Published in final edited form as:

Int Forum Allergy Rhinol. 2016 May ; 6(5): 544–548. doi:10.1002/alr.21703.

Treatment of severe refractory epistaxis in hereditary hemorrhagic telangiectasia using a two-flap nasal closure method

Benjamin H. Timmins, MS¹,

University of Utah School of Medicine, SLC, UT, USA

Benjamin N. Hunter, BS,

University of Utah School of Medicine, SLC, UT, USA

Kevin F. Wilson, MD, and

University of Utah Division of Otolaryngology and Head & Neck Surgery, SLC, UT, USA

P. Daniel Ward, MS, MD

University of Utah Division of Otolaryngology and Head & Neck Surgery, University of Utah Facial Plastic and Reconstructive Surgery, SLC, UT, USA

Abstract

Background—Nasal closure has been shown to effectively manage severe epistaxis refractory to other treatments in patients with hereditary hemorrhagic telangiectasia (HHT). The nasal closure procedure may be underutilized due to its surgical complexity and flap breakdown.

Methods—Retrospective review of thirteen HHT patients treated for severe epistaxis with nasal closure between 2005 and 2013. Operating room (OR) time, need for revision surgery, pre- and post-procedure epistaxis severity score (ESS), complete blood count values, and Glasgow Benefit Inventory (GBI) questionnaire results were collected for each patient. The technique is described. We characterize a typical nasal closure patient and compare outcomes based on our experience with the traditional three-flap closure and a simplified two-flap nasal closure procedure.

Results—The average candidate for nasal closure in this series had an ESS of 7.88, Hgb of 8.3 g/dL, and received multiple transfusions, iron therapy, and cautery/coagulation procedures. Average ESS subsequent to nasal closure using the two flap method is 0.92 and mean GBI score is 56.3. Comparison of five patients who underwent the traditional three-flap nasal closure procedure and eight patients receiving the two flap nasal closure showed no significant difference in post-op ESS or GBI metrics. Mean operating room times of the traditional and simplified methods were

¹All correspondence, reprint requests, and proofs will please be sent to this author. Contact info: ben.timmins@hsc.utah.edu, University of Utah School of Medicine, 30 N 1900 E, Salt Lake City, UT 84132. Phone: 435-764-4932, Fax: 801-585-5744.

Conflict of Interest Statement

No authors of this study have any conflicts of interest.

Financial Disclosures

None of the authors report financial disclosures

Data Presentation

Poster Presentation Combined Otolaryngology Spring Meeting, American Rhinologic Society, Las Vegas, NV, USA, May 14-18, 2014.

3.12 hours and 1.44 hours ($p=0.0001$). Mean time to first revision for eight nasal closure patients is 21.5 months.

Conclusion—In short-term follow-up, the two-flap procedure has shown comparable effectiveness with significantly reduced complexity and operative time compared to the traditional nasal closure method.

Keywords

Anemia; Arteriovenous Malformations; Operative Time; Patient Satisfaction; Quality of Life; Reoperation; Telangiectasia, Hereditary Hemorrhagic

Introduction

Hereditary hemorrhagic telangiectasia (HHT) is an autosomal dominant vascular dysplasia characterized by the presence of multiple arteriovenous malformations (AVMs) affecting at least 1/8,000 people.¹ Mucocutaneous telangiectasias can have a significant negative impact on quality of life as they often rupture causing epistaxis. In many individuals turbulent nasal airflow is traumatic enough to damage these telangiectasias and cause epistaxis. Severity of epistaxis ranges from infrequent minor nosebleeds to frequent large-volume hemorrhages leading to severe anemia and transfusion dependence.² Treatment options for severe epistaxis may include chemical or electrical cautery, laser photocoagulation³⁻⁵, hormonal therapy, antifibrinolytics, embolization, septodermoplasty⁶⁻⁷, and anti-VEGF injections (Avastin).⁸⁻⁹ After initial treatment, telangiectasias return over time requiring re-treatment to again temporarily reduce epistaxis.

