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Technic and Results of Circumcision in Patients With Haemophilia at the University Hospital Center of Libreville (Gabon)

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1. Abstract

1.1. Introduction

Circumcision remains a wide spread practice in sub-Saharan African countries. This surgical procedure, which causes complications, particularly hemorrhagic ones, can reveal the disease. The hemophilia field is an aggravating factor. The authors report the preliminary results of their experience on the practice of circumcision on a patient with hemophilia.

1.2. Patients and Method

This was a prospective, descriptive, longitudinal study over 5 months, from May to September 2019. It was conducted at the General Surgery Department of CHUL in Gabon, in collaboration with the clinical hematology and pediatrics departments. All patients had type a hemophilia and were known to pediatric and clinical hematology departments. Factor VIII injections (AFSTYLA®1000UI) were performed one hour before and at the end of the procedure, at 24 hours and 48 hours after the surgery, that is at least 4 doses. In case of hemorrhage, an additional dose was administered. The act was performed under sedation in the facial mask in the operating room.

1.3. Results

Four male patients with hemophilia were selected. The average age was 7 years [3-25 years]. Indications of circumcision were socio-cultural in 3 patients and medical in one. The average duration of intervention was 24.75 minutes [15-37 minutes]. A case of focal hemorrhage on the suture area was found. The average dose of coagulation factors injected was 4250 IU per patient. The average duration of hospitalization was 3.25 days [2-5 days]. The average duration of healing was 9 days [7-12 days].

1.4. Conclusion

Hemophilia is a hereditary hemorrhagic coagulopathy. It remains difficult to manage in our regions because of the high cost and the unavailability of coagulation factors. Preliminary results are positive. This act requires effective multidisciplinary collaboration to ensure safe circumcision.

2. Keywords: Hemophilia; Circumcision; Libreville.

3. Introduction

Circumcision remains a wides pread practice in sub-Saharan African countries driven by religious, socio-cultural or medical reasons [1]. This surgical procedure is a source of complications, especially hemorrhagic ones, all the more serious if they occur on people with hemophilia. The social pressure linked to non-circumcision is such that it becomes a bad experience for the patient and his relatives. This inherited genetic condition of coagulation remains an obsession for practitioners, because it can be revealed after circumcision [2] by hemorrhage.

The authors aim to report the results of their preliminary experience on the practice of circumcision on people with hemophilia at the

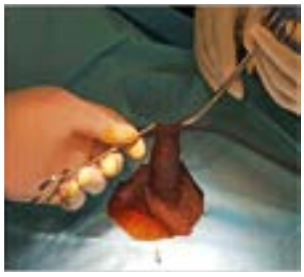

Figure 1: Intraoperative view. Marking of the section level about 2 mm below the projection on the foreskin on the base of the glans arrow).

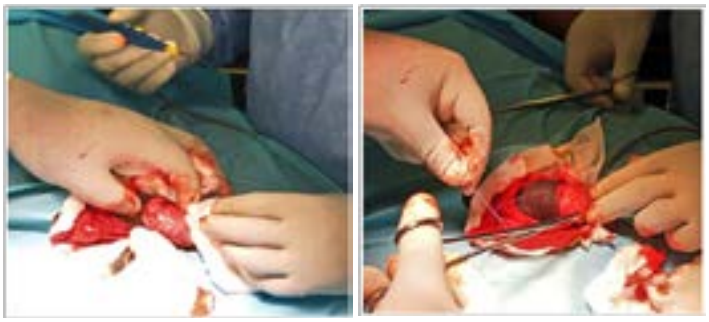

Figure 2: Intra-Operative Views. A: Hemostasis of penile sheath performed with electric bistoury. **B**:then supplemented with sutures with resorbing Vicryl® 3-0 thread.

University Hospital of Libreville (CHUL), Gabon.

