Use of Purified C1 Esterase Inhibitor in Patients with Hereditary Angioedema

To the Editor:—Wall, Frank, and Hahn recently reported their experience with 25 patients with hereditary angioedema requiring surgery.1 We are most surprised that use of purified C1 esterase inhibitor concentrate was not suggested as prevention, especially for patients undergoing surgical procedures at high risk (dental and ENT surgery) or those requiring tracheal intubation. Its efficacy in surgery as well as for the treatment of a crisis has long been known.2-5 Of course, fresh frozen plasma (FFP) contains the missing enzyme, but it also carries all the complement proteins, especially C4 and C2, which could keep a crisis going. Moreover in contrast to C1 esterase inhibitor concentrate, FFP may also transmit viral diseases.

We recently had to anesthetize two patients with hereditary angioedema, both of whom received C1 esterase inhibitor. Patient 1 required extensive dental surgery and patient 2 underwent removal of

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In Reply—We appreciate the interest and constructive comments of Dr. Y. Kubota and Y. Toyoda. They propose a modified technique for selective catheterization of right and left bronchi and also of branches of the upper lobes. Although in general the location of the catheter within the bronchial tree does not appear to be critical in the bacteriologic diagnosis of nosocomial pneumonia,1 the possibility of a selective catheterization of some lung segment could be important in specific cases such as unilateral bacterial pneumonia. I agree with their proposition and will soon modify our catheter according to their recommendations.

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varicose veins in the prone position. Tracheal intubation was mandatory for both patients. Despite receiving danazol (600 mg/day) for the week prior to surgery, C4 esterase inhibitor concentrations remained unmeasurable, and C4 concentrations were very low (fig. 1). Fifteen hundred units of C1 esterase inhibitor (50 ml, corresponding to 1500 ml FFP) were given intravenously 1 h before anaesthesia. The concentration of C1 esterase inhibitor rose to 2.8 U/ml in patient 1 and 3.5 U/ml in patient 2 (normal values = 0.9–4.1 U/ml). There were no mishaps during surgery or afterwards. Twenty-four hours later, C1 esterase inhibitor concentration was again unmeasurable in patient 1, while it remained normal in the other patient.

Purified C1 esterase inhibitor concentrate is ideal for preparing those patients for surgery who are at risk to develop upper airway edema; it should be preferred to FFP because it presents less risks. However, because it is very costly, we suggest it be kept for procedures in those patients in whom danazol does not increase C1 esterase inhibitor concentrations, as well as for pregnant women and children who cannot be given danazol. FFP should be kept for emergencies only when purified C1 esterase inhibitor is not available.

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