This text represents a compilation of papers presented at a conference held on October 28-29, 1994, in Marseille, France addressing the issues of pathogenesis and therapy in amyotrophic lateral sclerosis (ALS). It is remarkable in the rate of publication following such a conference and hence still remains quite current. As such, it is a text that will find a place in the libraries of clinicians interested in the treatment of ALS, clinician/scientists attempting to frame concepts of etiopathogenesis, and to basic scientists attempting to understand the clinical relevance of studying ALS.

The text, on the whole, is well-written, topical and adequately referenced. There are several chapters that are outstanding and present excellent reviews. The chapter by Munsat on trial designs is a good, balanced overview and presents a historical perspective of drug trials in ALS. This chapter should be read in the company of those by Brooks et al. and by Meisinger et al. on attempts at quantitation of disease progression and regional onset in ALS. The chapter by Pouget et al. on the diagnosis of ALS is perhaps the best to date that I have reviewed. My only concern is the inadequacy of the discussion on primary lateral sclerosis (PLS), and the omission of key references by Pringle et al. (Brain, 1992; Canadian Journal of Neurological Sciences, 1990) and Hudson et al. (Brain Research Bulletin, 1993). These three references delineated the clinical, pathological, and diagnostic criteria for PLS, and yet are not mentioned at all in this chapter. Rowland’s paper provides some useful insight into the diagnostic difficulties that arise in the finding of a paraproteinemia in a patient with motor neuron disease. My only concern with the chapter was a paragraph on page 97 on trangenic models of neurofilament expression. While this is included in a section on anti-neurofilament antibodies, it seemed out of place and was not brought into the relevance of the overall chapter. The chapter by Rothstein on the excitotoxic mechanisms of neuron death in ALS, and particularly the glutamate-induced neurotoxicity, is well-written, clear and concise.

These positive features are off-set by a number of minor annoyances within the text itself. The organization of the text is somewhat unusual. Clinically-relevant material is left to the latter half of the text. While useful for researchers this may not be so useful for the mainstream neurologist who wishes to pick up the text and have an initial overview of the diagnostic difficulties and the classification of ALS prior to reading about pathogenesis. The paraproteinemia and immune-based chapters are scattered in that three are grouped (Appel, Drachman and Jeagar) and then 3 chapters later appears the chapter of Rowland. While the chapter by Mitsumoto and Piero discussing animal models spends considerable time discussing the Wobbler mouse, the editors have stated in the preface “the Wobbler model has been extensively investigated, but its relevance to ALS is a concern”. Indeed, it is a useful model for understanding pathogenesis of motor neuron dysfunction, and one of the most useful models to date for therapeutic trials. It is disconcerting from my point of view to find that the aluminum neurotoxicity models are scarcely mentioned, and when discussed, inaccurately. As stated in the chapter, “chronic encephalopathic signs” were not described in the model and hence this section is inaccurate. Similarly, the equine model was