4. Patients and Methods

This was a prospective, descriptive, longitudinal study over 5 months, from May to September 2019. It was conducted at the General Surgery Department of CHUL in Gabon, in collaboration with the clinical hematology and pediatrics departments. All patients with type a hemophilia were known to pediatric and clinical hematology departments. They had previously received injections of coagulation factors previously for a various reasons. They were diagnosed Type A hemophilia based on TCA elongation, with a factor VIII rate below 30%. These were minor to moderate forms. The following protocol was systematically performed for each patient, including a dose of 1000 IU/L of factor VIII (AFSTYLA®) one hour before the operation, at the end of the operation, then administered at 24 hours, then 48 hours after the treatment, that is at least 4 doses. In case of hemorrhage, an additional dose was administered at 20m/kg of tranexamic acid every 12 hours, intravenously and locally applied to the dressing. Niflumic acid at a dose of mg/kg intrarectal and paracetamol at a dose of 60 mg/kg formed the analgesic protocol. The prophylactic antibiotic treatment based on cefuroxime was systematic. All patients underwent surgery at the block under sedation with facial mask. Interventions were performed by the same surgical team. The operative protocol included the following steps: Marking the section level about 2 mm below the projection on the foreskin on the base of the glans (Figure 1). Pushing the glans under the predefined level and setting up a Kocher forceps preventing the glans to go up. Cutting through the foreskin using the bistoury blade. Hemostasis performed with electric bistoury and supplemented with Vibryl® 3-0 absorbable suture (Figure 2 a and b). Mucocutaneous approximation by eight separate points with Vicryl® 3-0 absorbable suture (Figure 3A). Fitting of a fat pad soaked in povidone-iodine around the suture area remaining in place for seven days with bi-daily soaking of povidone-iodine (Figure 3b). Systematic hospitalization in the postoperative surgery department. Removal of the fat compress on the 7th day with povidone-iodine contact until complete healing.

Case 1: 3 years old KDS child, type A hemophilia, known and followed since 1 year. He had a brother with hemophilia. He had received 4 doses of 1000 IU of factor VIII (AFSTYLA®) peri-operatively, that is 4000 IU according to the protocol. No complication was noted. He left the hospital on the 2nd post-operative day. Healing was observed on the 8th day.

Case 2: AJ child, 6 years old, type A hemophilia, known and followed since the age of 2 years. He had already received several injections of factor VIII (AFSTYLA®) especially for hemophiliac arthropathy. His parents, Muslims, decided to circumcise their child. The intervention was conducted according to the protocol. He received 4 doses of 1000 IU of factor VIII (AFSTYLA®) peri-operatively, that is 4000 IU. On the 2nd post-operative day, he went back home. Healing was observed on the 7th day.

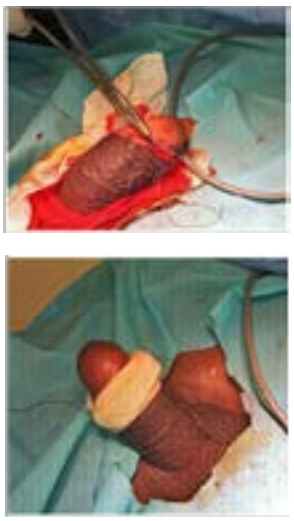

Figure 3 A: Intra-operative view, mucocutaneous approach by separate points. **B:** post-operative view: aspect at the end of the procedure, affixing an oily dressing soaked with povidone iodine.

Case 3: OBY child, 4 years old, type A hemophilia, known and followed since 2 years. There was no trait of hemophilia in the family.

He had focal hemorrhage on the mucocutaneous area at H8. He received a complementary suture, an additional injection of factor VIII (AFSTYLA®) and tranexamic acid to limit the complication. He had received 5 doses of 1000 IU of factor VIII (AFSTYLA®) that is 5000 IU. He left the hospital on the 3rd postoperative day. Healing was observed on the 9th day.

Case 4: NMBF, 25 years old, type A hemophilia, known and followed since 20 years, has an elder brother with hemophilia. He had received doses of factor VIII (AFSTYLA®) for coagulation 8 months before for traumatic hematoma of the left thigh. The intervention was conducted according to the established protocol and he received 4000 IU of factor VIII (AFSTYLA®). He left the hospital on the 2nd postoperative day. There were no complications noted. Healing was observed on the 12th day.

