Duchenne muscular dystrophy; the use of the 'traffic light' system to plan discussions and Advance care planning.

Willis T[1,2], Jing Yeo[3], Katie Nevin[3], Pippa Jackson[3], Channa Hewamadduma[3,4], Willis D[5].

1.Robert Jones and Agnes Hunt hospital, Oswestry. 2.Chester University Medical School. 3.Academic Neuromuscular Unit, Sheffield Teaching Hospitals NHS Foundation. 4.Trust Sheffield Institute for Translational Neurosciences (SITRAN), University of Sheffield. 5.Severn Hospice, Shropshire.

BACKGROUND

Duchenne muscular dystrophy (DMD) is severe neuromuscular condition and continuously evolves as a person's disease progresses, and is often depicted in reaching certain 'milestones', such as 'time to lose ambulation'. Throughout the disease journey, DMD care requires input from a multidisciplinary team, ranging from primary care physicians to specialists in neuromuscular, cardiology and respiratory, physical therapists, and nursing care, among others. As the disease progresses, it has been well recognised that palliative care and planning with advance care plans (ACP), together with the men living with DMD and their families, can benefit from this MDT involvement.

These discussions have been documented as being important to both the young men and their families[1], however such discussions may be challenging to have with patients due to various factors such as disease complexity, family dynamics, neurodiversity etc. It is paramount to know when the 'right time' is and how best to approach.

Having a system within the team, that highlights whether a young person is approaching that stage and is something that should start to be discussed is important, such a trigger i.e: starting ventilation, an ITU admission.

We highlight how using a 'tool' can be helpful to the clinical team, planning their ongoing care.

AIMS/OBJECTIVES

To review the use of a 'traffic light system' (TLS) to identify patients that an 'emergency care plan' and possibly ACP should be considered, and discussions started.

To review outcomes of deaths and use of the TLS and other factors that may be helpful in determining the right time for discussions.

METHODOLOGY

A 'traffic light' system relevant to patients with neuromuscular disease, and particularly DMD, was developed and called the 'Oswestry model'[2]. This 'traffic light' system (TLS) 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 used Robert Jones and Agnes Hunt hospital, UK, for over 5 years and highlighted the need for identifying patients for discussion around ACP and 'symptom control' clinic. Those categorised as 'amber' in the Traffic light system require 'consideration for discussion' and those who were 'red' prompt referral to a palliative care clinic.

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

	Blue	Green	Events	Amber	Events	Red	Events
Respiratory		No respiratory support		Overnight NIV, significantly reduced lung function	Starting overnight NIV	Daytime NIV, unrecordable peak flow	Starting NIV during day
Cardiac		Normal cardiac function or mild cardiomyopathy		Moderate cardiomyopathy	Insertion of ICD	Severe cardiomyopathy, arrhythmias	
Locomotor		Ambulant, or wheelchair user, able to transfer	Loss of ambulation	Wheelchair user, unable to transfer		Unable to self-feed, dependent for all care	
GI		Orally feeding		Supplemental gastrostomy feeds	Insertion of gastrostomy	Dysphagia, at risk of aspiration	
Acute hospital admissions		Occasional admission only		Increasing frequency		With life threatening event	ICU admission
Prognosis	Condition not expected to be life limiting.	Expected to be life limiting, but to have a period of stability. Not expected to die within the next few years		Would you not be surprised if this patient dies within the next few years?		Would you not be surprised if this patient dies in the next 12 months?	

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RESULTS

We present the data from 2 neuromuscular centres in the UK, who have been using this system for their adult DMD population and what the outcomes have been. This includes a total of 64 adult DMD patients, their 'TLS', ACP status and the outcomes over the last 5 years including information on the DMD deaths and nature of their outcomes with or without an ACP or palliative care involvement.

Fig 2 demonstrates that the number of patients where ACP discussions had happened were seen in 100% of the 'red' and 65% 'amber' category, and referral of known to palliative care was again higher in the 'red' category at 80%, however some of these were in the referral phase and were not yet actively receiving input from palliative care.

