On-Demand Clinical News

Management of Amyotrophic Lateral Sclerosis (ALS) in the Hospice Patient

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Disease Background

ALS, sometimes referred to as Lou Gehrig's disease, is a neurodegenerative disease that is progressive and affects nerve cells in the brain and spinal cord. ALS affects motor neurons, which control voluntary movement and muscle control. As ALS destroys these nerves, the brain loses the ability to initiate and control muscle movement, and the muscles become weaker.

The vast majority of cases (90-95% in the U.S.) are of the sporadic type, meaning it can strike anyone and a specific trigger is unknown. Only a small percentage of cases (5-10% in the U.S.) are of the Familial type and are due to an inherited gene mutation. ALS is considered to be 100% fatal, and there is currently no cure. Average age at diagnosis is 55, and average life expectancy is 2-5 years after diagnosis, although a small percentage of patients (5%) will survive 20 years or longer.

Amyotrophic

Lateral

Sclerosis

Presentation, symptoms, and rate of progression of ALS can vary among patients, although it is typically gradual in onset. Common symptoms include the progressive loss of the ability to talk, swallow, walk, move, and breathe; progressive muscle weakness and paralysis are experienced by all ALS patients. Because ALS attacks the motor neurons, muscles of the eyes and bladder are generally not affected, and the five senses (sight, touch, hearing, taste, smell) are also not affected. Up to 50% of ALS patients will have some degree of change in thinking or behavior, with approx. 25% developing "full blown dementia."

Continued on Page 2

Upcoming Lunch and Learn Presentations

September
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"Strategies for Management of Pruritus"

Presenter: Kiran Hamid, RPh

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Wednesday, Oct 12, 2016 at 12:00pm ET

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Presenter: Steve Stoyanov, PharmD

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Wednesday, Nov 9, 2016 at 12:00pm ET

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Management of ALS continued from page 1

Treatment:

Riluzole (Rilutek®) is the only FDA approved medication indicated for the treatment of ALS. It can extend survival time and/or time to tracheostomy, although the survival benefit is relatively modest, with a difference in median survival of only approx. 60 to 90 days. In studies, riluzole demonstrated no statistically different difference in mortality, and did not show a benefit in measures of muscle strength and neurological function. Typical adult dosing for riluzole is 50mg PO q12 hours. The average retail price for generic riluzole is approx. $37 per tablet.

It is typically recommended to discontinue riluzole when patients are enrolled in hospice or when they become ventilator dependent, for the following reasons:

- Riluzole is a treatment intended to prolong survival and/or extend time to tracheostomy, and that survival benefit is typically only 2-3 months.
- It also has not been shown to palliate ALS-associated symptoms or improve quality of life.
- In addition, riluzole is cost-prohibitive, and has adverse effects that typically improve once the medication is discontinued (nausea and weakness are among the most common).

Symptom Management

Spasticity: Non-pharmacologic measures may include physical therapy, range of motion, and stretching, and muscle relaxants include baclofen or tizanidine. Note that both of these medications list drowsiness and weakness as possible adverse effects, and it is possible that the relief of spasticity could decrease tone and increase falls – Recommend to start with low doses of these medications and increase gradually, if needed. Both of these medications require tapering prior to discontinuation.

Hypersialorrhea (excessive saliva production): Oral/sublingual medications are used first-line, as they are typically more cost-effective and more accessible than some of the other options.

- Preferred meds include: atropine ophthalmic drops and hyoscyamine tabs or liquid, given via SL route.
- Other non-preferred agents include: glycopyrrolate, scopolamine patch, amitriptyline, trihexiphenidyl, or pseudoephedrine.
- For thickened secretions, guaifenesin, adequate hydration, and/or humidification of room air can be helpful.

Pulmonary Issues: Respiratory failure is the leading cause of death in ALS, and in the absence of any underlying intrinsic pulmonary disease, this failure is purely mechanical, with respiratory muscle weakness preventing the lungs from fully inflating during inspiration.

- Air hunger due to ventilatory failure occurs in up to 85% of patients, typically in the later stages of ALS. Non-invasive ventilatory assistance (e.g. non-invasive positive pressure ventilation) is typically used first-line.
- Supplemental oxygen should typically be avoided in ALS patients because it can suppress the respiratory drive and/or exacerbate alveolar hypoventilation.
- Studies have shown morphine to be safe and effective for the treatment of dyspnea in the ALS patient, and opioids in general for dyspnea have not shown any excess mortality in this population.

