Amyotrophic Lateral Sclerosis Regional Variants (Brachial Amyotrophic Diplegia, Leg Amyotrophic Diplegia, and Isolated Bulbar Amyotrophic Lateral Sclerosis)

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KEYWORDS

• Motor neuron disease • Amyotrophic lateral sclerosis • Flail arm • Flail leg
• Leg amyotrophic diplegia • Brachial amyotrophic diplegia • Isolated bulbar ALS

KEY POINTS

- Regional variants of ALS include brachial amyotrophic diplegia, leg amyotrophic diplegia, and isolated bulbar ALS, and can overlap with classic ALS presentations.
- Regional ALS variants have symptoms isolated to a single spinal cord region for periods of time greater than 1 year.
- Brachial and leg amyotrophic diplegia are regional variants of progressive muscular atrophy.
- Regional ALS variants may have slower disease progression so are important clinical distinctions.

INTRODUCTION

ALS is a rapidly progressive disease characterized by degeneration of motor nerves in the brain and spinal cord; it is invariably fatal, with overall median survival between 3 and 4 years.[1,2] Three classic clinical presentations have been described; they have clear

Disclosure statement: See last page of article.
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Neurol Clin 33 (2015) 775–785
http://dx.doi.org/10.1016/j.ncl.2015.07.003 **neurologic.theclinics.com**
0733-8619/15/$ – see front matter