

CIRCUMCISION IN HEMOPHILIA AN OVERVIEW

Revised Edition

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CIRCUMCISION IN HEMOPHILIA: AN OVERVIEW

Introduction

The World Health Organization describes health as being “not only physically healthy but also psychologically and socially healthy”. Therefore, when dealing with the health issues of people with hemophilia, medical professionals should address not only the physical but also the psychosocial aspects of their care. The ultimate aim of modern hemophilia treatment is to integrate people with hemophilia socially and culturally into society (1). Such an approach is vital when considering issues related to circumcision in people with hemophilia.

The status and importance of circumcision in various societies

Religious and cultural aspects

Although it is not known for sure when and where circumcision was first performed, there is some evidence that Egyptians were performing circumcision in the 23rd century BC (2). In some societies, circumcision is entirely a religious rite, whereas in others, it may be a traditional or cultural practice.

Circumcision is mandatory in Judaism (3, 4). It is stated explicitly in the Old Testament that “the Prophet Abraham and his successors will be circumcised” (Gen. 17:10-14).

In Islam, circumcision is performed only to conform to the practice of Prophet Mohammad and is not a religious necessity. Even in the words of Prophet Mohammad, the references date back to the practice of Prophet Abraham (5). Over time, circumcision has become an important tradition in the sociocultural life of Muslims and is practiced by almost all Muslim societies.

In some societies, such as African cultures, including Nigeria and Senegal, circumcision is a local tradition or cultural practice (6, 7).

Patient and family perspective

In societies where circumcision is a religious, social, or cultural practice, people with hemophilia and their families view being uncircumcised as unacceptable (8, 9). For example, in Turkey, circumcision is considered the first step toward being a man and a sign of becoming a member of the Turkish society (10, 11). The situation is similar in other countries with a tradition of circumcision (12). In these societies, an uncircumcised boy cannot view himself as a man and may suffer severe psychosocial problems. His family also feels this social pressure very strongly. The inability to be circumcised can result in feelings of inferiority, both in the person with hemophilia and his family. In a survey by Kavakli et al. of 105 people with hemophilia and their families, 94% of the families wanted their children to be circumcised, and 60% of the boys and 82% of the families reported feelings of inferiority for this reason (11). Loutfi et al. stated similar reasons in their study and concluded that in order to satisfy the religious and social beliefs of people with hemophilia and their families, the risks and costs of circumcision should be accepted (13). Since it is considered an ordinary procedure, families may go to centers that do not have knowledge and experience on hereditary bleeding disorder for the circumcision of their children.

Medical reasons for circumcision

In countries where circumcision is not a traditional practice, the main reasons for the procedure are medical. Although there have been debates on the medical benefits of circumcision, it has many advantages.

In circumcised males, the rate of sexually transmitted diseases (STDs), including HIV, is significantly lower (14, 15), and it was shown that circumcision has a protective effect against penile cancer development (16). Similarly, the risk of cervical cancer in partners of circumcised males is lower than the partners of uncircumcised males (17, 18). Circumcision also reduces or prevents problems related to the foreskin, such as phimosis, paraphimosis, or balanitis

(6, 19). But there are some studies that report that phimosis can be treated with topical corticosteroids, at least in some patients, with a lower cost when compared to circumcision (20, 21).

Early circumcision significantly decreases the risk of urinary tract infections in boys. The rate of urinary infections in uncircumcised boys is 8-10 times higher than that of the circumcised boys (22, 23, 24). Although not confirmed with multiple studies, it was suggested that circumcision reduces prostate cancer incidence (25). Despite demonstrated benefits of circumcision, a study performed in Israel showed an increase in urinary tract infection prevalence after early ritual circumcision (26). However, most of these ritual circumcisions were not performed by doctors. Similarly, studies examining the postsurgical complications of circumcision show that the risk of complications is higher when the procedure is not performed by qualified doctors (27, 28).