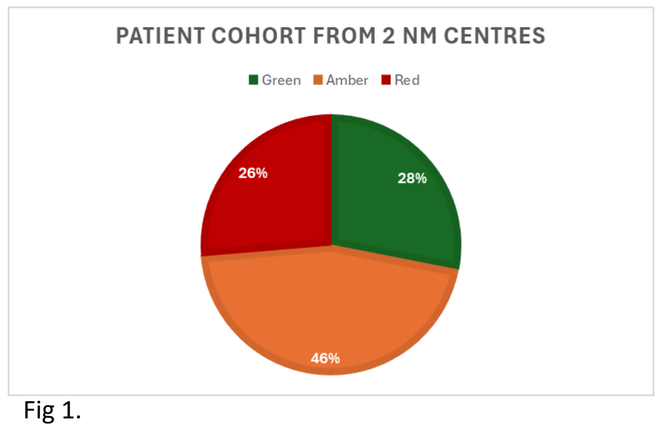

Fig 1.

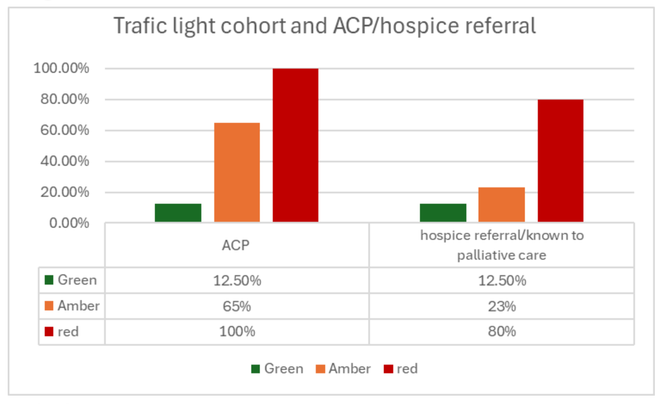

Fig 2.

Table 1 demonstrated that those that were categorised 'red' were significantly worse clinically compared to 'amber' and numbers ventilated full time and during the day were much higher. This reinforces that the right patients were being selected for discussions regarding ACP and palliative care involvement.

Function	Amber	Red
Respiratory function (mean values)	1.28L	0.39L
Ventilation nocturnal (% Cohort)	54%	6%
Ventilation Nocturnal and part daytime (% Cohort)	12%	27%
Ventilation full time (% Cohort)	0%	67%
Cardiac function (Mean LVEF)	55%	31.8%

Table 1.

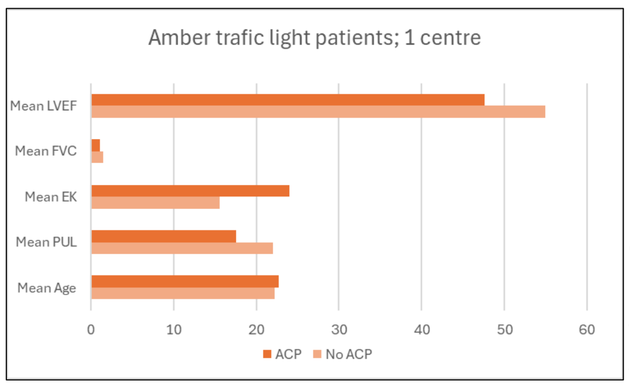

Fig 3.

In Fig 3 we were able to analyse the Outcome measures; Mean Left Ventricular Ejection fraction (LVEF), Forced vital capacity (FVC), Egen Klassification (EK) scale (reversed), Performance of Upper Limb (PUL) and age, in those that had an ACP in the 'amber 'category'. This showed a trend in those that were older, with reduced functional score and increased EK scale (reversed), with a reduced FVC and reduced cardiac function.

CONCLUSIONS

These results show that the process of TLS is effective in DMD at identifying appropriate patients at the appropriate time to start discussions regarding advance care planning and palliative care involvement. This proves that the TLS is a useful tool in the continuum of provision of high-quality care in DMD.

REFERENCES

1.Abbott, D. W. F., & Carpenter, J. S. W. (2014). 'Wasting precious time': young men with Duchenne muscular dystrophy negotiate the transition to adulthood. Disability and Society, 29(8), 1192-1205. https://doi.org/10.1080/09687599.2014.916607

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