

Full Length Research Paper

Social impact of delayed male circumcision in a group of hemophiliacs' patients from Sudan

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Abstract

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Haemophilia is the most common hereditary disorder of blood coagulation; there are two types, according to the factor deficient, haemophilia A in which there is factor VIII deficiency and haemophilia B with factor IX deficiency; and known as Christmas disease. In addition to life-long bleeding and other complications, a large number of haemophiliacs endure other sufferings. Social stigmatization, drug addiction, neglect, dropping out of school, loss of the father, these are only some of their problems. This prospective study was carried out on haemophilic patients who present themselves for circumcision, usually past the time for this procedure as compared to their peers. The study aimed to critically assess and address the social problems that arise from delayed circumcision in male haemophiliacs in the Sudan. The study also assessed effect of circumcision on the problems of the individuals. The guardians/patients were enlightened about their disease and the surgical operation that was going to be performed. Following informed consent by guardians of the children or the children themselves if they are older than 18 years of age, a pre-designed questionnaire was filled by interviewers. In addition to demographic data, the questionnaire also contained detailed medical and treatment histories. The surgical operation was performed under factor concentrate cover according to the Sudanese National Haemophilia protocol and using cautery knife to minimize the chance of post-operative bleeding. 87 patients with haemophilia were enrolled in the study, 82 with haemophilia A and 5 with haemophilia B. The mean age of 12.8±5.6 year. The post-operative period went smoothly, apart from manageable breakthrough bleeding in three patients. All haemophiliacs and their families consider that circumcision is a very important and as a mandatory procedure.

Keywords: Factor eight, coagulation, partial thromboplastin time, bleeding, X-chromosome, hereditary

INTRODUCTION

Haemophilia

The word haemophilia is derived from the Greek word "blood" and to "love". Haemophilia was called a royal

disease because Queen Victoria, Queen of England, was a carrier and she passed the disease on several generat-

ions of the royal family (Aronson, 1973). Haemophilia was first described during the second century where it was encountered in Jewish texts. Rabbinical rulings exempted male boys from circumcision if two previous brothers died of bleeding after the procedure. The same ruling applied to the sons of a woman who had married twice, which indicate the hereditary nature of the condition. The Arab physician Albucasis (1013-1106) also described a family whose males died of bleeding after trivial injury (Criangraude, 1997). So, haemophilia is the oldest known hereditary bleeding disorder, which results from a deficiency of blood coagulation factor VIII resulting in haemophilia A or factor IX resulting in Haemophilia B, and is caused by a defect in the corresponding genes located in the tip of the long arm of the X- chromosome. The genetic defect can result from deletions, point mutations, insertions or inversion in this region. Haemophilia accounts for > 90% of all severe coagulation disorders. Congenital Haemophilia is inherited in an X-linked recessive manner; these genes are carried by the female line and phenotypically expressed in males. Approximately one third of female carriers have some problems with bleeding. In approximately one third of cases there is no family history and haemophilia results from a spontaneous mutation. Haemophilia is classified as mild, moderate or severe according to the residual activity of the affected clotting factor. It is dangerous to perform even simple surgical procedures, such as tooth extractions, in patients with haemophilia without correcting the deficiency (Haemophilia. First edition, 1997).

In some societies circumcision is entirely a religious ritual, or it may be traditional or a cultural practice. It is mandatory in Jewish (Glass, 1999). In Islam circumcision is performed only to conform to the practice of the prophet Mohammed and is not a religious necessity (Rizvi et al., 1999). Over time, circumcision has become an important tradition in the sociocultural life of Muslims and is practiced by almost all the Muslim society. Circumcision is a local tradition or cultural practice (Shittce and shokunbi, 2001). In societies where circumcision is a religious, social or cultural practice, people with haemophilia and their families view being uncircumcised as socially unacceptable. In Sudan, being uncircumcised, a boy suffers severe psychological problems, and his family also feels this social pressure very strongly. This results in inferiority complex both in the haemophilic boy and his family. Loutfi et al. stated similar reasons in his study and concluded that in order to satisfy the religious and social believes of haemophilias and their families the risks and costs of circumcision should be accepted (Loutfi et al., 1988). People with haemophilia in the developing countries have many problems and view themselves as being handicapped all their lives. It should be the responsibility of health care professional not to subject these boys to an additional

psychosocial trauma from not being circumcised (Rickwood, 1999).