Despite the evidence of benefit stated above, which has been accepted by the World Health Organization (29), health organizations of many countries believe the evidence is too vague and state that the risk of complications and the functional role of the foreskin make routine circumcision unacceptable. But still, they suggest that the decision of circumcision can be made by taking the opinion of family and child, if the child is old enough to declare his wish to be circumcised (30, 31). Although a review of the literature may create the impression that advantages of circumcision outweigh the disadvantages, the child or his parents should not be under any kind of pressure during the decision making. When a decision in favor of circumcision is made, then the procedure should be carried out as a safe surgical procedure by qualified medical professionals, under adequate pain preventive measures, and under hygienic circumstances. Following a workshop held in 2012, the German Ethics Council (Deutscher Ethikrat) decided that circumcision could be done under certain conditions as stated above, based on an examination of medical, religious, cultural, criminal, and constitutional law (32).

Balancing social desire, risk, and cost

In developing countries, people with hemophilia face many problems, such as lack of adequate factor supply and the

undertaking of surgical procedures without proper precautions that threaten the life of people with hemophilia. A survey conducted in Turkey found that one in every three families with hemophilia had experienced a death related to circumcision (33). It has been reported that, especially in developing countries, perioperative hemorrhages may be fatal. In a study reported from Nigeria, post-circumcision hemorrhage was observed in 52% of people with hemophilia (6). A review of the literature found 101 published reports of bleeding from circumcision in newborns (34). It is noteworthy that this problem maintains its importance even in the publications in recent years (35, 36). However, circumcision can be performed in hemophilia patients with low complication rates, when necessary precautions are taken (9, 37, 38). A serious problem that can be seen due to circumcision is the development of inhibitors due to exposure to factor concentrates (39).

One of the main obstacles to the circumcision of people with hemophilia is the high cost of the operation, which may be as much as US\$10,000, mainly due to the cost of clotting factors that are not readily available. In a developing country with limited resources, it is almost impossible for parents to have their sons circumcised if they do not have health insurance coverage (40). However, despite high costs, the social pressure imposed by religious and cultural beliefs remains high, even for boys with hemophilia (1, 13). This demand is so strong that most families will risk their children's lives by having them circumcised by unauthorized people, without taking the necessary precautions, or by not disclosing the bleeding status. Thanks to innovations in coagulation factors and non-factor replacement products, faster coagulation, shorter healing of wounds, and reduced cost of replacement products are all possible (36, 41, 42). In addition, the use of pharmacokinetic guided therapies in the perioperative periods allow for less factors to be needed compared with standard body weight-based dosing (43).

In short, we need an optimal approach to address the social demand for circumcision of people with hemophilia, despite the risks and costs. People with hemophilia who have many unmet needs may view themselves as being handicapped all their lives. It should be the responsibility of healthcare professionals to protect these boys (or men who decide later in life) from the additional psychosocial problems resulting from not being circumcised. Currently, due to the widespread use of prophylactic factors, the

cost of factor concentrates needed for circumcision has become bearable.

Suggested measures for the circumcision of people with hemophilia

Circumcision of people with hemophilia should not be considered a minor procedure and should not be performed without taking adequate precautions. Before operating on a patient with hemophilia or another bleeding disorder, all necessary laboratory tests should be done to confirm the factor deficiency, level of factor, and presence of inhibitors. Informed consent should be obtained from the family. The insurance or reimbursement agency should be notified and necessary products (factor concentrates, packed red blood cells, etc.) and other medications (tranexamic acid, DDAVP, local tissue adhesive, analgesics, antibiotics, etc.) should be obtained beforehand.

Increasing local hemostasis in the surgical wound may decrease the risk of postoperative bleeding. The general risk of postoperative bleeding in people with hemophilia is about 15-20%. Therefore, adequate precautions should be taken for any surgical procedure, including circumcision. Parents or patients should be informed that bleeding may occur despite all these precautions (including factor replacement) (44, 45).

