

## Foreword



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*Consulting Editor*

*There is no other source of knowledge but the intellectual manipulation of carefully verified observations—in fact, what is called research...and no knowledge can be obtained from revelation, intuition, or inspiration.*

—Sigmund Freud (1856-1939) [1]

Motor neuron diseases—amyotrophic lateral sclerosis (ALS) and related disorders—continue to be an enigma for the medical community. At the time of this writing (May 2008), only one drug has been approved by the Food and Drug Administration for the treatment of ALS. That drug, Rilutek, has only a modest effect. With high hopes, other drugs have been studied but have not cleared the bar of randomized, controlled trials.

It is clear that ALS is not just one disease: some ALS is sporadic and some is hereditary. There are some data that indicate there may be a “clustering” of the sporadic diseases, thus suggesting some communicable, environmental, or situational link. But the current state of knowledge is that the medical community has so far failed to identify the cause of sporadic ALS.

So what is known? It is known that one to two persons per 100,000 in the United States develops ALS in any given year. Males seem to be more likely to get it than females. The most common age of onset is between 40 and 60, although some of us in ALS clinics suspect that we may be seeing the disease even earlier. We know that the hereditary form of ALS accounts for 5% to 10% of all cases, and that some of these are linked to a mutation in copper/zinc dismutase, an enzyme responsible for scavenging free radicals. In

addition, in three regions of the Pacific—most prominently in Guam—there is a very high prevalence of ALS.

Consequently, motor neuron disease continues to be both a puzzle and a challenge to the medical community. I am fortunate to be Co-Director, with Dr. Thomas Bird, of the University of Washington Muscular Dystrophy Association (MDA) Clinic. Within our MDA Clinic we have an ALS Center, directed by Drs. Gregory Carter and Michael Weiss, the Guest Editors of this issue. ALS is generally a rapidly progressing neuromuscular disease, producing progressive weakness of the skeletal muscles, including the muscles of respiration. The outcome is that after diagnosis, patients need a number of years of rehabilitative care.

As the Consulting Editor of the *Physical Medicine and Rehabilitation Clinics of North America*, I search out Guest Editors who can bring together outstanding physicians to present in a single issue the important clinical and research points necessary for a clinician to manage diseases. We strive for a relevant issue, but with the latest research underpinnings.

This issue has far, far surpassed my expectations. Drs. Michael Weiss and Gregory Carter, as Guest Editors, have taken a single issue of the *Clinics* to a new height. With their enthusiasm and contacts with the best ALS physicians and researchers available, they have filled this issue with 16 useful articles. The reader will note that this is probably the thickest issue of the *Clinics* that they have ever received. But there is tremendous sadness as well.

Tremendous sadness.

During the preparation of this issue, one of the article authors was diagnosed with ALS. She had been scheduled to write an article on EMG as well, but was unable to carry this out. Dr. Lisa Krivickas, an old friend of the Consulting and Guest Editors, has recently been diagnosed with a familial form of ALS.

This issue is dedicated to Dr. Lisa Krivickas, a remarkable physician, Harvard academician, role model, and mother. Drs. Weiss and Carter have more to say about Dr. Krivickas and her many contributions in their dedication.

Thank you, Lisa, for contributing so much, being such an outstanding physician, and for being a friend.

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**Reference**

- [1] Freud, S. New introductory lectures on psychoanalysis. 35;1933. In Pruner, HW. Freud: His Life and His Mind. New York: Grosset and Dunlapp; 1947. p. 13.