Indications for circumcision

Circumcision is the removal of a fold of loose skin (the foreskin) that covers the head (glands) of the unerect penis. The medical indications are phimosis, recurrent balanoposthitis, balanitis, and paraphimosis (Stenram et al., 1986).

Phimosis is a medical condition, occurring in many uncircumcised males, in which the foreskin cannot be fully retracted over the head of the penis. Paraphimosis is a serious condition where the foreskin has been retracted, and cannot return to the normal position. The preputial ring is too tight, squeezing the shaft of the penis, and possibly cutting off the blood supply (Stenram et al., 1986).

Advantages of circumcision

Circumcision has a role in prevention of other conditions as urinary tract infection sexually transmitted diseases, herpes genitals, candidiasis, gonorrhea (Singh et al., 2005; Diseker et al., 2000). There is increased evidence which suggests that there is an increased risk of female to male transmission of HIV and human papilloma virus in uncircumcised men (Baily et al., 2007; Auvert et al., 2009). Women with circumcised partners had a lower risk of cervical cancer than women whose partner were uncircumcised (Ogilvie et al., 2008). It reduces carcinoma of the penis, cancer of prostate and it improves personal hygiene (Hayes et al., 2000).

Complications of circumcision

Apart from pain and distress and the side effects of anaesthesia, there are many reported complications of circumcision. The most frequent acute problem is haemorrhage. Infection is usually minor, but rarely septicemia and meningitis may occur as well as accidental trauma (Gee et al., 1996; Kravitz et al., 1992). Long term complications include meatal ulceration, the incidence of meatal ulceration among circumcised infants is very common. Stenosis, meatal stricture results from prolonged or repeated episodes of meatal ulceration. The repeatedly irritated meatus becomes narrowed. This results in pain and difficulty with urination. In extreme cases this can result in infections and kidney problems (Graves, 1998).

Other complications as urethral fistula, phimosis of remaining foreskin, urinary retention, cutaneous tags, glandular ulceration and necrosis can occur (Kaplan,

1977). Poor cosmetic appearance, and psychological trauma are common complications. Children with prominent prepubertal fat may have a concealed penis following surgery, which tends to resolve at puberty (Kaplan, 1977; Trier and Drach, 1973).

Circumcision in Sudanese Haemophiliacs

In Sudan circumcision is a religious, social and cultural practice, and uncircumcised boys have a feeling of inferiority and it is a social stigma. In Sudan people with haemophilia face many problems, the main problem is the lack of adequate factor supply because of its high cost. In spite of this high cost there is a huge social demand for circumcision of boys with haemophilia. Circumcision of haemophiliacs should not be considered as a minor procedure and should be performed taking adequate precautions.

Objectives

The objectives of this study are:

- 1- To study the social impact of delayed male circumcision on a group of haemophilic patients and their families.
- 2- To revise and update the circumcision protocol previously developed by the Sudanese National Haemophilia Society.

MATERIAL AND METHODS

This is a cross sectional hospital base study, conducted in Khartoum Teaching hospital. 87 Sudanese males' patients with haemophilia, 82 of them with haemophilia A and 5 with Haemophilia B were circumcised according to the Sudanese National Haemophilia society circumcision protocol.

The patients and their parents were interviewed using a designed questioner before and after the circumcision procedure. A written ethical consent was taken from the patients and care takers.

Investigations done to all patients before circumcision were as follows:

Complete blood count:

- Hb ,TWBC and platelets count
- Platelets aggregation

Coagulation profile

Bleeding time (BT), prothrombin time (PT), activated partial thromboplastin time (APTT) and thrombin time (T.T).

