

Circumcision in a combined factor V and factor VIII deficiency using desmopressin (DDAVP)

To The Editor

Dr. Devocioğlu et al.'s recent case report in the Journal (2002; 44: 146-147) entitled "Circumcision in a Combined Factor V and Factor VIII Deficiency Using Desmopressin (DDAVP)" gives me an opportunity to emphasize our experiences briefly in two patients with these deficiencies¹.

Six- and 11-year-old boys who had decreased factor X activity (20 and 29%) in addition to low factor V (8 and 12%) and factor VIII (4 and 8%) levels were seen at İhsan Doğramacı Children's Hospital before 1980. Because of mildness of factor X deficiency in these patients, it could easily be missed without assay¹.

Dr. Devocioğlu et al.'s patient had mild Factor VIII deficiency clinically and by laboratory (32% AHG level) assay. The patient's Factor V level was also not very low (28%). Despite mildness of the disease he was circumcised after fresh frozen plasma, DDAVP administration and tranexamic acid which was continued up to the 7th day. Therefore, hemostatic success in this case could not solely be related to DDAVP administration, although its administration increased Factor VIII level up to 130% prior to circumcision.

Dr. Devocioğlu et al. should be congratulated for safe circumcision of the case. But cost of the approach must also be taken into consideration, especially in underprivileged countries such as Turkey. Corticosteroid and/or adrenalin has been used by us for elevation of Factor VIII levels in mild hemophiliacs before circumcision as in our

two patients with combined Factor V and Factor VIII deficiencies without major bleeding². One of the patients was prepared for circumcision by corticosteroid (3 mg/kg daily for 5 days) administration and the other was given adrenalin (5 µg/kg iv) infusion before the operation, which was continued daily for six days. Eventhough our patients' factor levels were much lower than the authors' patient, adrenalin was used instead of DDAVP. The cost was 136 to 300 times less than DDAVP administration according to the brand of desmopressin.

In addition to the cost, it should not be forgotten that "DDAVP is not a panacea for children with bleeding disorders" as stated by Sutor⁴.

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PREFERENCES

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Reply

We recently read the letter from Professor Şinasi Özsoylu related to our article about "Circumcision in a Combined Factor V and Factor VIII Deficiency Using Desmopressin (DDAVP)" in the Turkish Journal of Pediatrics¹. We know that corticosteroid and adrenalin were the most popular therapy in the 1970's. Professor Özsoylu published a report in which he used fresh frozen plasma (FFP) together with corticosteroid and/or adrenalin for his patients². In contrast, the Factor X level of our patient was in normal range, and FFP was given only once due to the low level of Factor V. We did not observe any problem during the post-operative period as hemostatic levels of Factor VIII remained above therapeutic levels for four days. In addition, we can even defend that DDAVP alone may be adequate for minor surgeries of this patient.

Professor Özsoylu has also stressed on the cost-effectiveness of DDAVP therapy. But, in our opinion, replacing DDAVP with FFP will cause more risk for blood-borne infections, and factors

will be more expensive as they will be used for at least four days³.

In conclusion, any operation like a circumcision can be problematic in patients with bleeding disorders, if appropriate measurements are not taken. A successful circumcision can be done safely using DDAVP.

REFERENCES

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