Several issues relating to baclofen dosing and expected duration of therapeutic effect will need further study. It is possible that the test doses of intrathecal baclofen may be higher in stiff-person syndrome than in other conditions. In our patient, who experienced no adverse reactions to the initial testing, we do not know if a therapeutic response would have been obtained at baclofen doses higher than the standard test doses we administered. In general, we would advocate the use of blinded test-dose trials of intrathecal baclofen and placebo for each patient prior to pump placement. Because stiff-person syndrome is a rare entity, it may prove difficult to conduct a controlled, double-blind, randomized trial of intrathecal baclofen with large numbers of patients. Thus, we may need to depend on isolated case reports employing this technique in order to define the long-term effectiveness of intrathecal baclofen in stiff-person syndrome as well as the ranges of expected benefit and toxicity.

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Stanley Fahn, MD
New York, NY

Reply from the Author: The failure of intrathecal baclofen at normal test doses to help Ford and Fahn's patient with well-documented stiff-person syndrome certainly takes the steam out of our claim that GABA agonist treatment will work. Are there any differences in the patients we treated and theirs? The only obvious difference is that their patient had very late-stage disease with "extreme" rigidity and ankylosis. In such a patient with fixed abnormal posture, videotapes might be difficult to interpret and a mild effect could be missed. A higher test dose may be in order. Another possibility is that a GABA_B agonist is not enough and, as the disease advances, a GABA_A agonist is needed. As Ford and Fahn point out, our patient, followed over two years, has needed much higher doses as his underlying disease has progressed. The possibility was also raised that our second patient may have had

only pain relief due to intrathecal baclofen, not control of the movement disorder. However, clinical observations clearly showed that her spasms were decreased by treatment. Furthermore, in spasticity patients who have pain unrelated to spasms, intrathecal baclofen has not been a good analgesic. We have just completed a retrospective survey of more than 50 intrathecal baclofen patients with chronic pain and have found that pain is relieved only when spasms and spasticity are the cause.

Where does that leave us? The hypothesis is wounded, not dead. We now know, through informal communications, of several other patients with stiff-person syndrome who have responded to intrathecal baclofen. For the present, until we know who might respond, a reasonable approach is to test all stiff persons with an intrathecal test dose. Unfortunately, the response to oral baclofen in our two patients was poor and gave no clue as to the outcome of the intrathecal dose. Only an intrathecal test will do, and that requires close observation for 24 hours. Because stiff-person syndrome is so rare, no single center will be able to test enough patients with intrathecal baclofen to evaluate the success rate of the treatment. If neurologists let us know their experience, good or bad, I will be pleased to provide a tally of results.

Richard D. Penn, MD
Chicago, IL

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Correction

In "Is the incidence of dementing illness changing? A 25-year time trend study in Rochester, Minnesota (1960-1984)," by Kokmen et al, which appeared in the October 1993 issue (*Neurology* 1993;43:1887-1892), there was a labeling error in figure 1. In the mislabeled figure, the triangles denoting "total" actually indicate "males" and the dark squares denoting "males" actually indicate "total" or combined male plus female rates. The figure appears below with correct labeling.

Age- and Sex-Specific Incidence Rates/100,000 Person-Years for Alzheimer's Disease in Rochester, MN

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Is the incidence of dementing illness changing? A 25-year time trend study in Rochester, Minnesota (1960–1984)

Neurology 1994;44:1368-1368-a
DOI 10.1212/WNL.44.7.1368-a

This information is current as of July 1, 1994

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