5. Results

Four patients were selected. They were all male. The average age was 7 years old with extremes at 3 and 25 years old. There were 4 cases of type A hemophilia. Indications about circumcision were socio-cultural for 3 patients. It was indicated for a phimosis in a patient. The average duration of intervention was 24.75 minutes with extremes at 15 and 37 minutes. No intra-operative incident was found. Focal bleeding at the suture area was a complication in a patient with postoperative H8. He had undergone an elective suture at the bleeding point with a dose of 1000 IU of Factor VIII (AFSTYLA®) and a tranexamic acid dose intravenously and locally applied. There was no infection or hematoma. The average stay at hospital was 3.25 days with extremes at 2 and 5 days. Complete healing occurred on average after 9 days with extremes at 7 and 12 days.

6. Discussion

Hemophilia is a rare pathology in our country and not widely documented [3]. This inherited hemorrhagic disease is characterized by the lack or decrease of coagulation factors VIII in type A and IX in type B. The severity depends on the percentage of existing factors. Circumcision is practiced mostly for religious or socio-cultural reasons in Africa [4], as in our series. The age of the child with hemophilia at circumcision varies by region. The average age in our study was around 7 years, which is lower than those of other authors such as Sagna [4] and Padaro [5], who respectively had an average age of around 10 years and 12.5 years. The family trait of hemophilia was found in half of the cases, placing our results between those of Noufissa (44%) [6] and those of Sagna [4] which recorded this family trait in more than half of the cases. The handling of hemophilia requires the administration of coagulation factors that are not always available in our context. In developing countries, it is still perceived as a disease with disastrous medical and social consequences due to unavailability and high cost of treatment [6]. The national health insurance does not take into account the coagulation factors whose cost is not accessible for the average patient. Through some associations for the fight against hemophilia, the acquisition of these products is made possible at the national level. Circumcision has a great religious value and social recognition in some countries, and it is difficult for the patient and his parents to cope with its abstention [6].

Circumcision is a common circumstance for the discovery of hemophilia [2, 5, 6]. Sometimes, spontaneous hemorrhagic phenomena, especially in severe forms, such as hemarthrosis or visceral hemorrhage, can reveal the disease [7]. Patients of the series were all already known as hemophiliac and were followed at the CHUL hematology department at the time of circumcision. There was a family trait among half of them, as Sagna et al. [4] also observed. This act, still tagged as "minor surgery" when practiced in hospital, is often practiced by paramedical qualified - or not - staff, who have random notions of anatomy, surgery and asepsis [8], under local anesthetic on an out patient basis. It is still performed by traditional healers in some of our regions, due to their belief and sometimes lack of financial means. The conditions of child immobilization and the hemorrhagic risk are likely to make people fear circumcision on people with hemophiliac under these conditions, hence our option to systematically operate patients with hemophilia under general anesthesia in the operating room. This provides better technical comfort and safety against peri-operative circumcision accidents such as amputation of the glans [8, 9]. The shorter duration of almost 10 minutes of circumcision compared to the Sagna series [4] could be explained by our conditions of practice. The hemostasis control is an obsession for the surgeon on a patient with hemophilia. One can use various techniques for this purpose, such as the cutaneous mucosa method in 2 steps under guide pliers used by Sagna et al [4] or the use of biological glue as recommended by Avavoglu et al [10]. Complications such as hemorrhage are common in case of circumcision on the hemophiliac patient [6]. One quarter of the patients had a hemorrhagic complication in our series. The use of biological glue and increased amounts of administered factors appear to reduce these hemorrhagic complications [11]. Other infectious complications on patients with hemophilia were recorded [4, 8], unlike in our series where none was recorded. Aseptic conditions in the operating room could explain this result.

7. Conclusion

Hemophilia is a hereditary coagulopathy exposing to hemorrhagic risk. It remains difficult to manage in our regions because of the

high cost and the unavailability of coagulation factors, a real cornerstone for treatment. The preliminary results of circumcision on patients with hemophilia in our context are positive. This act requires effective collaboration between different specialists (surgeons, pediatricians, hematologists and anesthesiologists) in order to minimize the risk of hemorrhage associated with this condition. The inclusion of these products by health insurance would facilitate their accessibility and thus optimize the management of this condition.

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