Systemic and local measures to prevent bleeding include:

- Factor replacement (1, 46, 47)
- Desmopressin (DDAVP) (1, 46, 47)
- Inhibitors of fibrinolysis (10, 38, 47)
 - Tranexamic acid
 - Epsilon aminocaproic acid (EACA)
- Local tissue adhesive (e.g., fibrin glue, blood stopper) (48, 49, 50, 51)
- Laser surgery (52, 53)
- Meticulous surgical technique and hemostasis (13, 36)

Application of fibrin glue, which reduces the amount of factor used and hence the costs, may be considered safe. Martinowitz et al. used fibrin glue in circumcision in 1992 (49). Avanoglu et al. combined fibrin glue with continuous factor infusion for 48 hours and reduced the duration of factor replacement therapy and costs (48, 50). Similarly,

Yilmaz et al. showed that usage of tranexamic acid and fibrin glue decreased the necessary factor dosage and cost (54). Oner et al. stated that in a patient with continuous bleeding despite factor replacement before and after circumcision, Ankaferd Blood Stopper produced results in minutes (51). However, this result needs to be supported with further studies.

Turkish method for circumcision of people with hemophilia

In Turkey, we have developed a technique for circumcision of people with hemophilia. Since 1996, we have used a bloodless circumcision technique, which we call the Turkish method, that reduces the amount of factor needed by half (55). Our goal was to develop a safe, practical, acceptable, and comfortable method for both the patient and his family. This approach minimizes the two main obstacles to circumcision: the risk and the cost.

Our novel technique has been cited in the pediatric urology guidelines of European Association of Urology in 2018 and 2021, under the “phimosis” chapter. The EAU Paediatric Urology Guidelines Panel, which consists of an international group of clinicians with expertise in this area, stated that circumcision can be performed in children with coagulopathy with 1-5% suffering complications (bleeding), if hemostatic agents or a diathermic blade are used (56).

In the authors’ center, 147 people with hemophilia A, hemophilia B, factor VII deficiency, factor XIII deficiency, factor V deficiency, Glanzmann thrombasthenia, and von Willebrand disease (VWD) have been circumcised until 2010 (Table 1). The same surgical method was used in all cases. However, factor concentrates and other hemostatic agents changed during the course based on the disease entity. The results of these patients, including length of stay in the hospital, the amount of factor concentrates used, hemostatic treatment plan, and total costs (in US dollars) had been published before (38, 55).

The authors’ current treatment protocol with standard factor concentrates is shown in Table 2. In this protocol, the hospitalization period is limited to 48 hours. Duration of factor support ranges from 4 days in mild-moderate type hemophilia, VWD, and other rare factor deficiencies,

to 11 days in severe cases. However, it may vary in cases with inhibitors.

Recently, two severe hemophilia A cases on extended half-life factor VIII prophylaxis were circumcised with the same method. Both cases received two additional doses (50 IU/kg/dose) PEGylated rFVIII and tranexamic acid for 7 days, with successful results (41). Another 10-year-old patient with severe hemophilia A and high titer inhibitors, who was participating in an emicizumab clinical trial, was circumcised without any need for bypassing agents, using only tranexamic acid (57).

During the COVID-19 pandemic, the number of circumcisions decreased like other surgical procedures; however, three patients who had phimosis were circumcised. Circumcisions resumed during the post-pandemic period.

Another 41 patients were circumcised during major surgery under general anesthesia. In these cases, the foreskin was cut surgically, and sutures were placed afterwards. The preparation phase was similar in these cases; however, the factor replacement program was designed according to the primary surgical procedure.

Before the operation, tests were completed on each patient to determine complete blood counts, factor levels, and presence of inhibitors. Then, a hemostasis plan was

designed. Necessary medicines such as factors, tranexamic acid, DDAVP, and creams were supplied. All patients were hospitalized on the day of circumcision four hours prior to surgery.

