Original Article

Circumcision in patients with bleeding disorders: Can it be done safely?

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ABSTRACT

Purpose: The purpose of our study was to review outcome of circumcision among children with bleeding disorders at our institution and also to determine the impact of optimization leading to safe circumcision.

Methods: Data representing boys (age 0-16 years) who underwent routine circumcision at the Aga Khan University Hospital (AKUH) between1988-2014 was retrospectively reviewed. Children with bleeding disorder were identified using International Classification of Diseases (ICD) Code 64.0. Data was retrieved and confidentially was maintained. SPSS version 19 was used for statistical analysis.

Results: During 26 years 13,200 circumcisions were performed at AKUH. Amongst these 8,463 (64.11%) were done by using Plastibell, while 4,737 (35.88%) by open slit method. Only 23 (0.17%) children were identified with bleeding disorder. Two groups were made, Group-A (n:15) children with known bleeding disorders having circumcision and Group-B, (n:8) those in whom bleeding disorder was diagnosed after circumcision.

Median age of children in Group-A was 9 years. All children in Group-A underwent open circumcision. 10 patients had Factor VIII deficiency, 2 had Glanzmann's thrombasthenia, 1 had Factor IX deficiency, 1 had Quebec platelet disorder, and 1 had Von Willebrand disorder.

Median age of children in Group-B was 3 months. 7 out of 8 underwent plastibell while one had circumcision by open technique. 7 were diagnosed as Factor VIII deficiency and 1 diagnosed later to have Glanzmann's thrombasthenia. Statistical analysis showed significant difference among these two groups' p-value with respect to age (p-value 0.00) and family history (p-value 0.04- Fisher's exact test). Both groups had similar postoperative length of stay. Overall bleeding complication rate after optimization was 13.33%.

Conclusion: With the help of hematologist and adequate Factor replacement, these children can be managed as daycare. We suggest risks and benefit should be discussed with parents before procedure.

Key words: Bleeding, circumcision, hemophiliacs

INTRODUCTION:

Circumcision is one of the oldest surgical procedures with the origin dating back to ancient times[1]. In some societies i.e. Muslims and Jews, circumcision is entirely a religious rite, whereas in others it may be a cultural and traditional practice[2]. However, in some societies in Africa it is a local cultural practice. According to WHO, 30% of males are circumcised, majority being Muslims. Its prevalence has risen from 48% to 61% during the years 1988 to 1991[3]. Age at circumcision shows differences between societies. Jews as a continuation of God's covenant, circumcise boys 8 days after birth, whereas usually muslim males are circumcised before puberty[4]. It is one of the commonly performed surgical procedures in pediatric surgery practice in our country due to religious reason.

Bleeding is the most common early complication after circumcision followed by infection[5]. The complication rate depends on multiple factor including anatomic variations and surgical technique used. Mostly bleeding occurs along skin edges or from discrete blood vessels, surrounding frenular area[6]. Reported incidence of bleeding after circumcision in normal individuals is reported to be 0.1-35%. However, post circumcision bleeding in children with bleeding disorder is higher which varies between 23-56%[7]. As majority of circumcisions are performed in neonatal period, excessive post circumcision bleeding is often the first sign of congenital bleeding disorder as reported by Shahida et al to be nearly 62%[8]. This higher incidence of bleeding in such children can lead to catastrophic bleeding events. However limited data is available about management of these children.

In developing countries where routine screening is not done, many individual are diagnosed with bleeding disorder following circumcision. In Pakistan, 90-95% circumcisions are performed by village barbers or paramedical staff. And rest being performed within a proper medical setup[9]. Due to paucity of local literature on this subject, this study was conducted too review our experience of dealing with these

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Received: 25-08-2017
Revised: 18-05-2018
Accepted: 05-06-2018

situations.

