SUCCESSFUL CATARACT EXTRACTION IN A SEVERE HAEMOPHILIAC*†

BY

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THE life expectancy of haemophiliacs has increased over the last 30 years with the increasing availability of effective therapeutic materials, such as fresh frozen plasma and Factor VIII concentrates. As a consequence, more and more haemophiliacs are surviving to enter the age group in which pathological changes such as senile cataract formation are encountered.

During the past 10 years there have been four previous reports describing cataract extraction in patients with haemophilia or Christmas disease. Mehra, Gupta, and Kapoor (1963) gave no laboratory data confirming the diagnosis of haemophilia. The patient described by de Grósz, Borbély, Szabados, and Rado (1965) had a prolonged clotting time and an abnormal thromboplastin generation test compatible with a defect in Factor IX; this patient was treated with steroids and whole blood. Poweleit (1965) treated his patient with fresh plasma; no haematological details are given. Rubenstein, Albert, and Scheie (1966) described an uncomplicated cataract extraction with complete iridectomy in a man with mild Factor IX deficiency; repair of an iris prolapse was followed by massive vitreous haemorrhage 4 weeks later. This cleared over a 6-month period but the final visual acuity was hand movements only.

The present paper describes successful cataract extraction in a severe haemophiliac.

Case Report

A 68-year-old professional rat catcher was referred with a history of 10 years' progressive loss of vision in the left eye. He had also suffered from bleeding haemorrhoids for many years, and was known to have haemophilia, diagnosis being based on:

- (1) A history of bleeding consisting of occasional epistaxes, bleeding for 3–4 weeks following dental extractions, haematuria on one occasion, and occasional haemarthroses affecting both elbows and both wrists;
- (2) A family history (see Fig. 1, opposite) of bleeding, which showed the pattern of classical sex-linked recessive inheritance;
- (3) *Laboratory Findings:*

Whole blood clotting time (Lee and White method): 8'30" × 4 (normal in this unit = less than 5') (Lee and White, 1913);

Bleeding time (Ivy method): 2', 2'30", 3' (normal = 2'5"–7') (Ivy, Nelson, and Bucher, 1941);

Prothrombin consumption index: 20 per cent. (normal = below 10 per cent.) (Merskey, 1950);

Factor VIII assay by the 2-stage method: 1 per cent. average normal (Biggs, Eveling, and Richards, 1955);

Platelet count: 180,000 per cu. mm.;

Haemoglobin: 7.5 g. per cent.

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FIG. 1.—Family tree showing classical sex-linked recessive inheritance. Arrow refers to patient. Numbers within symbols refer to multiples of unaffected children.

□ = Male ○ = Female. No history of bleeding
 ▨ = Male History of bleeding
 ■ = Male History of bleeding and factor VIII assay

Ophthalmological Findings.—The patient had a mature cataract in the right eye, visual acuity being reduced to hand movements; sight in this eye had been deteriorating over the past 10 years; the eye was somewhat divergent. There was a moderate cataract in the left eye, the best vision being 6/24, with correction; the patient could read N9 slowly with this eye. Intra-ocular pressures were within normal limits.

Treatment and Progress.—The patient was admitted to hospital primarily for cataract extraction, but the haemorrhoids were also to be treated. Packed cells from three bottles of blood were transfused to raise the haemoglobin to a safe level for surgery. The following day fresh plasma was infused and the haemorrhoids were injected, and 2 days later the cataract extraction was carried out. Porcine Anti-Haemophilic Globulin (AHG) (6,400 units) was infused immediately before operation. Subsequent doses of porcine AHG were given with the aim of maintaining the Factor VIII level above 25 per cent., up to and including the tenth day post-operatively.

One unit of AHG is equivalent to 1 ml. plasma with 100 per cent. Factor VIII activity. In this study porcine AHG manufactured by Maws was used; at the time of writing, their product was still calibrated according to the old Oxford unit, *i.e.* 1 unit is equivalent to 4 ml. plasma of 100 per cent. Factor VIII activity. In this paper units have been expressed as new units.

Details of this therapy and the daily Factor VIII levels of the patient immediately before and after the doses are given in Fig. 2.

FIG. 2.—Details of therapy.

Operation.—Cataract extraction (Mr. J. P. F. Lloyd) was performed under general anaesthesia; a facial block was performed but no retrobulbar injection given. Lid sutures were inserted without previous anaesthetic injection.

A von Graefe section emerging on the corneal side of the limbus was made without conjunctival flap in the expectation of avoiding the limbal vessels. A solution of thrombin was sprayed into the area, but no abnormal bleeding occurred. Two corneo-scleral sutures of virgin silk were pre-placed in the wound. Iridectomy was not performed.

The superior iris being held retracted, the lens was extracted in its capsule without incident by means of a Cryostat (Pierse). Pilocarpine 3 per cent. was instilled into the wound and the sutures tied. Air was not injected. Neomycin and Pilocarpine 3 per cent. drops were applied, and a vaseline gauze, pad, and bandage.

At the first post-operative dressing after 48 hrs., the anterior chamber was shallow but the eye was otherwise satisfactory. The anterior chamber remained shallow and the eye hypotonic for 5 days, and a choroidal detachment was observed. Diamox (250 mg. four times daily) and potassium bicarbonate (500 mg. twice daily) were given by mouth for 5 days.

During the next few days the anterior chamber deepened. At no time after the operation did bleeding take place.

Result.—The final visual acuity was 6/6 and N5, with aphakic correction.

Discussion

In this case porcine AHG was used as it enables a higher level of Factor VIII to be maintained without the infusion of large volumes of fluid daily to a 68-year-old man. This regime has been used for major surgery in severe haemophiliacs since 1955 (Macfarlane, Mallam, Witts, Bidwell, Biggs, Fraenkel, Honey, and Taylor, 1957). The main disadvantage of porcine AHG is that, since it is a foreign protein, patients are liable to develop antibodies. However, at the age of 68, it is hoped that this patient may not require further major surgery and in view of the extreme importance for the success of the operation of preventing any bleeding whatsoever the use of animal protein seemed justified.

Summary

A case of a successful cataract extraction in a severe haemophiliac is presented with full haematological investigations. The final visual acuity in the operated eye was 6/6, with aphakic correction.

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