The Turkish method is performed as follows:

All patients are hospitalized four hours prior to the procedure, informed consent or assent for surgery is obtained, and the required factor replacement is administered. Local anesthesia (2-4 mL 2% lidocaine hydrochloride) is injected around the base of the penis to obtain a ring blockage.

A modified straight clamp similar to the Mogen clamp is applied distally to the glans penis by pushing the glans back with the thumb (Shield technique) and index finger to avoid inadvertent glandular damage (Figure 1). After the genital area has been scrubbed and prepared, the foreskin is stretched distally with two straight clamps in the 6 o'clock and 12 o'clock positions (Figure 2, 3).

The clamped foreskin is crushed and squeezed (pressed) between two jaws of the modified Mogen clamp. Then the foreskin on the distal side of the clamp is excised using the diathermic knife developed for bloodless circumcision in our country. The cutting electrode of the device is simply applied parallel to the distal surface of the modified Mogen clamp after activation. The excision takes no longer than 10 seconds (Figure 4).

TABLE 1. Characteristics of patients circumcised using the Turkish method from 1996-2010

	Year 1996-2010
Total patients	147
Hemophilia A	122
Hemophilia B	12
von Willebrand disease	9
Glanzmann thrombasthenia	1
Factor VII deficiency	1
Factor XIII deficiency	1
Factor V deficiency	1
Age – median (range)	11 (1.5-37) years
Body weight – median (range)	33 (9-102) kg

TABLE 2. Factor replacement protocol (Turkish method)

Factor Replacement (IU/kg) *, **		
Protocol II		
	Severe hemophilia	Moderate/mild hemophilia and VWD
Day 1 Preop	25	15
Postop	15	10
Day 2-7	15	10***
Day 9, 11, and 13	15	10 (day 9 only)
Total	175 IU/kg	95 IU/kg

* Double doses for Hemophilia B, DDAVP not used

** Tranexamic acid 25 mg/kg/day for 7 days + 10 mg/kg during surgery

*** Plus DDAVP 0.3 mcg/kg/day for first 3 days

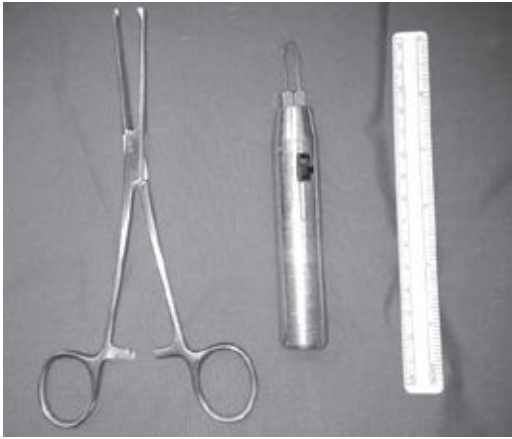


FIGURE 1. Modified circumcision clamp and battery-operated diathermic knife.



