PERIOPERATIVE MANAGEMENT OF A PATIENT WITH CONGENITAL HYPOFIBRINOGENEMIA

To the Editor—Congenital severe hypofibrinogenemia and hypodysfibrinogenemia are rare disorders complicated by hemorrhage and/or thrombosis, which pose a significant therapeutic dilemma. Anesthesiologists generally only use fibrinogen infusions to increase the fibrinogen blood level, but thrombotic events have been described after those infusions in such patients.

A 57-yr-old, 75-kg weight male with congenital severe hypofibrinogenemia was scheduled for a right sympathectomy because of recurrent painful episodes of lower limb ischemia. He had been suffering for years from both severe subcutaneous hematomas and peripheral arteritis. His sister also presented with hemorrhagic and thrombotic disease and died from postoperative pulmonary embolism. He had been given fibrinogen infusions for active bleeding nearly every 4 months during the last 20 yr; antifibrinogen antibodies were evaluated every two infusions, first by measuring postinfusion rise and half-life in plasma fibrinogen level, and second by a mixing test. No antifibrinogen antibodies could be detected. The serologic tests for HBV and HIV were negative. He had very low plasma fibrinogen levels as demonstrated...
by Elisa assay (0.08 g/l). The classical chronometric assay (Von Clauss) showed a concentration less than 0.2 g/l. Platelet fibrinogen tested by Elisa was about 90% of control level. The activated partial thromboplastin time (APTT) ratio was greater than 5.00.

On the day before surgery, low molecular weight heparin (enoxaparin, 40 mg) was administered subcutaneously, followed 60 min later by an iv infusion of 3 g of solvent detergent refined fibrinogen in a 30-min period. One hour later, the fibrinogen level was 1.7 g/l and the APTT ratio was 1.18. On the operative day, APTT ratio was 1.18 and fibrinogen residual level was 1.1 g/l. As we had decided to keep the fibrinogen level within the normal range, a second 3-g injection was given. One hour later, the fibrinogen level increased to 1.68 g/l and the APTT ratio was 1.15. General anesthesia was then induced and sympathectomy was performed without any abnormal bleeding.

During the postoperative period, 20 mg of enoxaparin was administered 8 h after the completion of surgery, then 40 mg every morning for 10 days. The blood fibrinogen concentration was assessed every morning. When a concentration less than 1.6 g/l was found (nearly every second day), a fibrinogen infusion of 3 g was administered. During the postoperative period, we did not find any clinical evidence for bleeding or thromboembolic complications.

Thrombotic complications have been reported in the literature after infusion of fibrinogen in patients with congenital severe hypofibrinogenemia. Such treatment was associated with pulmonary embolism in the propositus’ sister. The mechanism for these thrombotic episodes is still unknown, but did not involve in this patient the formation of antifibrinogen-fibrinogen complexes as no antifibrinogen antibodies could be detected. This case report suggests that when fibrinogen is used to prevent bleeding, low molecular weight heparin can be safely used with the aim to avoid thromboembolic complications in such cases.

E. CALENDA, M.D.
Departement d'Anesthesie-Réanimation Chirurgicale

J. Y. BORG, M.D.
Laboratoire d'Hématologie

C. PEILLON, M.D.
Clinique Chirurgicale

Anesthesiology

REFERENCES


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Needle Bevel Direction and Postlumbar Puncture Headache

To the Editor.—The study by Norris et al.1 claims there is an important relation between needle orientation and the severity of headache following inadvertent dural puncture.

The analysis used an unspecified chi-square test. I have organized their data into the 2 X 2 table (table 1).

Let P1 (P2) be the proportion of individuals in group 1, two who required a blood patch. The null hypothesis is that the proportions are equal: P1 = P2. The alternative hypothesis then is P1 ≠ P2. Fisher’s exact test, which seems the most reasonable for evaluating two small independent samples, yields a P of .052. The Yates corrected chi-square result is P = .078. Using an alpha level of .05, the authors’ chosen critical value, there is insufficient evidence to reject the null hypothesis.

This discrepancy is from application of a large sample procedure to the limited number of observations contained in the 2 X 2 contingency table. The chi-square distribution is continuous. Its approximation is strained when the number of cells and their expected frequencies is small. Test results in these circumstances tend to exaggerate significance.2

Were the differences “statistically significant”? Would they be clinically important? The Woolf/Taylor series 95% confidence interval for this data is 0.981, 7.027.* This interval, as expected, includes the null point. Further, it provides information about the magnitude of the effect and the precision of the estimate. In my opinion, the data suggest that bevel orientation and subsequent development of severe headache is clinically unimportant.