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Diagnosis of hemophilia in newborn circumcision: A case presentation

James L. Liu, Jason E. Michaud, Wesley W. Ludwig, John P. Gearhart*

The James Buchanan Brady Urological Institute, Division of Pediatric Urology, Johns Hopkins University School of Medicine, Baltimore, MD, USA

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ABSTRACT

Circumcision is often the earliest surgery performed in a young male's life. Though complications in this procedure are rare, prolong postoperative bleeding may be the first sign of undiagnosed hemophilia. Hemophilia is a rare X-linked bleeding disorder and if not treated prophylactically or promptly during surgical intervention can be fatal. In this case presentation we describe the diagnosis of hemophilia in a child presenting with post-operative bleeding from circumcision. We review the literature regarding the history of this disease with early surgery and highlight the current treatments.

Introduction

Circumcision is one of the earliest surgeries performed globally. The United States Center for Disease Control and Prevention (CDC) estimates the prevalence of circumcision to be 80.5% for males age 14–59 years, the majority performed during the neonatal period.[1] Circumcision is linked to several religious beliefs and has demonstrable health effects including decreasing rates of penile cancer and sexually transmitted disease.[1]

Hemophilia is a group of X-linked recessive inherited bleeding disorders. Hemophilia A and Hemophilia B lead to deficiency in the clotting factors VIII and IX, respectively, and potentiate internal bleeding or spontaneous external bleeding following minor trauma or surgery.[2] Hemophilia A occurs in 1 in 5000 male births the United States, while Hemophilia B is more rare presenting in 1 in 25,000 male births.[2] Since circumcision is often the first surgical procedure for males, prolonged post-operative bleeding may be the first sign for new diagnosis for hemophilia.

In this case report we report a patient with no prior family history of hemophilia or bleeding disorders who presented to our service post circumcision with recurrent bleeding leading to diagnosis of new onset hemophilia. We also review the literature and management of this disease in the early post-surgical setting.

Case presentation

CY a DOL (Day of life) 7 male presented to our emergency department for bleeding following circumcision. He had undergone bell clamp circumcision 24 hours earlier. On exam he had sub-coronal oozing of blood from the ventral aspect of the penis. No reported family history of bleeding disorders. The wound was reinforced with pressure dressing which resolved the bleeding. He was discharged after brief observation. Hemoglobin was 16.7 g/dL during this encounter. Urology was not consulted.

The patient re-presented to the ED on DOL 9 with recurrent bleeding. Urology was consulted. On exam he had slow oozing from the ventrum and soaked 2 diapers in the morning. He was asymptomatic and vitals stable. Pressure dressing was reapplied which stopped the bleeding and he was observed shortly before discharge. Since the patient re-presented for bleeding hematology was consulted. However, given no immediate and extended family history the patient's family declined further work up.

On DOL 11, the patient re-presented to the ED with persistent bleeding from the circumcision site. He was hemodynamically stable. On exam the bleeding was again seen as venous ooze from the ventrum. Urology placed two sutures along with fibrin glue bedside to stop the bleeding. Though this initially worked, bleeding recurred within a few hours. Hemoglobin was 8.6 g/dL-a significant drop from days prior. At this point patient was admitted for formal evaluation in the operating theater and monitoring.

The patient was taken to the operating theater on DOL 12, and the source of bleeding near the coronal margin was identified and cauterized with electrocautery. This was further reinforced with sutures and fibrin glue was applied to the incision. There was no further bleeding upon leaving the operating theater. Given the recurrent nature of bleeding, labs were drawn to rule out a bleeding disorder. Prothrombin time, INR, Von Willebrand, ristocetin, and factor 9 activity were all within normal limits. However, Factor VIII was < 1% and APTT

* Corresponding author. The James Buchanan Brady Urological Institute, Division of Pediatric Urology, Charlotte Bloomberg Children's Hospital, 1800 Orleans St Suite 7304, Baltimore, MD, 21287, USA.
E-mail address: jgearha2@jhmi.edu (J.P. Gearhart).

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prolonged at 58.9 seconds, suggesting diagnosis of Hemophilia B. Hematology was consulted and the patient was given recombinant Factor VIII. On post-operative day 1, his hemoglobin dropped to 6.6 g/dL, and he was transfused red cells. Following transfusions, his Factor VIII was 127% and hemoglobin was 12.4g/dL. The patient had no further bleeding. His parents were counselled on the genetics of hemophilia and he was discharged to follow up with Hematology and Urology.

Discussion

Circumcision is one of the oldest operations traced back as early as the stone age- 15,000 years BC.[3] It remains a longstanding, historical tradition in Jewish and Muslim faiths. Interestingly, the concept of hemophilia from post-circumcision bleeding has been understood since early times. As noted in the Talmud, a Jewish ceremonial and civil law text, for women who deliver two sons who die from bleeding after circumcision the next son is absolved from circumcision.[3] This hints to early understanding of the genetic association of bleeding disorders. Today, circumcision continues to be widely practiced for medical, religious, and social reasons.

Overall, circumcision is a well-tolerated and refined procedure. Postoperative bleeding has a reported incidence of 0.1–35%.[3] Bleeding can often be resolved conservatively with pressure, silver nitrate, and fibrin glue before pursuing surgical intervention.[4] However, in hemophiliacs postoperative bleeding can be severe.

The exact incidence of bleeding encountered from circumcision in hemophilia patients is variable, but authors estimate it is between 0.1 and 35%.[3] In these scenarios, local bleeding control is performed concomitantly with factor deficiency replacement. Certainly, for those diagnosed prior to surgery several studies highlight preoperative factor replacement. However, given the concern for inhibitor development, some argue against circumcision, or recommend bundling with other surgeries.[5] There is relative consensus for factor replacement in existing cases of hemophilia, prior to circumcision. The 2009 European Hemophilia Therapy Standardization Board[5] noted circumcision in their survey and recommended replacement of factor to between 80 and 100% preoperatively and to continue replacement therapy for 3–4 days postoperatively.

In this case presentation, the patient was undiagnosed and had no family history prior to circumcision. His initial presentation is consistent with the occasional postoperative bleeding and treated in standard form. However, with subsequent presentations hematological work up was indicated, and a diagnosis of Hemophilia B was made. The treatment with systemic Factor VIII replacement paired with localized hemostatic control was able to stop the bleeding.

Conclusion

Hemophilia is a rare X-linked recessive bleeding disorder that may be diagnosed by postoperative bleeding. Likewise, postoperative bleeding in hemophilia patients, even from benign procedures can have life-threatening consequences. Since circumcision is often the earliest surgery performed in males, it can be the first incidence to diagnose the disease. Early diagnosis and factor replacement are key to stop bleeding. In patients diagnosed postoperatively, local hemostatic control and factor replacement with monitoring is critical.

Conflict of interest statement

None.

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