FIGURE 2. Stretched penile skin



FIGURE 3. Clamping the foreskin

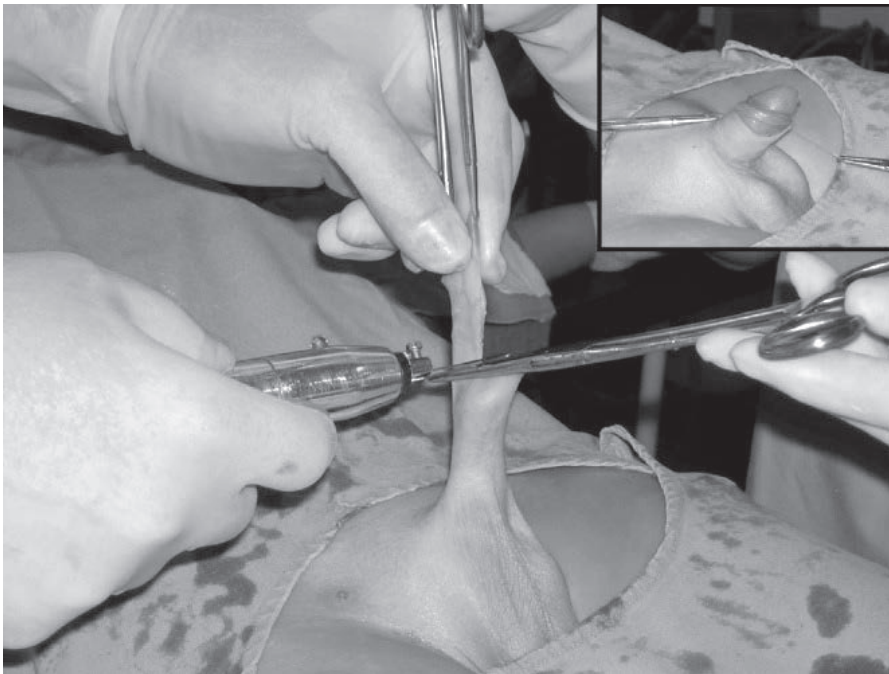


FIGURE 4. Clamped foreskin and excision with diathermic knife; and final appearance after circumcision (small picture).

Following completion of foreskin excision, no bleeding is seen in the surgical field. The remaining skin and mucosa are sutured interruptedly with 5-10 rapidly absorbable polyglactin sutures (Figure 4).

Antibiotic ointment is applied to the wound, and no dressing is necessary. When necessary, paracetamol (acetaminophen) 15 mg/kg/dose is administered orally

for analgesic purposes. After the operation, the patients are kept under observation and treated in the hospital for an average of 2 (1-3) days.

No significant bleeding or wound infection occurred. Only seven patients (4.7% [7/147]) had transient minimal bleeding because of delay in factor supply,* and responded quickly to factor administration. One of them needed

* The delay in factor application is due to the patient not following the instructions given to them, and delaying factor thinking that the wound has healed.

sutures. Hematoma on the ventral aspect of the wound at the frenular junction was seen in one case on the ninth post-operative day and was treated successfully with cleaning, pressure bandage, and factor injection. Mild to moderate edema and hyperemia seen along the suture line in almost all cases lasted for three to five days. No other complications were encountered.

Several biopsies taken from the excision line to evaluate the histological effect of the diathermic knife on penile tissue revealed the same diathermic effect in character and depth as a laser incision. There was no harmful effect on deep tissues, vessels, or nerves.

The period of complete wound healing varied from 7 to 21 days. The patients returned to their routine daily life within a week. Excellent patient and family satisfaction was reported.

After 2005 using this same surgical method, the average factor consumption was less: 175 IU/kg (range: 165-210) for severe hemophilia, and 95 IU/kg (range: 85-215) for moderate hemophilia, mild hemophilia, and VWD. In congenital factor VII deficiency, 125 IU/kg FEIBA was used (total: 8500 IU). In congenital factor V and XIII deficiencies, 6 units of fresh frozen plasma were used. In Glanzmann thrombasthenia, 16 units of platelets were used. Early wound healing in the first period was the determinant in the decreased consumption of factor.

Forty-one people with hemophilia who were circumcised with other operative surgical indications had general anesthesia and were hospitalized for a median of 10 days (range: 5-22 days). They used more factor as a result of their primary problem, with no bleeding observed at the circumcision site in these 41 patients. Wound healing began after the fifth day. Patients started on a physiotherapy program on the fourth day because of their primary problem, and the addition of the circumcision procedure did not hinder the management of their primary condition. Factor consumption in this group of patients was 550 IU/kg (range: 500-590) for severe hemophilia and 405 IU/kg (range: 360-480) for moderate and mild hemophilia. The total cost of these procedures could not be calculated because each patient underwent a different operation.

