

Circumcision in boys with mild Hemophilia A – the Yogyakarta experience

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Abstract

Background In boys with hemophilia, there is a risk of hemorrhage resulting from circumcision. There has been no standardised management in Indonesia. We report our experience with 4 hemophiliac boys who underwent circumcision at Dr. Sardjito General Hospital, Yogyakarta, in south-central Java.

Objective To develop a safe, practical and accessible method to prevent bleeding in boys with hemophilia undergoing circumcision in Indonesia.

Methods Written informed consent was obtained from parents requesting the procedure. All boys had mild Hemophilia A (VIII: C level ≥ 7.8 U/ml) diagnosed at a median age of 4.4 years (range 0.75–9 years). Median age at time of circumcision was 7.5 years (range 0.8–12 years).

Results There were no intraoperative or post-circumcision bleeding problem encountered in any of the patients, who were discharged from hospital three days post-surgery. On follow up eight days and 12 days post-circumcision, they remained free of complications.

Conclusion Circumcision can be safely performed in boys with mild Hemophilia A following adequate pre-operative planning and prophylactic therapy. A standardised protocol for the Yogyakarta Bleeding Disorders Program, evolving from this experience, is being planned. [Paediatr Indones 2007;47:71-73]

Keywords: *circumcision, mild hemophilia A, electrocautery, laser surgery*

Circumcision has a long recorded history as an ancient medical procedure.¹ It was not only related to social, cultural and religious traditions in Islam and Judaism,^{2,3} but is also proposed to have medical benefits,

including prevention of urinary tract infection, penile malignancy and sexually transmitted diseases.^{4,5,6} However, there is ongoing debate as to the benefits and risks of circumcision in boys with hemophilia.^{7,8,9}

Circumcision in male infants and boys is an integral part of religious tradition in Indonesia, a country with the largest Moslem population in the world. Since there has been no standardised management of this procedure within the country, we undertook a pilot study for prophylactic therapy of boys with hemophilia undergoing circumcision at Dr. Sardjito General Hospital, Gadjah Mada University, Yogyakarta, Indonesia. Our objective is to develop a safe, practical and accessible method applicable to the majority areas of Indonesia, an archipelago consisting of over 13,000 islands.

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Table 1. Schedule of antihemophilic factor (AHF) and tranexamic acid (TA) administration

Day	d-1			d0			d1			D3		d4		d5	
Date	23	-11	-04	24	-11	-04	25	-11	-04	26	-11	-04	27	-11	-04
Hour	h-24	h-12	h-2	h0	h12	h24	h36	h48	h60	h72	H84	h96	h108	h120	h132
TA, oral 25 mg/kg	•	•		•	•	•	•	•	•	•	•	•	•	•	•
TA, iv bolus 10 mg/kg				•											
AHF, iv drip 25 U/kg	•	•		•	•	•	•	•	•	•	•	•	•	•	•
AHF, iv bolus 25 U/kg			•												

Keterangan : d= day, h= hour, TA= Tranexamic Acid, AHF= Anti Hemophilic Factor

Methods

Four boys, two of them from the provinces of Yogyakarta Special Territory (South-Central Java) and another two from Central Java, with mild hemophilia A (VIII: C level = 7.8 U/ml) diagnosed at median age of 4.4 years (range 0.75–9 years) underwent circumcision at median age of 7.5 years (range 0.8–12 years). Parents who requested circumcision were provided with a detailed explanation regarding the procedure and the risks that may develop. Informed consent was obtained. All circumcisions were performed by the same pediatric surgeon as an elective procedure in the operating room under general anesthesia. Either electrocautery or laser surgery was used, recombinant factor VIII (rVIII) and tranexamic acid were administered as summarised in **Table 1**. The children monitored for bleeding prior to, during, and following circumcision as an outcome measurement.

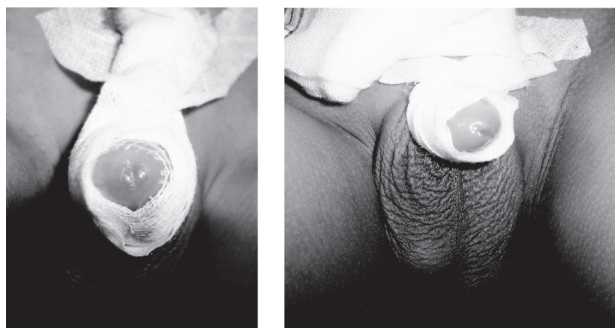


Figure 1. Very minimal bleeding occurred, as seen on day 2 (left) and on day 3 (right) post- circumcision

Results

In contrast to undiagnosed or untreated hemophiliacs who underwent circumcision in whom bleeding usually occurs intra- or post-operatively, very minimal bleeding occurred during the first day post-circumcision and resolved by the 2nd day post-circumcision and hemostasis was maintained until discharged from hospital on the 3rd post-operative day in all four children, although rVIII administration was originally planned to be administered for five days post-circumcision (**Figure 1**). On outpatient follow-up eight and 12 days post-circumcision, they remained free from bleeding or infectious complications.

Discussion

Hemophilia is the most common inherited X-linked recessive bleeding disorder in males.¹⁰ Its severe form results in recurrent, spontaneous bleeding episodes with resultant morbidity and mortality. Boys with hemophilia A suffer from a lifelong inherent inability to produce hemostatic quantities of Factor VIII coagulant protein (VIII:C) in normal amounts (100-150 U/ml). The degrees of deficiency are classified as mild, moderate, and severe, based on VIII:C levels less than 1 U/ml, 1-5 U/ml and 6-30 U/ml respectively. In our report, all four boys had mild hemophilia A with VIII:C levels ranging between 7.8 -20.8 U/ml.

Hemostatic level of VIII:C above 30 U/ml is usually sufficient to prevent minor soft tissue bleeding in patients with mild Hemophilia A. However, in

view of geographical factors and the unavailability of rVIII outside of major treatment centres, it was decided to prophylactically administer rVIII in a dose of 25 U/Kg body weight every 12 hours for circumcision. As an added precaution, tranexamic acid, an antifibrinolytic agent, was included to the prophylactic regimen. Even though this combination therapy could be considered excessive in developed countries, the situation in developing countries requires modification of published guidelines into "tailor-made" protocols that reflect regional socio-economic and health care realities.

Our prophylactic method, namely the 'Yogyakarta method', for prevention of bleeding from circumcision in patients with mild hemophilia A produced results similar to those reported by Zulfikar *et al*¹¹ where no significant bleeding or wound infection occurred postoperatively. While the total amount of rVIII used at our center was higher (225 U/Kg vs 125 U/Kg body weight), we required a total shorter duration of rVIII administration postoperatively (three days vs eight days), thereby reducing the length of stay in hospital. For economical and practical reasons it is worth to consider reduction of AHF requirement. It is recommended that circumcision be performed in conjunction with other elective surgical procedures (e.g., joint surgery or other surgical procedures, dental extractions, etc.).

The results of this pilot study should be followed by the development of a standardised prophylaxis protocol to prevent bleeding during, and following, ritual circumcision in Indonesian boys with hemophilia.

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