MATERIALS AND METHODS:

This was a retrospective chart review. The study was conducted at the section of pediatric surgery, The Aga Khan University Hospital Karachi, Pakistan. We reviewed files of all children with bleeding disorders who underwent circumcision from April 1988 till May 2014. ICD code 64.0 (procedure code), 99.06, 155.0, 286.0, 286.3, 286.9, 287.1, 287.5, 570, 571.5 was used to identify patients. Children were divided into two groups, Group-A included children with known bleeding disorders prior to circumcision and Group-B included those in whom bleeding disorder was diagnosed after performing surgery, Figure 1. Data including patient's demographics, type of bleeding disorder, reason for circumcision, Bleeding disorder status whether known before or after the procedure, pre and postoperative strategies used for homeostasis and length of hospital stay. Data was collected in coded form on a specifically designed questionnaire. All the possible efforts were made to prevent patient disclosure. Data was entered and analyzed using SPSS version 19.

RESULTS:

A total of 13220 children were circumcised during the study period. Plastibell technique was used in 8,463 (64.11%) (Mabis Healthcare USA/ Hollister) followed by open method 4,737 (35.88%). Total 26 children developed bleeding after circumcision. However, the record of 3 children were incomplete, therefore 23 were included in our study (Figure 1). We divided the children into two groups. Group A; with known bleeding disorders prior to surgery included 15 children and Group B; diagnosed later with bleeding disorder had 8 children. The surgery was done as an elective procedure for all children. The indication for circumcision was religious grounds in all of the cases. In majority of patients, procedure time was observed to be 15-30min (39%) followed by 30-60 minutes (34%). However in 2 children procedure time was more than one hours. Sutures and bipolar diathermy were used to secure homeostasis in 21 out of 23 children

In Group A, 7 children underwent procedure under penile block, while 6 underwent procedure under general anesthesia. All had circumcision by open method. Majority of the children (73%) were hemophiliacs. Two children were found to have Glanzmann's thrombasthenia. One child had Quebec syndrome and another had Von Willebrand disease.

Preoperative optimization of the children was done according to the nature of the bleeding disorder. Guidelines of world federation of hemophiliac for developing country were followed including pre-operative levels of factor (40-80 IU/DL), factors transfused1-2 hours before surgery, post-operative levels between 20-50 IU/DL for 1-5 days transfusion(10). Since the majority of children were hemophiliacs, therefore factor levels were optimized before and after the procedure in accordance with these guidelines in collaboration with hematologist. Those with platelet deficiencies were transfused platelets. The same strategy was used for postoperative optimization i.e. based on the nature of the bleeding disorder, as given in Table 1.

Three children had post-operative complications in Group-A. One patient developed hematoma on day 15. Other had postoperative bleeding on day 12, while third had bleeding on day 9. This child was initially diagnosed as Factor XIII deficiency but the bleeding did not stop despite adequate factor replacement. Later he was found to have Glanzmann's thrombasthenia, Table 1.

In Group-B, 3 children were operated at our institute, while 5 children were referred to us after having had procedure from some other facility. All of them were hemophiliacs with Factor VIII deficiency as shown in Figure 2. 7 out of 8 underwent plastibell circumcision. Postoperatively these children received plasma and plasma products as they had no prior diagnosis of congenital bleeding disorder, Table 1.

DISCUSSION:

Male circumcision is one of the oldest surgical operation known, its history dates to ancient times (15000 BC)[11]. Worldwide prevalence is 20-30%. It is ritual among Jews and Muslims. Complication rates vary from 1-15%. It's most common complications is bleeding, rate varies from 0.1-35%[12]. Risk associated in children with bleeding disorder is even greater and if proper measures are not taken it can be life threatening[13]. In our country because of religious obligation parents wish to have their child get circumcised despite a known bleeding disorder[14].

In countries, where circum is not a traditional practice, the main reason for the procedure is a medical indication, they include lower rate of urinary tract infection seen only in infancy, 10 times higher in uncircumcised boys, 3 times of higher risk of penile cancer, and the risk of cervical cancer in partners of circumcised males is low[15,16]. Patient on clean intermittent also gets benefit from circumcision[17].

