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ENT surgery in children with inherited bleeding disorders

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Abstract
Inherited bleeding disorders are frequently considered an absolute contraindication to tonsillectomy and other ENT procedures. Over a 15-year period we have performed ten elective tonsillectomies and five bilateral myringotomies on children with inherited bleeding disorders. All procedures were carried out with the close co-operation of the Haematology Department in the hospital. All patients underwent uneventful surgery. One patient returned after tonsillectomy with a secondary haemorrhage which did not require surgical intervention. We present our team approach to the management of these children and demonstrate that necessary surgical intervention can be undertaken safely in this select group of patients.

Key words: Tonsillectomy; Haemophilia; Von Willebrand's disease

Introduction

Children with inherited bleeding disorders present the otolaryngologist with a clinical dilemma. Primary and secondary tonsillar haemorrhage are well recognised complications in children without coagulation difficulties and thus many surgeons are reluctant to operate on children with known bleeding diatheses. We report on our management of ten patients who have undergone tonsillectomy at our unit over the past 15 years. We also report on our management of five children requiring myringotomy and grommet insertion. Although this latter procedure carries a lower risk of serious haemorrhage, there are few studies published with regard to myringotomy and ventilation tube insertion in patients with abnormalities of haemostasis.

All procedures were undertaken at The National Children's Hospital, Dublin, with the close co-operation of the Haematology Department. This hospital is the national referral centre for children with inherited bleeding disorders. We adhere to a strict protocol which has been developed over the years and we believe this has aided in achieving a low complication rate in the operative and post-operative period.

Methods

We reviewed the charts of all patients with haemostatic disorders who had undergone ENT procedures in our unit since 1980. We did not include patients requiring nasal cautery for recurrent epistaxis in our study. During this period ten patients underwent tonsillectomy, one of whom had bilateral myringotomies at a later date. Four other patients underwent bilateral myringotomies alone. All children were managed according to the following protocol.

Protocol

The patients are admitted on the day prior to surgery, under the joint care of the Haematology and ENT departments. Routine full blood count, platelet count and cross match are ordered. All intramuscular or subcutaneous injections and arterial sampling are forbidden. We also ensure that aspirin or NSAIDS are not proscribed.

All patients have had Hbs Ag, Anti-Hbs, Hep C and Anti-Hep A IgG performed within the previous 12 months and liver function tests and prothrombin time are obtained on all hepatitis-positive patients.

The presence of anti-factor VIII antibodies (inhibitors) is determined pre-operatively and their presence is a contraindication to proceeding with surgery.

All patients with haemophilia A and von Willebrand's disease undergoing tonsillectomy receive Factor VIII concentrate 30 minutes prior to surgery. Patients with haemophilia B similarly receive Factor IX concentrate 30 minutes prior to tonsillectomy. The dose is calculated to produce a 100 per cent factor rise, to a normal post-infusion level. Post-infusion factor levels are obtained at 20 minutes to confirm a 100 per cent rise. If the rise is found to be insufficient further treatment with factor concentrate is administered. Tranexamic acid, which acts as a fibrinolytic agent, is given in a dose of 1 gm

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intravenously followed by 500 mg orally q.i.d. post-operatively. Following surgery, factor VIII or IX concentrate is administered to give a rise of 100 per cent every day for up to 10 days.

Patients with mild haemophilia A (Factor VIII greater than 10 per cent) or mild Von Willebrand's disease undergoing myringotomy are managed with desmopressin (DDAVP), which acts by raising factor VIII levels. A standard dose of 0.4 μg/kg body weight of DDAVP in 100 ml of normal saline is infused over 15 to 30 minutes one hour pre-operatively.

Patients with moderate or severe Haemophilia A or B and patients with severe Von Willebrand's disease undergoing myringotomy are administered factor VIII or IX concentrate prior to myringotomy in accordance with the procedure used for tonsillectomy. A factor rise to 50 per cent is considered adequate cover for myringotomy and grommet insertion and post-operative concentrate is not administered unless bleeding occurs. We do not carry out myringotomy as a day case in these children.

Tonsils are removed using bipolar diathermy dry dissection and no attempt is made to remove the adenoids. All patients undergoing tonsillectomy are placed on prophylactic broad spectrum oral antibiotics.