Comparison of techniques

As it is evident from our study, an open surgical approach consisting of “meticulous surgical technique + scrupulous hemostasis + factor replacement” (that is, other than the Turkish method) for the circumcision of people with hemophilia is not economical, due to the large amount of factor needed. DDAVP and inhibitors of fibrinolysis are not powerful enough to stop bleeding and they should be combined with other agents.

In a randomized study of 230 children including some with hemophilia, Mendez-Gallart et al. found that those in whom the procedure was performed with bipolar scissors had less bleeding than those in whom the classic surgical technique was used (58). Although it was reported that there is less bleeding in circumcisions performed with different laser devices when compared with classical surgical circumcision, there is not enough supporting literature to recommend using these devices in hemophilia patients (59, 60, 61).

Studies have reported that local use of fibrin glue is a safe and cost-effective method for treating patients with hemorrhagic disease for minor surgical procedures, including circumcision (48, 49). In our study, the method used for circumcision has proven to be safe, effective, and cheaper than local application of fibrin glue in males with hemophilia. In addition, this practical and quick procedure can be done under local anesthesia, avoiding the additional risk of general anesthesia. Avanoğlu et al. reported the average cost of factor replacement was US\$366/kg/case in a group of patients with moderate and severe hemophilia circumcised using fibrin glue, and US\$472/kg/case in the control group, while the average cost was US\$127.8/kg/case for patients with moderate hemophilia and US\$225/kg/case for those with severe hemophilia in our study using protocol I (48).

Circumcision in infants with hemophilia

People with hemophilia can be circumcised as newborns when their body weight is low. This reduces the amount of factor used, and hence the cost.

Developmental hemostatic changes may increase infants' bleeding tendency (due to lower levels and activity of vitamin K, contact factors, and impaired platelet function) whereas higher levels of von Willebrand factor and its multimers, together with reduced activity of the natural coagulation inhibitor proteins, play a protective procoagulant role during the early post-natal period (62).

In the Jewish tradition, it is recommended to perform circumcision early (within the first 8 days of life). However, since the risk of developing inhibitors is high during the neonatal period, postponing circumcision after six months of age is preferred and 6-18 months is deemed a better age for circumcision. Despite this risk, no inhibitors were found in two different studies in Israel, where early circumcision was performed. However, 16 of 20 babies diagnosed with sporadic severe hemophilia A between 2010 and 2018 were admitted to the clinic due to bleeding after circumcision (63).

In terms of reducing contact with factor concentrates, it was promising that bleeding occurred in only 3 of 40 babies who were circumcised and were given one dose of factor and tranexamic acid for 3 days in the preoperative period to reduce the inhibitor risk (64). Despite these practices, the option of using the Turkish method, where surgical technique is important even in the neonatal period, should also be remembered.

Suggestions for the economical circumcision of people with hemophilia

The following are suggestions for the Turkish method in particular, and for circumcision in general, to make the procedure more economical for people with hemophilia:

- If circumcision is an elective procedure and the patient is going to have dental therapy, orthopedic intervention, or surgery where factor replacement is inevitable, the circumcision can be done at the same time. This saves factor and reduces fear.
- People with hereditary bleeding disorders who were circumcised should be included in the prophylaxis program and closely monitored for inhibitor development.
- If the cost of coagulation factors can be reduced or non-replacement products can be used, the overall cost will be significantly reduced.
- In people with hemophilia, the circumcision procedure should be performed electively by an experienced surgeon and hematology team in a resourced hematology treatment centre with access to clotting factor concentrates (45).
- In people with hemophilia undergoing circumcision, intraoperative care should be taken to cauterize all bleeding vessels (45).
- For people with hemophilia undergoing circumcision, the WFH recommends adjusting clotting factor replacement to the clinical course of the procedure. If continued clotting factor replacement is required, the goal would be to maintain factor levels above 50 IU/dL for the first 3 days, and above 30 IU/dL for the subsequent 4-8 days (45).

