A Managed Care Perspective on Scientific Advances in Amyotrophic Lateral Sclerosis

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Sample of Online Posttest

Choose the best answer for each of the following:

- 1. NS is a 63-year-old Caucasian man diagnosed with amyotrophic lateral sclerosis (ALS). He has lived in Arizona his entire life. NS reports that his father, who died at age 75 years, had diabetes, myocardial infarction, hypertension, and chronic kidney disease; and his mother has atrial fibrillation, hypertension, and breast cancer. He is a smoker and works on a farm. Which of the following issues are associated with the development of ALS?
 - A. Gender, smoking status, occupation, and age
 - B. Geography, family history, smoking status, and age
 - C. Smoking status, family history, geography, and occupation
 - D. Occupation, gender, smoking status, and geography
- 2. Which of the following is a gene associated with ALS development?
 - A. Glutamate
 - B. Urinary p75
 - C. SOD1
 - D. Nogo-A
- 3. By approximately how much did a daily dose of 100 mg of riluzole decrease the risk of death or tracheostomy in patients with ALS at 18 months, as compared with placebo?
 - A. 20%
 - B. 35%
 - C. 50%
 - D. 65%
- 4. What is the presumed mechanism of neuroprotective action of edaravone?
 - A. Antioxidant
 - B. SOD1 inhibitor
 - C. Antiglutamatergic
 - D. Voltage-gated Ca2+ channel blocker

- 5. A 70-year-old African American woman presents to her primary care physician with progressively worsening dyspnea over the preceding 6 months. She has a 50-pack per-year history of smoking and a forced vital capacity (FVC) of 65%. However, further tests reveal that she does not have chronic pulmonary obstructive disease, but instead, her reduced FVC is a result of ALS. Her ALS specialist suggests she start riluzole. What do you counsel her on about taking riluzole?
 - A. Liver function abnormalities are common.
 - B. Taking riluzole will improve her breathing strength.
 - C. Ask about bisulfite allergy, because riluzole is prepared with bisulfite preservative.
 - D. Adverse effects include dizziness, disorientation, nausea, visual changes, auditory deficits, and occasionally QT prolongation.
- 6. With such a debilitating disease as ALS, early diagnosis can be beneficial in a number of ways. Which of the following is a potential benefit to early diagnosis of ALS?
 - A. Reduced risk for tracheostomy
 - B. Increased cure rate
 - C. Extended time to medication initiation
 - D. Reduction in overall healthcare costs
- 7. Which of the followings statements is true regarding riluzole and edaravone?
 - A. Riluzole and edaravone are more effective when administered early in the disease process.
 - B. Typical wholesale acquisition costs are essentially equal for both drugs.
 - C. Edaravone and riluzole have the same route of administration—by mouth.
 - D. Edaravone and riluzole have the same route of administration—by intravenous infusion.

- 8. An open-label extension study of edaravone expanded on a 6-month, double-blind clinical trial of the drug. All patients from the double-blind study were eligible to receive 6 cycles (6 months) of edaravone treatment in the open-label extension. Which of the following best describes the results of the open-label study?
 - A. All patients exhibited the same decline in the revised Amyotrophic Lateral Sclerosis Functional Rating Scale (ALSFRS-R) in the open-label study.
 - B. Patients who initially received placebo in the doubleblind trial experienced a significantly smaller decline in ALSFRS-R in the open-label extension study.
 - C. Patients who initially received edaravone in the doubleblind trial experienced a significantly smaller decline in ALSFRS-R in the open-label extension study.
 - D. All patients exhibited no decline in ALSFRS-R in the open-label study.

- 9. As a complex disease that leads to substantial physical disability, the American Academy of Neurology has emphasized the need for a multidisciplinary healthcare approach in ALS. Which of the following is a potential benefit to multidisciplinary, patient-centered care for patients with ALS?
 - A. Improvements in measures of quality of life for patients
 - B. Improved access to medications via "right to try" avenues
 - C. Lack of ethical issues when multiple healthcare professionals are involved
 - D. Decreased time to tracheostomy and other respiratory interventions
- 10. Approval for the use of riluzole and/or edaravone in ALS may require prior authorization with certain patient criteria. Which of the following best describes typical prior authorization criteria for these drugs?
 - A. Patients may need to match the inclusion criteria of the phase 2 trials that supported the FDA approval of each respective agent.
 - B. Patients may need to match the inclusion criteria of the phase 3 trials that supported the FDA approval of each respective agent.
 - C. Inclusion criteria of riluzole may be more restrictive based on length of time on market.
 - D. Patients may need diagnosis of confirmed or probable ALS and that the prescription is being prescribed in consultation with a pharmacist.

