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Adenoidectomy in a girl with haemophilia

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Abstract

Haemophilia A is a sex-linked inherited disease in which those affected are usually males, and females are usually asymptomatic carriers. This paper presents a haemophilia A carrier who has a low factor VIII level first, to remind readers that females can have low factor VIII levels with consequent increased tendency to bleed; second, to stress the importance of routine questioning regarding a bleeding tendency in the patient or the patient's family and third, to illustrate the importance of pre-operative diagnosis to allow adequate correction of the defect so that surgery such as adenoidectomy can be undertaken with safety in such a patient.

Case report

An 11-month-old girl was referred to one of the authors (HH) because of nasal blockage, obstructed breathing at night and sleep disturbance. Clinical ear, nose and throat examination was normal except for the presence of mouth breathing and noise on respiration suggestive of nasal obstruction. A diagnosis of adenoid hypertrophy was made and was supported by a lateral X-ray examination of the post-nasal space.

Adenoidectomy was recommended despite the patient's age because of the respiratory obstruction and sleep disturbance. However, routine questioning at the time of presentation had revealed that the mother's brother suffered from haemophilia and that the patient herself bruised easily. Although inherited bleeding disorders are more common in males, females can be affected and she was therefore referred to the Haematology Department of the Royal Alexandra Hospital for Children, (RAHC) Sydney, Australia for pre-operative assessment and operative planning.

At RAHC it was discovered that, because of the family history, blood tests had been performed there on the patient when she was six months of age; they had revealed a factor VIII level of 22 per cent of normal (Table I). A diagnosis of haemophilia carrier had been made but the mother did not recall this. Operative preparation was by the infusion of factor VIII prior to operation and post-operatively twice daily to maintain factor VIII levels above 50 per cent until significant risk of bleeding had passed.

Adenoidectomy was performed and blood loss of 20 ml was measured. There was no post-operative respiratory difficulty or significant bleeding. Factor VIII as cryoprecipitate was administered for seven days after which the patient was discharged from hospital.

TABLE I
LOW FACTOR VIIIC AND LOW FACTOR VIIIC: AG RATIO CONSISTENT WITH HAEMOPHILIA A CARRIER

	Patient	Mother	
Factor VIIIc	22%	35%	(N50–200%)
Factor VIII Ag	68%	70%	(N50–200%)
Ratio C/Ag	0.32	0.5	

At follow-up two weeks after the operation, the patient had a clear nasal airway and was no longer mouth breathing. The mother stated that there had been no bleeding since discharge from hospital and that the patient's sleep was undisturbed.

Telephone enquiry eleven months after surgery confirmed that the breathing improvement had been maintained but revealed that shortly after the last attendance, the patient had sustained a minor head injury. This had resulted in a haematoma prompting presentation to a hospital casualty where junior medical staff had expressed disbelief at the possibility of a bleeding tendency in a female due to haemophilia carrier status. There were no sequelae.

Discussion

Although the average factor VIII level in carriers of haemophilia is approximately 50 per cent of normal, carriers can have a wide range of factor VIIIc activity, in accordance with Lyon's hypothesis of random inactivation of the X chromosome (Wintrobe *et al.*, 1974). Data presented by Shen (1982) showed 48.6 per cent of carriers to have a factor VIIIc level below 40 per cent and 18.9 per cent below 25 per cent which is similar to the author's experience. Rizza (1976) states that factor VIII levels below 25 per cent are associated with severe bleeding following surgery and Carruth (1969) reported bleeding after tonsillectomy in an unrcognized carrier who had a factor VIII level of 28 per cent. Livingstone (1965) stated that a factor VIII level continuously above 25 per cent until healing was well advanced was satisfactory for most operations but that tonsillectomy may require higher levels; he mentioned a tonsillectomy patient who bled with a level just over 40 per cent. Adenoidectomy is as likely to cause bleeding as is tonsillectomy (Capper and Randall, 1984); the authors therefore believe that the subject of this report, who had a factor VIII level of 22 per cent, would have been very likely to suffer from serious bleeding if adenoidectomy had been performed without correction of her haemostatic defect. Her development of a haematoma after minor head injury also supports this contention.

The diagnosis of haemophilia A can be so occult that it may not be made until well into adult life (Harrison *et al.*, 1972) and may only be revealed when a major test of haemostasis is faced such as significant trauma or surgery. Sufferers of mild inher-

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ited disorders of haemostasis are particularly at risk of being subjected to operative procedures (especially tonsillectomy) without their bleeding tendency being diagnosed beforehand (Kerr, 1963; Livingstone, 1965) and therefore without preoperative correction of their haemostasis. The authors believe that females may be particularly at risk of this because some doctors appear unaware that females may be affected by inherited bleeding disorders, as demonstrated by the attitude shown by casualty staff towards this patient. The figures quoted from Livingstone (1965), Shen (1982) and Rizza (1986) in the previous paragraph suggest that an unrecognized carrier of haemophilia has almost a 20 per cent chance of post-surgical bleeding and that almost 50 per cent are predisposed to bleed after tonsillectomy and therefore adenoidectomy (Capper and Randall, 1984).

Questioning as to abnormal bleeding in the patient or relatives before surgery is more likely to reveal a bleeding disorder than are routine screening tests (Livingstone, 1965). A history suggestive of a bleeding disorder should prompt thorough investigation.

Whilst surgery on a haemophiliac should not be undertaken without good cause and adequate preparation in a suitable centre, necessary surgery can generally be performed without undue risk from bleeding (Livingstone, 1965; Lusher, 1987). The 'bleeding history' may be vital in making the requisite preoperative diagnosis, especially in mild haemophiliacs or in carriers. It is therefore wise to take such a history routinely in all patients (even females) before surgery to lessen the risk of a diagnosis of haemophilia being made when the patient is already bleeding dangerously after unprepared surgery.

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