Conclusion

The healthcare of people with hemophilia includes not only replacing missing clotting factors, but also maintaining the social, economic, and psychological health of these people. Therefore, for those people who request it, circumcision is very important for improving not only their physical but also their psychological and social well-being. Various techniques have been used all over the world with considerable success. In societies where circumcision is a religious, sociocultural, or psychological obligation, there is no doubt that the procedure should be done if a person with hemophilia wants to be circumcised, medical reasons notwithstanding. The goal is to perform the operation with the safest, most effective, and cheapest method. Performing circumcision by experienced medical staff under optimal conditions will prevent negative outcomes (6, 11). In this regard, the circumcision of people with hemophilia can be done safely using the above-mentioned method and equipment. Bloodless circumcision with a diathermic knife using the Turkish method is a reliable and practical surgical alternative for boys with hemophilia (38). Moreover, since this technique requires less factor concentrates, it is much cheaper than other surgical techniques, especially if it is performed early in life.

Appendix 1 – Circumcision in neonates with hemophilia – The Israeli perspective

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Consensus evidence-based guidelines regarding prophylactic treatment for circumcision in infants with hemophilia are currently lacking. Notably, the guidelines provided for circumcision care in hemophiliacs of older age may not be relevant during the neonatal period. Developmental hemostatic changes may increase infants' bleeding tendency (due to lower levels and activity of vitamin K, contact factors, and impaired platelet function) whereas higher levels of von Willebrand factor and its multimers, together with reduced activity of the natural coagulation inhibitor proteins, play a protective procoagulant role during the early post-natal period (62).

The Israeli National Hemophilia Center serves a total of 723 patients with hemophilia, prospectively followed by our comprehensive team since diagnosis. As Jewish tradition recommends circumcision by day 8 after term delivery, early hemophilia diagnosis among new families usually occurs due to bleeding following this surgical challenge. For children born to known carriers with a family history of hemophilia, prenatal or perinatal diagnosis dictates proper periprocedural prophylaxis.

Thus, most hemophilia patients are neonatally exposed to either replacement factor concentrates and / or blood products (in case of major bleeding complications). Interestingly, despite concerns regarding potential immunogenicity of early extensive FVIII exposure, the early single factor dose exposure in most Israeli hemophilia neonates did not result in increased risk of inhibitor formation (65).

A retrospective analysis of the National Hemophilia Center's database disclosed that between 2010-2018, a total of 20 novel sporadic severe hemophilia A patients were diagnosed; for 16 of these patients, bleeding after circumcision was the presenting symptom (63).

In another study, we retrospectively examined the results of circumcision of 40 consecutive neonates with hemophilia, diagnosed after delivery. Our protocol included application of a single dose of factor replacement prior to the

procedure and 3 days' oral treatment with tranexamic acid. Only 3/40 neonates with hemophilia experienced mild post-circumcision oozing. No major or any life-threatening bleeds were encountered. As the therapy administered was minimal and the procedure simple and feasible, we concluded that circumcision in neonates with hemophilia should be performed according to the family's and physician's discretion. Avoidance of circumcising male babies with hemophilia due to fear of bleeding complications is not supported by the results shown within our patient cohort (64).

Our current protocol consists of a single dose of FVIII (50-75 IU/kg) or FIX (75-100 IU/kg) administered intravenously 1-3 hours before the procedure and additional tranexamic acid, given orally, 3-4 times daily for a total of 3 days. Surgeons or the Rabbi performing the procedure are advised to control local hemostasis and use sutures meticulously, if possible, to avoid delayed bleed. The use of fibrin glue, abundant in the past, is not routinely recommended anymore, as results are now favorable with single dose replacement therapy. Notably, a second prophylactic dose (that may be applied if needed, 1-2 days afterward) is rarely required.

Similar principles for replacement therapy administration are recommended in case of some other rare bleeding disorders (e.g., severe FVII deficiency, severe Von Willebrand disease [VWD], severe FXIII deficiency), whereas for some other coagulopathies (eg, severe FXI deficiency, hypo-dysfibrinogenemia, VWD type II, Factor V / Factor V + Factor VIII deficiency), the sole use of antifibrinolytics may be sufficient.