Factor VIII and factor IX assay and inhibitor screening were done. Circumcision procedures were done under complete aseptic precautions using diathermy and under supervision of the urological team.

According to the following protocol:

- Weigh the patient.
- Calculate the dose according to the formula $30 \times \text{wt}$ in kg/2 for factor VIII, $30 \times \text{wt}$ in kg for factor IX.
- Give the patient the dose half an hour preoperatively, then repeated 8-2 hourly postoperatively according to the status of homeostasis as judged clinically in the first 48 hours for haemophilia A.
- once daily for haemophilia B
- In the third day give the dose once for seven to ten days till healing.

RESULTS

Absence and drop out of school were seen in all patients more than ten years and in 20% of patients less than ten years, as seen in figure (1). The majority (80%) of patient tended to stay in doors and not participate in any outdoor social activities.

2% of the fathers abandoned their families due to the cost of the treatment and the social stigma of the daughters being carriers of the disease, as shown in figure (2).

95% of the patients and 80% of the parents viewed circumcision as a mandatory procedure, as shown in figure (3). 55% of patients and 80% of their parents had anxiety because of the bleeding risk, as seen in figure (4).

All parents viewed circumcision as a mandatory procedure and they were ready to take the high risk. After circumcision, none of the patients had significant bleeding or complication during the procedure. 82 patients stay in hospital for 3 days and 5 of them stay for 7 days. All of them discharge in a good condition. 4 patients developed break-through bleeding due to physical trauma after discharge, 2 of these patients received 3 additional doses of factor 8 the 3rd received 10 additional doses of the factor, as shown in figure (5) .

The average cost of circumcision was calculated at >500.000 SDG per patient, for patient without postoperative complication. Break-through bleeding increase the cost of the procedure by 50-100%.

Following circumcision all parents were happy and satisfied and all patients in school age express their willingness to go back to school, the unstable environment in the house was relieved and some of the fathers who abandoned their families previously return home.

