PALLIATIVE CARE CASE OF THE MONTH

"How to Predict Prognosis in Amyotrophic Lateral Sclerosis"

by

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Case: Mrs. K is an 80 year-old woman with newly diagnosed Amyotrophic Lateral Sclerosis (ALS). One month prior to my consultation, the patient was admitted to the hospital, diagnosed with ALS and discharged to a skilled nursing facility for physical and occupational therapy. She is now admitted with urinary retention and hyponatremia. She has diffuse weakness with inability to move her arms other than at the wrists and inability to move either leg against gravity. She attempts to mouth words but her family has not been able to understand her speech. She has dysphagia and uses a feeding tube for nutrition. We are consulted for goals of care.

On my visit, the patient was not strong enough to articulate her advance care preferences. However, her family reports she has a strong preference of quality of life over prolonging life and that she does not find her current quality of life acceptable. The family asked how long we thought Mrs. K would live.

Discussion: There are several methods to prognosticate for Mrs. K. Using **disease non-specific predictors** for prognosis such as the Palliative Performance Scale (PPS)[1] or the Karnofsky[2] score, we can use her functional status to prognosticate. Mrs. K has a Karnfosky score of 20. Her PPS is 30%. This data suggest a prognosis of days-weeks.

Given that we attributed her abrupt decline to the recent diagnosis of ALS, we wondered if we could have predicted this trajectory based on **diagnosis alone predictors.**

There is one evidence-based model for predicting prognosis in ALS.[3] The ENCALS model was developed to predict survival without tracheostomy or non-invasive ventilation for more than 23 hours per day in patients with ALS. The study authors collected data from 11,475 European patients from 1992-2016.

This model uses clinical, cognitive and genetic variables to predict survival. The clinical predictor variables include diagnostic details and functional status: site of onset (spinal v. bulbar), age at onset of weakness or bulbar symptoms, time from onset of weakness or bulbar symptoms to diagnosis, revised El Escorial criteria (definite v. probable or possible ALS), forced vital capacity (FVC), the revised ALS Functional Rating Scale (ALSFRS-R). The cognitive predictor is the presence of frontotemporal dementia, and the genetic variable is presence of the C9orf72 mutation (one of the most common genetic mutations found in ALS).

This model does not include treatment with riluzole, enteral nutrition or ventilation as prognostic variables. Riluzole is the only medication shown to improve survival in ALS, and its use is common (about 75% of European patients).

The authors note that the survival benefit of riluzole (HR=0.84 in a Cochrane meta-analysis; increased median survival of three months) is substantially smaller than the combined effect of the predictors included in the model (HR up to 15.29).[4]

The model can report survival outcomes without tracheostomy or non-invasive ventilation for more than 23 hours per day in three ways: as individualized prognostic estimates (specific patient compared to average patient), prognostic groups (very long, long, intermediate, short or very short), or as a point estimate within a survival curve (See Figure).

The model is available to health care providers (after registering on the website) at http://www.encalssurvivalmodel.org. The ALSFRS-R score is needed to use the ENCALS model. The ALSFRS-R scale is here: http://www.outcomes-umassmed.org/als/alsscale.aspx.

There are several caveats to the use of this specific model. First, some patients may be outliers, in which case the model will not accurately predict his/her prognosis. Second, some patients with ALS have plateau periods, which make disease-specific prognostication difficult. Third, prognosticating for a patient based on population based data and being able to communicate this estimated prognosis to an individual patient and/or a surrogate are separate skills. Although this model provides data for the former, the model's appropriate and effective use in clinical practice requires a thoughtful approach and complex communication skills. Although outside the scope of this case, much has been written about helpful methods for communicating prognosis.[5-6]

Resolution of the Case: We discussed the ENCALS model as well as the functional status predictor models with the family medicine team caring for Mrs. K and planned a family meeting with her daughter and husband. Based on the ENCALS model, Mrs. K's prognosis was about 12 months, which put her in the "very short" prognostic risk group. Unfortunately, Mrs. K's condition worsened over several days as we worked to coordinate a meeting time with her family. Unbeknownst to us, as we arranged the family meeting, Mrs. K and her family were discussing goals of care and making funeral arrangements. By the day of the family meeting, Mrs. K's breathing was labored due to suspected aspiration. At that time, the family updated us that the focus would be her comfort. She died a day later in the hospital.

Personal details in the case published have been altered to protect patient privacy.

For palliative care consultations please contact the Supportive and Palliative Care *programs at PUH/MUH, 412-647-7243, pager # 8511, Shadyside, 412-647-7243, pager # 8513, Perioperative/ Trauma Pain, 412-647-7243, pager # 7246, UPCI Cancer Pain Service, pager 412-644-1724, Magee Women's Hospital, pager 412-647-7243 pager # 8510, VA Palliative Care Program, 412-688-6178, pager # 296. Hillman Outpatient: 412-692-4724. For ethics consultations at UPMC Presbyterian-Montefiore and Children's pager 412-456-1518*

With comments about "Case of the Month" call Dr. Robert Arnold at (412) 692-4834.

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