With the recently evolving novel non-replacement therapies, whose use is expanding for hemophilia prophylaxis at all age groups, one should remember that circumcision, as a surgical procedure, still deserves attention, and the use of additional replacement therapy is compulsory to avoid bleeding complications. Unlike treatment for adult patients with hemophilia, the intensity of treatment (dosing regimens, treatment timing, and measures of additional care) may be minimal if the procedure is performed during the neonatal period.

Appendix 2 – Circumcision in hemophilia – The Moroccan perspective

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It is common when asking an individual why he wants to be circumcised or parents why they want their child to be circumcised, the answers are the same: “To become a man; boys make fun of me, I will never go back to school; my family and friends don't know that I am not circumcised; to fully be a part of my religion; etc.”

Circumcision is part of religious and socio-cultural practices and provides a sense of belonging. In the Occidental culture, circumcision is a medical procedure that is not covered by the healthcare system. This forces immigrants to practice circumcision in their country of origin. In the author's opinion, the biggest obstacle to protecting this medical practice is the consumption of factor concentrate, which makes the cost of the surgery significantly higher for a child with hemophilia compared to a child without hemophilia. It is recommended to circumcise the child either before he is one year old or after he is five years old when there is the least risk of trauma.

It is, therefore, essential to consider a procedure that would be safer and less costly. The objective of this protocol is to:

- Adapt a technique that can be used for the circumcision of a child with hemophilia.
- Reduce the cost of the procedure.
- Result in few or no hemorrhagic incidents.

For a child without hemophilia, there is no need to take hemostatic precautions. However, for a child with hemophilia, there is a risk of bleeding during and after surgery, even if the procedure is considered a minor surgery with a hemorrhagic risk. If, during the pre-operative procedure, the administration of concentrate factor is mandatory, we asked ourselves the following questions:

- Is administering factor VIII or factor IX mandatory during the post-operative period?
- What can cause bleeding during the post-operative period?
- Does factor replacement injection fully protect the child?

The author's protocol is for patients with hemophilia A and B, any age, all stages, and all degrees of hemophilia severity (mild, moderate, or severe).

- One week before the procedure, a test for inhibitors should be performed. If the test results are negative, the circumcision can proceed.
- After receiving the parent or caregiver's consent, healthcare providers should offer education on therapeutic options since the post-operative follow-up is done either at a day hospital or at home by the parents if they know how to inject the product. The education of parents and care staff are key elements to the successful outcome of this protocol.
- One hour before the procedure, an injection of factor VIII or factor IX (10-20 IU/Kg) is given to the individual.
- The surgical procedure is carried out in the same way that it would be for a child without hemophilia.
- Upon leaving the operating room, the child is placed for observation in a day hospital with his parents at his side. If no bleeding is observed, the child is placed in the care of his parents and observation will continue at home. Although the dressing is not obligatory, we administer ointment and dressing on the surgical site. The venous line is kept in place.
- In the post-operative period, the injection of clotting factor concentrate is done only in the case of bleeding.

During the circumcision, there is an inflammatory reaction with local edema which acts like a mechanical and biological glue. If during the forecasted injection, the child is not bleeding, will the injection of factor avoid spontaneous bleeding?

- Yes, however, it is most likely that the post-circumcision bleeding is post-traumatic. Injection of factor will indeed reduce the degree of bleeding for bleeds that are far from the kidney. However, during the post-circumcision procedure, the area that will most likely bleed is the frenulum of the penis. A bleed that occurs at this level can only be stopped by a surgical procedure (stitches). Systematic injections will not be mandatory and will not avoid post-traumatic bleeding.

Do not hesitate to ask the surgeon to add stitches at the frenulum of the penis using excellent thread. The most significant risk of hemorrhage occurs between the fifth and tenth day after circumcision.

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