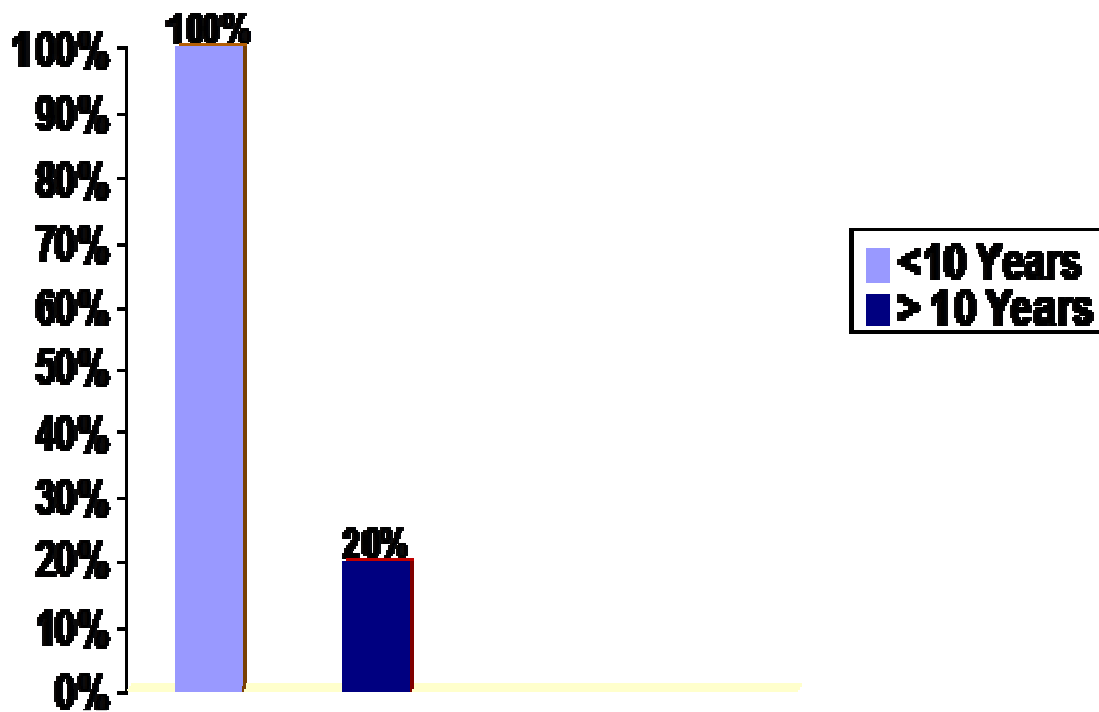


Figure 1. Drop out and absence from school

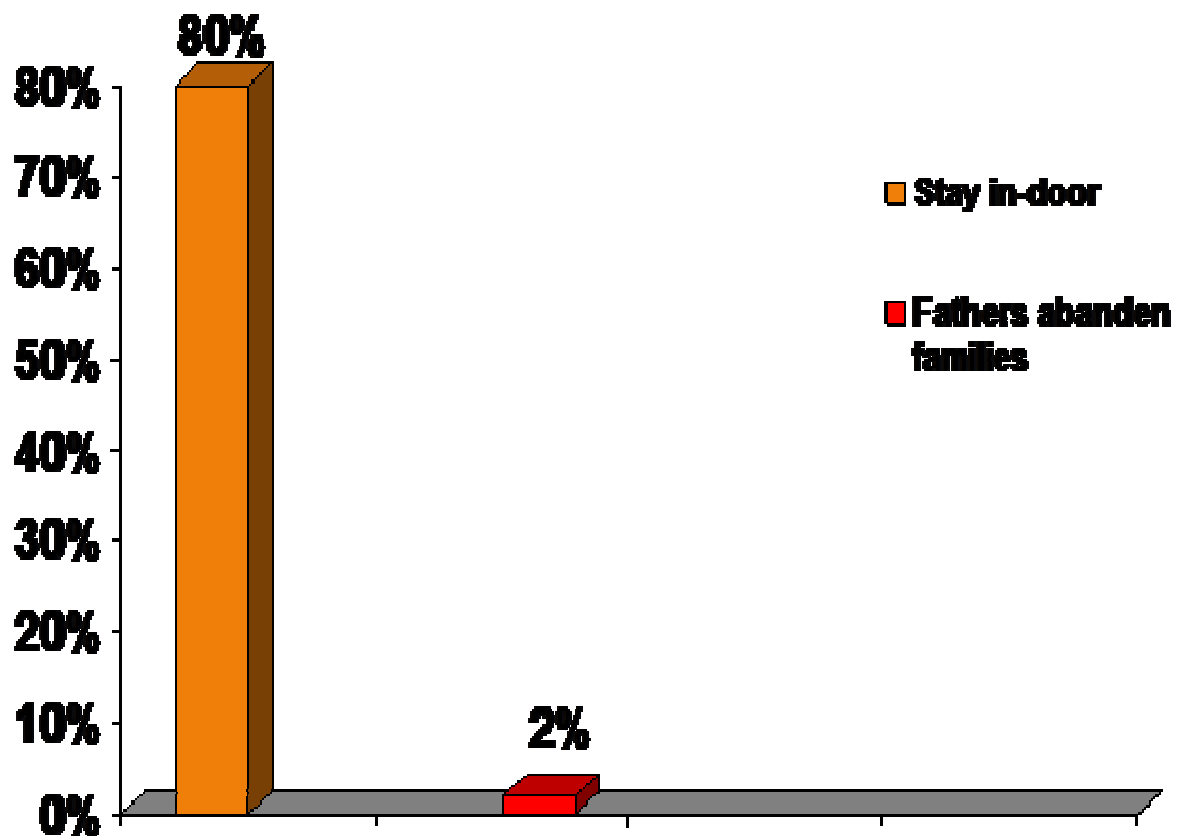