Bleeding disorders can be categorized into three groups: disorders of platelet function or number, disorders of clotting factors, and a combination of these. Initial laboratory investigations should include a complete blood count (CBC), prothrombin time (PT), activated partial thromboplastin time (APTT), platelet function screening and peripheral film examination should be done in patient with positive family history of bleeding diathesis[18]. A preoperative screen for a patient with a negative history and examination should include a CBC, PT, and APTT only.

Clotting factor disorder whether acquired or inherited can result in bleeding disorders[19]. Hemophilia is the most commonly the X-linked recessive diseases characterized by deficiency either of factor VIII (hemophilia A) or factor IX (hemophilia B, or Christmas disease). The incidence is 1 per 5000 live births with no racial tendency[20]. The diagnosis is suggested by an elevated APTT level in a male patient

Variables	Group A (N = 15) known diseased	Group B (N = 08) not diagnosed	P - Value
Median age	9 years	3 months	0.001
Preoperative optimization • Cryoprecipitate • Clotting factors • Platelets • Factors + cryoprecipitate	15 (100%) 03 08 03 01	0	
Surgical techniques • Open method • Plastibell	15 (100%) 0 (0%)	01 (12.5%) 07 (87.5%)	
Postoperative optimization • Factor 8 • Cryoprecipitate • Factor 9 • Cryoprecipitate + FFP • platelets	15 (100%) 06 04 01 01 03	0	
Postoperative complications	03 (20%)	0	
Postoperative Bleeding	02 (13%)	0	0.00
Family history	07 (46.66%)	0 (0%)	0.04
Length of hospital stay	05 days	05 days	0.5

