Trends in Pediatric Palliative Care Research (TPPCR) 2021; Issue #10: Commentary on *Battista et al.*

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Abstract:

This TPPCR commentary discusses the 2021 paper by Battista et al, “Advance Directives for Adolescents and Young Adults Living with Neuromuscular Disease: An Integrative Review of the Literature.” published in Journal of Hospice and Palliative Nursing.

This commentary is a part of the Trends in Pediatric Palliative Care Research commentary series. To learn more or to sign up for our monthly newsletter visit: <https://pediatricpalliative.com/research-blog/>

The world is now home to the largest cohort of adolescents in history – 1.2 billion people between the ages of 10 and 19. Both between and within countries there is much variation in the definitions used to describe the Adolescents and Young Adults (AYA) cancer population. In the United States, the 2006 Surveillance, Epidemiology and End Results (SEER) program report used the 15–29 age range,¹ while the 2006 Adolescent and Young Adult Oncology Progress Review Group (AYAO PRG) report used 15–39²—now the standard used by the National Cancer Institute (NCI) as well as the age range generally used by JAYAO. Canada generally uses the same 15–29 age range as SEER,³ while Australia tends to favor 15–25.⁴ In the United Kingdom, the Teenage Cancer Trust focuses on teenagers and young adults (TYAs) aged 13–24,⁵ though European members of EURO CARE employ the 15–24 bracket.⁶

Editor's recommendation for the age range defining the AYA oncology population is to subdivide NCI's 15–39 age range into three age cohorts: early young adulthood (15–18 years old), young adulthood (19–24), and late young adulthood (25–39); to best account for the differential physiological and psychosocial realities experienced by AYAs within each age category.⁷ AYA comprise a distinctive demographic whose priorities of care are not always congruent with those for older adults.⁸ The issues faced by AYA patients are complex, with psychosocial, interpersonal, and developmental challenges not typically encountered by other patient populations, particularly during transition from pediatric to adult services,⁹ or with the involvement of parents/guardians in the care though many AYAs are adults in the eyes of the law.¹⁰ This article was chosen to highlight this important but often neglected aspect of consultations in AYAs.

Out of 307 studies retrieved from database search, only 5 (1.6%) were included for this review, which reflects the paucity of literature in this aspect. Four themes emerged from the literature:

1. conversations about advance directives with adolescents/young adults with neuromuscular disease are not being conducted
2. only a small number of patients have documented advance directives
3. patients want to have conversations about goals of care and want to have them sooner
4. there is a lack of evidence in this area.

These findings will influence neuromuscular clinicians' practice surrounding the use of advance directives and increase their knowledge regarding the need for discussions regarding goals of care, and some preliminary work has been done in that regard as well.¹²

Using the 'The Spectrum of Children's Palliative Care Needs' classification to identify when to introduce advance care planning, a traffic light system has been developed specifically for neuromuscular patients. This categorises patients as 'red', 'amber', 'green' or 'blue' based on cardiac, gastrointestinal, locomotor and respiratory status, recent hospital admissions and prognosis. As with The Spectrum of Children's Palliative Care Needs, the over-riding factor is that of prognosis. This asks clinicians if they would not be surprised if the patient were to die within 12 months (red category) or a few years (amber). This is a commonly used concept in palliative care and distinct from expecting the patient to die within that time frame. The system also includes events that could lead to a change in category, which may prompt advance care plan discussions.¹²

	Blue	Green	Events	Amber	Events	Red	Events
Respiratory		No 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	ICD Insertion	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		Oral feeding		Supplemental gastrostomy feeds	Gastrostomy Insertion	Dysphagia; 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.	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 significant palliative comorbidity		You would not be surprised if this patient dies in the next 12 months? And/or patient has significant palliative comorbidity.	

NIV = Non-invasive ventilation; ICD = implantable cardioverter defibrillator; ICU = intensive care unit

Figure: Neuromuscular Traffic Light Tool. NIV, non-invasive ventilation; ICD, implantable cardioverter defibrillator; ICU, intensive care unit; GI, gastrointestinal. Adapted from Willis et al. (2021).¹²

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