Pseudobulbar Affect (PBA): Patients with ALS may develop PBA, which typically manifests as uncontrollable laughing or crying inconsistent with the patient's mood. Nuedexta® (dextromethorphan 20mg/quinidine 10mg) is the only FDA approved treatment for this condition, but it can be cost-prohibitive in the hospice patient.

- PBA spontaneously resolves in some patients, so it is appropriate to periodically reassess if continued use of Nuedexta® is necessary.
- If continuation is necessary, a more cost-effective option is administering a compounded quinidine oral suspension along with OTC dextromethorphan hydrobromide oral suspension (Delsym®).
- Tricyclic antidepressants (e.g. amitriptyline, doxepin, imipramine) and selective serotonin reuptake inhibitor (SSRI) antidepressants (e.g. sertraline, citalopram, fluoxetine, paroxetine) have also shown benefit for PBA.

Continued on Page 3

Management of ALS continued from page 2

Pain: Most patients with ALS will experience pain, typically worsening as the disease progresses, with severe pain reported in 20% of ALS patients.

- Pain in ALS is primarily due to immobility and related issues such as mechanical back pain, musculoskeletal pain, contractures/joint stiffness, and/or pressure areas on the skin; neuropathic pain is rarer, although not well studied.
- It is recommended to follow the World Health Organization's Pain Relief Ladder for Adults for patients with ALS.
    - Non-opioid analgesics such as acetaminophen and non-steroidal anti-inflammatories (NSAIDs, e.g. ibuprofen, naproxen, meloxicam) are typically very effective for ALS-related pain in the early stages, with opioids as a next step, if non-opioids are ineffective.

Depression: Just as in the general population, patients with ALS can also suffer from depression, especially if they have unresolved pain. SSRI antidepressants are typically very effective. Lorazepam or other benzodiazepines can also be used for anxiety, but titrate them carefully due to the risk of respiratory depression.

Conclusion

In general, we treat end-of-life symptoms for patients with ALS in much the same way as we would treat these symptoms in patients without ALS.

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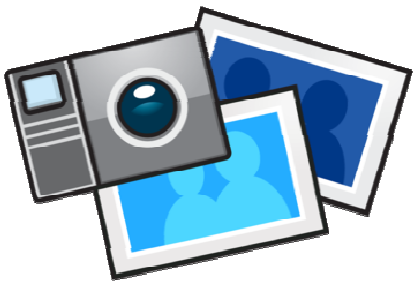
Fall Selfie Contest

Snap and win! Snap a selfie and email it to kedwards@procarerx.com to be entered into our monthly give-away! Every contestant wins a prize!



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Medication Cost Spotlight

Nate Hedrick, PharmD

Medication	Cost Changes*	Cost-Effective Alternatives	Clinical Considerations**
Atropine 1% Ophthalmic Solution	$40-$50 for 5mL bottle	Levsin Tabs (hycosycamine): $15-25 for 15-day supply	Levsin has an onset of action of only a few minutes but a duration of action of 4-6 hours. Atropine's maximum effect is usually seen at 30-60 min. As such, levsin should generally be dosed less frequently (q4-6h) than Atropine.
Chlorpromazine (Thorazine)	50mg tabs given QID: $275 for 15-day supply	*Refractory Agitation* Phenobarbital Tabs: $9 for 15-day supply Quetiapine Tabs: $14 for 15-day supply *Intractable Hiccups* Baclofen Tabs: $5 for 15-day supply Haldol Tabs: $20 for 15-day supply *Nausea* Haldol Tabs: $20 for 15-day supply Promethazine Tabs: $17 for 15-day supply	Thorazine can be used for multiple indications and the alternative agent selected to replace it should be dependent on that original indication.
Oxycodone Liquid (20mg/mL; 5mg/5mL)	20mg/mL: $300 for 30mL 5mg/mL: $1 per mg (30mL = $150)	Oxycodone IR Tablets: $20-$50 for 15-day supply Morphine IR Tablets: $20-$30 for 15-day supply Morphine (20mg/mL) Concentrate Liquid: $7-$10 for 15-day supply	Remember that oxycodone tablets can be crushed and given sublingually. Morphine's active metabolites may accumulate in patients with renal impairment. However it may be utilized acutely during the final days of life (up to approximately 7 days), even in patients with severe renal impairment, without anticipation of significant adverse effects

*Please note that drug costs may vary by geographic region and individual pharmacy. Prices provided above are estimates only and may not reflect the exact cost for a prescription for your hospice.

**Alternatives provided may not be appropriate for all patients and are provided as a general recommendation only. Please contact a ProCare Clinical Pharmacist for patient-specific recommendations.

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