Table 1: Comparative analysis of two Groups

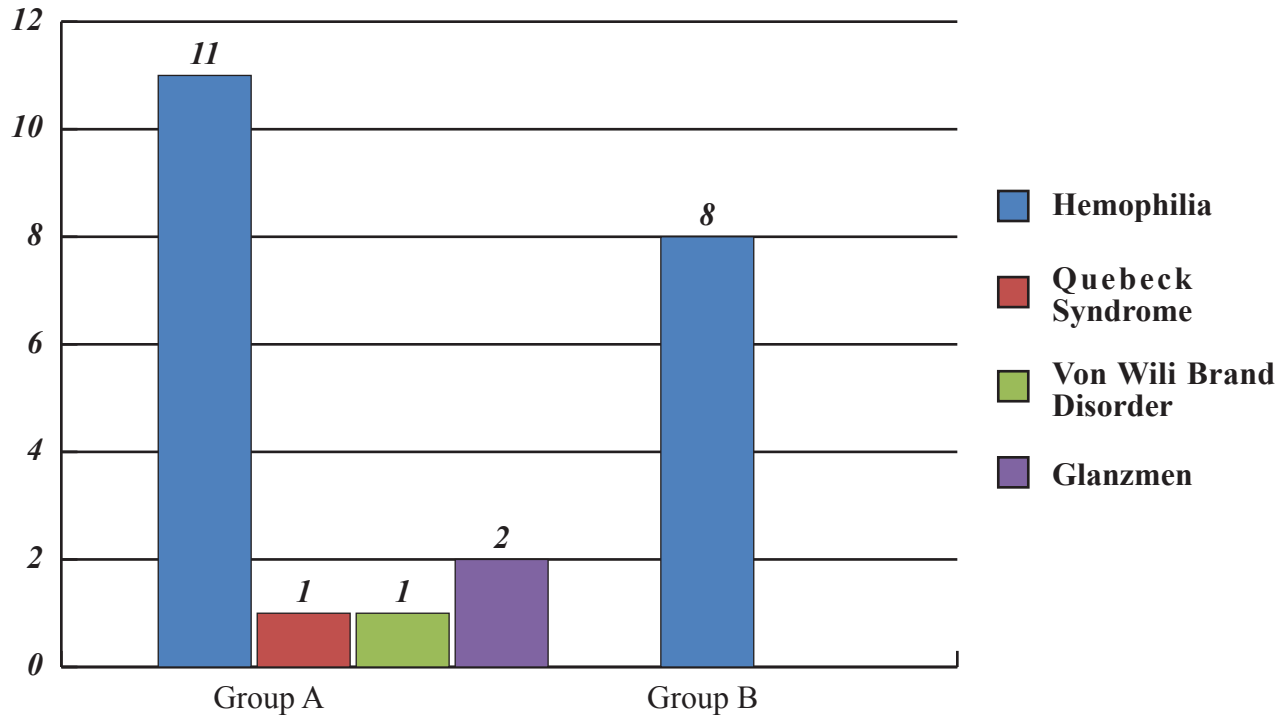

Figure 2: Diagnosis in two groups

with a positive family history. Factors VIII and IX levels are decreased in hemophilia's A and B, respectively[21].

Most surgical and invasive procedures can be performed safely in children with bleeding disorder with factor replacement therapy[22]. Appropriate pre-operative optimization of patient with factor replacement can decrease pre, peri- and post-operative bleeding complications. Replacement therapy is required for children undergoing circumcision. A target level of 80% and maintenance of replacement therapy during 3–4 days are recommended. Adjunctive treatment with antifibrinolytics and/or fibrin glue should be considered[23].

In Group-A preoperative and postoperative optimization was done specifically according to nature of disease. We have used Factor VIII in 8 children, cryoprecipitate in 3 children, cryoprecipitate and Factor IX in single child, platelets in 3 children. Whereas, postoperative optimization was done using Factor VIII in 6 children, cryoprecipitate in 4 children, cryoprecipitate and Factor IX in 1 children, cryoprecipitate and FFPs in one child and platelets in 3 children. However, no pre operative optimization done in Group-B as we were unaware of their diagnosis. All children in Group B received cryoprecipitate and FFPs postoperatively, Table 1. These are the patients had bleeding disorder but it was not diagnosed unless had circumcision and was diagnosed after circumcision. In our health care system it is possible as most of children had no access to screening or mostly because parents are not aware of other symptoms like bruising or unexplained bleeding.

A study conducted in Iran had a very large sample size of 424 children with previously diagnosed bleeding disorder and that of a US study had a sample size of 21 children. In comparison to these studies, our study had 15 children with previously known bleeding disorders. More than ¾ of the children in our study were hemophiliacs. All our previously diagnosed children were preoperatively optimized before the procedure, whereas in the US study 57% and in the Iranian study 35% children were preoperatively optimized[24,25]. We have found the lowest rate of bleeding complication in our study. On comparison, the children with previously known bleeding disorder were found to have slightly higher risk of bleeding 20%. The reason could be attributed to the fact that two children were circumcised before factors were available for optimization, while in third case diagnosis was delayed.

This study is an eye opener for us as ritual circumcision is a norm in our socially and because of lots of social and religious obligations parents are more the willing to proceed for circumcision. Unfortunately most of our children don't have an access to better screening due to poor socio-economic conditions. Although this procedure apparently seems a simple procedure but this can be a high risk procedure especially for this cohort of patient who hasn't been screened before or even if known to us due to various bleeding disorders. We believe without adequate optimization it can lead serious complications which are impossible to manage outside tertiary care setting.

Circumcision can be done safely in children with bleeding disorders but with prior adequate optimization and timely prep consultation with hematologist in management decreases the incidence of postoperative bleeding and circumcision may be done as a daycare procedure[26].

CONCLUSION:

The complication rates were found to be similar in children with and without bleeding disorder. With prior optimisation which we have mentioned above complications rates are similar between these high risk group and normal children.

Circumcision can be done safely in children with bleeding disorders but with prior adequate optimization. Although procedure appears to be simple, but without adequate optimization it can lead serious complications which are impossible to manage outside tertiary care setting.

The involvement of hematologist in management decreases the incidence of postoperative bleeding and circumcision may be done as a daycare procedure.

Conflict of interest:

There is no conflict of interest in our study

Acknowledgement:

We acknowledge the Department of Hematology, The Aga khan university hospital, Karachi, Pakistan for their continuous support without which this study was not possible.

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