


(Received for publication February 22, 2012.)

In Reply:

Drs. Loubser and Sheinbaum purport in their letter that “Based on the lessons learned from cerebrospinal fluid drainage, and in the interests of patient safety, we should view neurophysiologic monitoring during TAAA [thoracoabdominal aortic aneurysm] surgery not as an obscure modality as Vaughn et al. impugn, but as a standard-of-care.” Although it is fair to acknowledge that some centers have successfully adopted these techniques in the interests of patient safety, we disagree that these techniques should be considered “standard of care” (which has major medical–legal connotations).


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Confirmation of Nonanesthetic-induced Malignant Hyperthermia

To the Editor:

We read with interest the important study by Groom et al., Identical de novo Mutation in the type 1 Ryanodine Receptor Gene Associated with Fatal, Stress-induced Malignant Hy-
perthermia in Two Unrelated Families. Although we are pleased that this article merited an editorial, Nonanesthetic Malignant Hyperthermia by Lehmann-Horn et al., we have several concerns about its content.

First, we question the editorial’s statement, “The in vitro contracture test performed on a muscle biopsy of the boy reported in this article would be considered by Europeans as malignant hyperthermia (MH) equivocal.” As reported by Groom et al., case 1 had a mean response of 8.5 g (9.3, 9.0, 7.1 g) contracture in the presence of 3% halothane (less than 0.7 g contracture is designated non-MH susceptible) and a mean 2.4 g (1.9, 2.9 g, insufficient muscle to permit testing in triplicate) contracture in the presence of 2 mM caffeine (less than 0.3 g is non-MH susceptible) (Sheila M. Muldoon, M.D., Professor of Anesthesiology, Uniformed Services University of the Health Sciences, Bethesda, Maryland, written communication, December 6, 2011). This MH contracture test was conducted according to the standards of the North American Malignant Hyperthermia Group with clearly positive responses in all five muscle strips to both the halothane and the caffeine portions of this test.

The North American and European MH biopsy methods are similar but not identical. The most important differences are bolus versus incremental halothane exposure and the European designation of an equivocal research diagnostic category for subjects demonstrating positive contracture responses only to halothane or only to caffeine exposures.

Islander and Twetman have studied the concordance of the North American and European biopsy protocols. Although Islander and Twetman’s excellent study found an accordance in diagnostic outcome between the European and North American protocols of 87%, they noted a 100% accordance for individuals with contractures exceeding thresholds in at least five of six tested muscle strips. They observed diverging outcomes in subjects with less reproducible test results near the cutoff limits of their respective protocols. Because case 1’s results markedly exceeded North American diagnostic thresholds by an order of magnitude in five of five tested muscle strips to both halothane and caffeine exposures, we contend that this patient should be designated by both North Americans and Europeans to be MH susceptible and not equivocal and thus, a suitable genetic research subject.

Second, although we are aware of individual case reports, there have been no large-scale human studies that support the statement of Lehmann-Horn et al. that MH-susceptible individuals presenting with ophthalmoplegia and muscle hypotonia, hypertrophy, or spasms will be at risk for nonanesthetic MH. We believe that the authors should have clearly noted that this was only their opinion.

Finally, despite diligent parental care and aggressive medical interventions, children such as the one described in case 1 are at risk of death from this poorly understood condition. In such situations, blaming the parents helps no one.

In Reply:
We are happy to see that our editorial prompts discussion, even though the points raised by Larach et al. result mainly from the removal of the designated statement of the global context of the editorial. Three points of criticism were made: (1) the interpretation of the in vitro contracture test result of the boy (Groom et al.), (2) possible indicators for individuals at risk for nonanesthetic malignant hyperthermia (MH), and (3) the alleged blaming of the parents.

Regarding item 1: As Larach et al. correctly noted, the American and European protocols have been compared and found to be mostly concordant. However, Islander and Twetman differentiate between inclusion and exclusion of the MH equivocal results. Simply put, 9 of 74 MH-susceptible results according to the North American protocol using 3% halothane were MH equivocal according to the European protocol using 2% halothane as trigger; that is 12%. Therefore, the in vitro contracture test of the boy could very well be considered MH equivocal by European standards. But more importantly, the message of the editorial was that the in vitro contracture test may not reliably identify persons at risk. This message becomes clear in the editorial by the statement: “In addition positive In Vitro Contracture Test results were found in only 24% of 45 individuals with exertional heat stroke, and in 83% of 12 patients with exercise-induced rhabdomyolysis. Therefore more appropriate test protocols

References

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