Vestibular closure with a silastic obturator – an alternative to Young's procedure in bleeding diathesis*

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SUMMARY *Epistaxis is a common and difficult problem to manage in patients with bleeding disorders. We present a case of recurrent epistaxis in a patient with Bernard-Soulier syndrome (a platelet disorder) and describe a non-invasive but effective method of closing the nasal vestibule using a silastic obturator thus preventing the drying effects of airflow on the nasal mucosa which may precipitate epistaxis in patients with a bleeding diathesis.*

Key words: epistaxis, Bernard-Soulier syndrome, vestibular closure, non-surgical

INTRODUCTION

Bernard-Soulier syndrome is a rare platelet disorder that gives rise to a bleeding diathesis. It is usually inherited as an autosomal recessive trait and hence consanguinity has a role in its transmission. This disorder is characterised by thrombocytopenia, giant platelets, a failure of the platelets to undergo selective von Willebrand factor-dependent platelet interactions and defective interaction between platelets and thrombin. The defect in the platelets is due to abnormalities within the platelet glycoproteins GPIb/IX/V complex, which is a major receptor for von Willebrand factor. Epistaxis is the most common symptom of Bernard-Soulier syndrome (70%), followed by ecchymosis, menometrorrhagia, gingival haemorrhage and gastrointestinal bleeding, less frequently seen symptoms are post-traumatic bleeding, haematuria and cerebral or retinal haemorrhage (Bentler et al, 1995).

CASE REPORT

This 32 year old Asian male began to suffer from intermittent epistaxes at the age of 4 years. Three years later it was noticed that he had a tendency to bruise easily and following investigation, a diagnosis of Bernard-Soulier syndrome was made. A history of parental consanguinity was obtained; his parents being first cousins. He had previously been resident in London and was under the care of a local haematologist and otolaryngologist for his recurrent nose bleeds. The management of his nose bleeds varied according to their severity, minor epistaxes usually settled with rest and local pressure to the nose, whereas for bleeds that were more persistent and of greater severity he underwent nasal packing with BIPP by one of his relatives and these packs were later removed in the local ENT department. In addition he was administered tranexamic acid and Desmopressin (DDAVP) to facilitate his clotting mechanism. His usual platelet count was around 30 x 10^9 / L and if platelet transfusions were required these had to be HLA- matched because of the development of antibodies. Since moving upto Manchester in the last year, he has been under the joint care of the haematology department and our unit at the Manchester Royal Infirmary. The frequency and intensity of his nose bleeds had increased and one occasion he underwent anterior and posterior nasal packing and had to be electively ventilated for a few days. In view of the increasing severity of epistaxis it was decided to fit him with a silastic obturator so as to block the passage of air into his nose thus preventing his nasal mucosa from drying and bleeding (Figures 1 & 2). This mode of treatment has been very

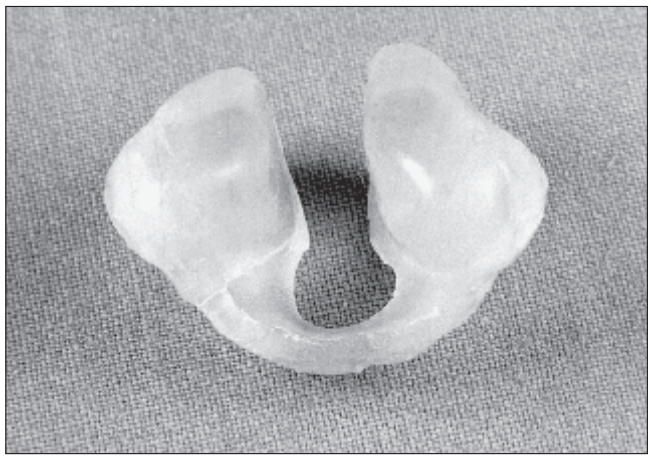

Figure 1. The silastic obturator.

