consistent incontinence of bowels and bladder	Full dependence for nutrition on 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 dies within the next few years and/or patient has significant palliative comorbidity.		You would not be surprised if this patient dies in the next 12 months. and/or patient has significant palliative comorbidity.	

METHOD

- A retrospective case note review of patients with DM1 in one tertiary neuromuscular centre, cared for by the service between 2017 and 2023
- Data gathered on percentage of patient caseload who accessed the symptom control clinic or palliative care services, those who died during follow up and percentage of patients with an ACP/ECP created.
- Data also collected from symptom control clinic on percentage of patients who have DM1, as opposed to another neuromuscular diagnosis.

RESULTS

Case load of DM1 patients between 2017-2023 – n=81

18.5% (n=15) of DM1 patients died during this time frame

40% (6/15 deaths) of DM1 patients who died within this time frame were involved with symptom control clinic or palliative care services

53% (n=43) of DM1 patients discussed and planned ACP/ECP

10% of all patients attending the neuromuscular symptom control clinic have DM1

CONCLUSION

- Patients with life limiting conditions, such as DM1, require a proactive approach to ensure a focus on quality of life, treatment choices and, when the time comes, a good death. Early consideration of Advance Care Planning is an essential part of this good quality palliative care.
- In this review, patients with DM1 were given opportunity and support for ECPS, ACP and symptom control.
- The symptom control issues identified and treated were found to be within the scope and competency of specialist adult palliative care units
- A "traffic light" system relevant to neuromuscular patients has been used successfully when reviewing patients in MDT to identify some of those who might benefit.
- We hope this model of care for patients with neuromuscular conditions, including DM1, has the potential to be replicated elsewhere, countrywide or internationally.

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No conflicts of interest