ALS patients differ on treatment choices in later phases of disease

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Two new studies analyzing treatment decisions in late-stage amyotrophic lateral sclerosis (ALS) patients shed light onto treatments aimed to extend the duration and quality of life in this progressively debilitating neuromuscular disorder. Researchers from the Perelman School of Medicine at the University of Pennsylvania found that waiting until the last minute to receive one treatment resulted in not living long enough to experience the benefits.

In a separate study, Penn researchers uncovered polarized preferences among patients regarding the value of an expensive, marginally effective disease-modifying drug. The research will be presented at the American Academy of Neurology's 64th Annual Meeting in New Orleans.

ALS, commonly known as Lou Gehrig's disease, is rare, affecting approximately 30,000 Americans. In later stages of the disease, it paralyzes ALS patients' bodies, while minds often stay sharp. Along with other treatments and supportive therapies used in later stages of the disease, many patients receive a feeding tube to ensure nourishment can be obtained when muscles are impaired.

One Penn Medicine study demonstrates that ALS patients who have feeding tubes placed before an emergency situation strikes fare better. Those having surgeries in non-emergent settings were much less likely to die within one month after surgery, compared to ALS patients receiving their feeding tubes under duress. Median survival after the feeding tube surgery was 6 months overall and longer for patients undergoing non-

emergent versus emergent placement (7 months vs. 4 months). In addition, mortality rates were worse for patients having procedures done at hospitals that did not regularly perform feeding tubes placement in ALS patients.

"Timing is crucial for placement of feeding tubes in ALS patients," said the lead author of both studies, Amy Tsou, MD, MSc, a fellow in Neurology and a Robert Wood Johnson VA Clinical Scholar. "We've shown that waiting too long can be detrimental and happens too often. In general, it is important for clinicians and patients to proactively prepare and reevaluate treatment decisions as ALS patients enter into different phases of the disease."

In a second study, researchers found polarized treatment preferences regarding Riluzole, the first FDA approved treatment to slow ALS. Patients had sharply polarized preferences about this expensive treatment, which modestly prolongs length of life of ALS patients. In a survey of 98 patients with ALS or Motor Neuron Disease, nearly two-thirds of the patients ranked Riluzole as either the most important (30 percent) or least important (33 percent) treatment option.

"It is important to ask patients how they value their treatments, as in this case, we learned that patients who are older and looking for a high quality of life, valued this drug considerably more than people with impaired walking ability, who instead preferred supportive therapies like adaptive equipment," said Leo McCluskey, MD, professor of Neurology and director of the Penn ALS Center. "Overall, medical care providers should work with patients to discuss treatment options throughout the progression of the disease to ensure a high quality of end-of-life care."

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