Figure 2. Stay in-doors and father abandon

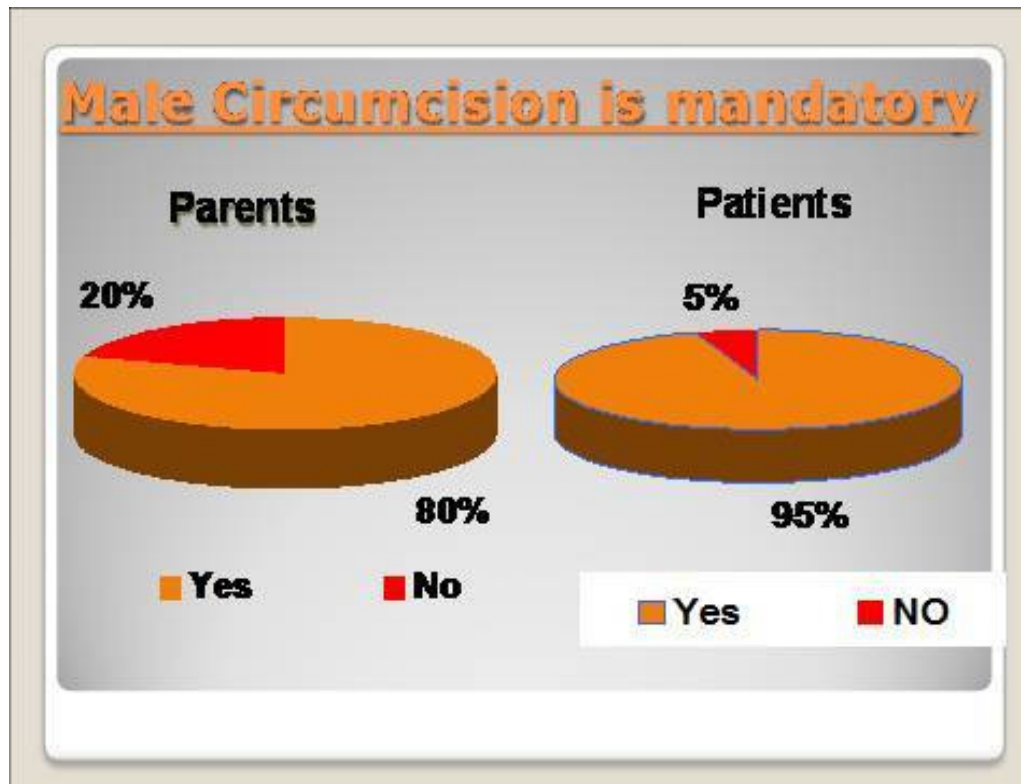


Figure. 3. Male Circumcision is Mandatory.