Results

Patient details are summarised in Table I. Ten tonsillectomies were carried out during the period under review. No excessive peroperative bleeding was encountered and primary haemostasis was easily achieved in all patients. One patient required re-admission for a secondary bleed at home. This bleed occurred on Day seven post-operatively and the parents reported that the patient had coughed up to five to 10 ml of blood. On examination the patient was found to have a small clot in the right tonsillar fossa with slight ooze. He was commenced on an infusion of DDAVP with immediate resolution of bleeding. There was no significant drop in haemoglobin level and the child was discharged four days later.

The hospital stay for patients undergoing tonsillectomy ranged from seven to 14 days, with a mean stay of 10 days. The group undergoing bilateral myringotomy had a hospital stay ranging from two to four days.

Discussion

The potential dangers of operating on patients with inherited bleeding disorders have been known for almost 2,000 years. Early Rabbinical texts forbade the circumcision of the third son of a woman whose previous two sons had died of bleeding from the procedure (Rosner, 1969). Mortality rates ranging from 25 per cent to 50 per cent were quoted in the first half of this century (Craddock *et al.*, 1948) and not until the development of cryoprecipitate and factor concentrates in the 1960s did surgery become a realistic option.

More recent reports have continued to urge extreme caution in the management of these patients resulting in a reluctance to offer surgery (Alusi *et al.*, 1995). While our threshold to operate on these patients is undoubtedly higher than that compared to children without coagulation difficulties we do believe that this surgery can be safely offered in selected cases.

There are few series of tonsillectomy in this group of patients reported in the literature and we believe our group is the largest published group in a paediatric population. Prinsley *et al.* (1993) reported on follow-up results in an older population of 10 tonsillectomies. His results were similar to ours with only one case of secondary tonsillar haemorrhage notably in the eldest patient in his series (31 years).

Tonsillectomy is only offered to patients who have had more than six attacks of acute tonsillitis per year during the previous two years with at least two of these episodes documented by an ENT surgeon. They must also have failed treatment with prophylactic antibiotic therapy. None of the patients in our series suffered from obstructive sleep apnoea. We avoid performing adenoidectomy in these patients as we feel control of primary or secondary haemorrhage from this site could pose significant difficulties. This policy has not resulted in any of our patients failing to gain resolution of their symptoms.

TABLE I

Patient no.	Sex	Age at operation	Diagnosis	Operation	Complication
1	F	4	vWd severe	Tonsillectomy	———
2	M	5	VIII mild	Tonsillectomy	———
3	M	3	VIII mild	Tonsillectomy	———
4	M	6	VIII mild	Tonsillectomy	———
5	M		VIII severe	Tonsillectomy	———
6	M	3	IX mild	Tonsillectomy	———
7	M	6	vWD mild	Tonsillectomy	———
8	F		VIII carrier Prolonged APPT	Tonsillectomy	———
9	F	5	VIII carrier	Tonsillectomy	———
10	M	4	vWD	Tonsillectomy	Secondary tonsillar haemorrhage
11	M	5	IX mild	Bilateral myringotomy	———
12	M	6	VIII severe	Bilateral myringotomy	———
13	M	7	vWD	Bilateral myringotomy	———
14	M	5	VIII mild	Bilateral myringotomy	———
15	M	4	vWD	Bilateral myringotomy	———

Two patients in our series were females with a family history of haemophilia A. Patient 8 had normal factor VIII levels but activated partial thromboplastin time (APTT) was elevated. In this patient no specific factor deficiency was determined and successful diathermy tonsillectomy was undertaken without concentrate cover. Patient 10 however did have reduced factor VIII levels (25 per cent) and was given pre-operative factor VIII cover. Significant post-operative haemorrhage in female carriers is well recognised and therefore it is important not to dismiss the possibility of excessive bleeding in female relatives of haemophiliacs (Carruth, 1969; Harrison and Lammi, 1991).

There are few reports of patients undergoing myringotomy in the recent literature. Scott *et al.* (1988) reported on three patients requiring myringotomy and tube placement. In one of these cases persistent bleeding from the middle ear necessitated removal of the ventilation tubes. Clearly the risk of dangerous haemorrhage in this procedure is significantly less than in those undergoing tonsillectomy. However, patients with severe factor deficiency will bleed excessively following minor trauma and we feel it is necessary to give these patients a 50 per cent rise. Using this protocol we have not experienced any difficulty in the management of this group.

In conclusion we feel that elective ENT surgery can be safely undertaken in children with inherited bleeding disorders providing it is performed in a specialised centre and close co-operation is maintained between the surgical and haematological teams. Elective surgery in such cases avoids prolonged morbidity and suffering and obviates the need for surgery at a later date when the procedure may be more difficult and the risk of bleeding even greater.

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