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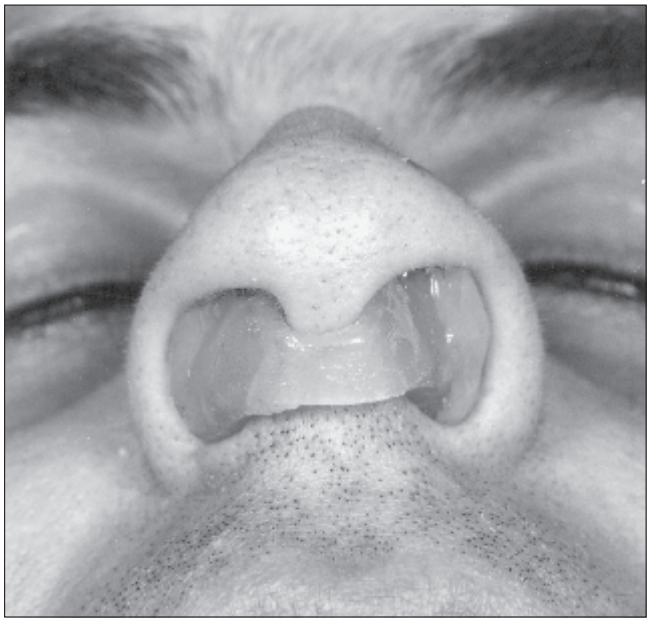

Figure 2. Case with the silastic obturator in situ.

successful and in the last six months, since he started wearing the obturator, he has had no further bleeding, which is the longest period he has ever been free of epistaxis.

TECHNIQUE

An impression of the nasal vestibules is taken with Otoform-AK2 (an addition-vulcanising silicone impression material) which is normally used in making impressions of the ear for the manufacture of hearing aid moulds. After cleaning the nasal vestibules with cotton wool, the posterior end of the vestibule is blocked using foam impression stops. The silicone impression material is injected to fill both the vestibules and a band of the material is left across the columella to join the two sides. After the material has set it is removed and the definitive obturator is made of soft silastic a polymer of dimethyl-siloxane by a commercial hearing aid mould manufacturer.

DISCUSSION

The Youngs procedure is an operation performed to achieve complete closure of the nasal vestibule thus preventing the drying effects of nasal airflow. It was originally designed and has since been successfully used in the management of atrophic rhinitis (Young , 1967; Sinha et al., 1977). Epistaxis in bleeding diatheses is thought to be secondary to trauma to the nasal mucosa caused by the drying effects of air flow, and thus bleeding is prevented by nasal vestibular closure. The Youngs procedure has been shown to be very successful in the prevention of epistaxis in cases of hereditary haemorrhagic telangiectasia (HHT) where all other forms of treatment have unsuccessful (Gluckman and Portugal, 1994; Hosni and Innes, 1994; Lund and Howard 1997).

Most patients suffering from HHT are apprehensive of undergoing a Youngs procedure with the fear that nasal closure may cause respiratory problems and also of the implications if further bleeding were to occur (Lund and Howard 1997). In view of this latter point and that the surgery itself may induce uncontrollable bleeding in a patient with a clotting mechanism disorder, we felt that a Youngs procedure was contraindicated. We therefore considered a prosthetic alternative and designed the silastic obturator as described above. This technique has also been used in the management of patients with atrophic rhinitis (Lobo et al., 1998), again to prevent airflow with its drying effects.

The authors feel that this non-invasive technique could be used as a permanent alternative to Youngs procedure in patients in a variety of bleeding disorders, whilst in individuals due to undergo vestibular closure, it may be utilised prior to surgery to increase patient confidence and demonstrate the efficacy of the planned procedure.

CONCLUSSIONS

In cases with a bleeding or clotting disorders where epistaxis can be troublesome and potentially life threatening, closure of the nasal vestibule using a silastic obturator is a useful option for prevention of epistaxis. It is a non-invasive, effective, cheap and can easily be made in any ENT department.

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