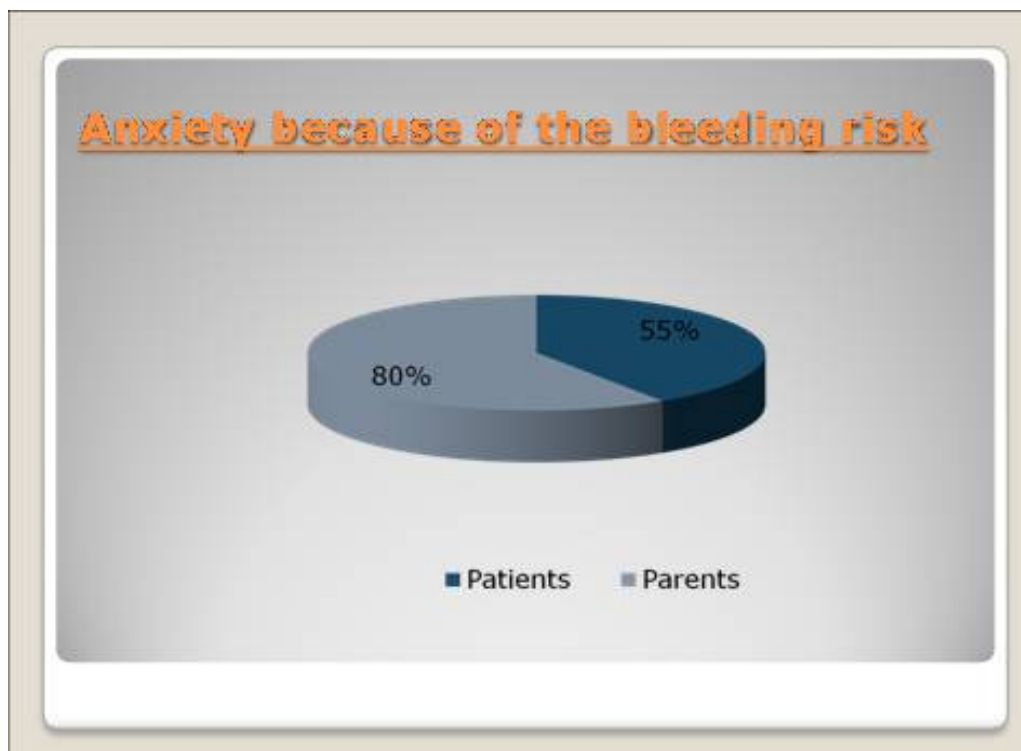


Figure. 4. Anxiety of Bleeding Risk in Haemophilic Patients.



Figure 5. Percentage of Haemophilic Patients admitted to Hospital.

DISCUSSION

In this study all participants consider that circumcision is very important and being un-circumcised result in complete absence and drop out of school which seen in all patients more than ten years and in few of them less than ten years. The majority of patient tended to stay in doors and not participate in any out-door social activities. Some of the fathers abandoned their families due to the cost of the treatment and the social stigma of the daughters being carriers of the disease. This result agreed with other study done in Turkey stated that haemophiliacs and their families consider that circumcision is a very important step to become a member of society and it is a social obligation for men in Turkey. All patients and parents viewed circumcision as a mandatory procedure and they were ready to take the high risk, and this agreed with the same study which stated that, although bleeding risk is high, almost all haemophiliacs would like to be circumcised in Turkish society (Yilmaz et al., 2010).

Most of the patients stay in hospital for 3 days and few of them stay for 7 days. All of them were in a good condition following the Sudanese National Haemophilia society circumcision protocol which is a simple and safe procedure. This agreed with above study in which only three patients (6%) showed bleeding complication and all

were resolved easily. All had at least one excuse from the protocol. Izmir protocol is safe, cheap and easy to carry out (Yilmaz et al., 2010).

This result also agreed with other study in Istanbul stated that transient minimal bleeding was observed in 5 patients and easily responded to factor administration. Moderate edema and hyperemia along the excision line owing to the diathermic effect. Bloodless circumcision with "diathermic knife" is a practical and reliable alternative for boys with hemophilia. Enhancement of local hemostasis using such an alternative device may reduce the need for factor substitution and, accordingly, the cost of circumcision in hemophiliacs, down to 50% (Ihsan et al., 2004). This agreed with our study in which most of the patients following the diathermic protocol and discharged in good condition without a significant bleeding, need less amount of factor costing the average calculated cost of circumcision, while break-through bleeding increase the cost of the procedure by 50-100%.

Following circumcision all parents were happy and satisfied and all patients in school age express their willingness to go back to school, the unstable environment in the house was relieved and some of the fathers who abandoned their families previously return home. This agreed with M. Ihsan in Istanbul who state that excellent patient and family satisfaction was reported (Ihsan et al., 2004).

CONCLUSION

Delayed circumcision has a profound and economic impact on the families. The social effect can easily be relieved by performing circumcision under hospital conditions and to avoid risk of bleeding. All haemophiliacs and their families consider that circumcision is a very important and as a mandatory procedure and they were ready to take the high risk of bleeding. Using the protocol of local hemostasis reduce the need for factor substitution and, accordingly, the cost of circumcision in hemophiliacs, down to 50%. After circumcision all patients and parents were happy and satisfied.

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