Surgical closure of the nares has demonstrated effectiveness in preventing epistaxis¹⁰⁻¹⁵ in patients who do not respond to other treatments or fail to achieve long-term resolution. Nasal closure, or Young's procedure, was first described for atrophic rhinitis in 1967¹⁶ and applied to HHT patients for epistaxis in 1991.¹⁰ The current standard Young's procedure is a modification of the original HHT nasal closure first reported by Lund and Howard.¹² This modified closure may be combined with lateral alotomy to create an additional internal flap closure. It is our experience that the described three-flap nasal closure method is complex and often results in wound breakdown and fistula formation at the flap closure trifurcation point. We have applied a modification to the procedure using two nasal vestibular flaps without a lateral alotomy to achieve nasal closure. Here we describe the technique and our initial experience with the first eight patients.

Materials and Methods

Surgical technique

The traditional three-flap modified Young's procedure has been described previously.¹² The procedure is usually combined with laser photocoagulation of the telangiectasias prior to nasal closure. The two-flap procedure begins with a circumferential incision through the skin at the mucocutaneous junction within the nasal vestibule. The skin is then back elevated over the cartilage to create the two skin flaps. Sufficient elevation of the flaps is required for

a tension-free closure. The two flaps are sutured in a single-layer closure using 4.0 Chromic suture (Figure 1). Intraoperative photographs are seen in Figure 2.

Data collection and analysis

The charts of all HHT patients (n=13) who underwent bilateral nasal closure at the University of Utah Hospital from 2005-2013 were analyzed after Institutional Review Board approval was obtained. The cohort was divided into two groups: those who received the three-flap method (2005-2011) and those who received the two-flap method (2010-2013). Telephone interviews were conducted for further data collection. Data analyzed included operating room (OR) time, need for revision surgery, pre- and post-procedure Epistaxis Severity Score (ESS) and Glasgow Benefit Inventory (GBI) questionnaire results. ESS is based on patient responses to six standard questions to quantify the severity of epistaxis over a three-month period.¹⁷ ESS scores range from 0 to 10 with 0-3 representing mild epistaxis, 3-7 moderate epistaxis, and 7-10 severe epistaxis. The GBI questionnaire¹⁸ was developed to quantify patient benefit after an intervention and consists of 18 questions that were adapted for the Young's procedure. Calculated GBI score ranges from -100 to 100 with -100 indicating much worse, 0 no change, and 100 much better. Laboratory values collected include hematocrit (Hct), hemoglobin (Hgb), mean corpuscular volume (MCV), and ferritin. When multiple laboratory and questionnaire results were available, the most abnormal value was used to indicate the severity of the patient's condition.

ESS, GBI score and OR time for the two groups were compared using a two-sample, two-tailed t-test with pooled variance assumption. Calculation of the t-test p values were made using MathWorks MATLAB R2013a. Simultaneous testing of post-operative ESS, GBI, and operative time necessitates the use the Bonferroni correction; statistical significance becomes $\alpha/3$ or 0.0167.

Results

Of the thirteen bilateral nasal closure patients, five received a three-flap nasal closure and eight received the two-flap nasal closure. None of the patients receiving either three or two-flap closure underwent alotomy. Seven of eight patients who received two-flap closure underwent laser photocoagulation of telangiectasias at the time of nasal closure increasing operative time. Average patient age was 63.4 years (33-75 years), 9 male and 4 female. Mean postoperative follow up time was 49 months (13.2-110.2 months). Mean pre-operative Hct, Hgb, and ferritin values were 26.8% (range, 20.3-39.9%), 8.3 g/dL (range, 6.5-13.2 g/dL), and 60.6 ng/dL (range, 2-321 ng/dL), respectively. Where data are available the mean pre-operative ESS was 7.88 (range, 4.53-10.0) with average ESS subsequent to nasal closure of 0.97 (range, 0-4.46). Table 1 provides a summary of the patient data used for comparison.

All of the three-flap nasal closure patients have required a revision due to closure dehiscence and subsequent resumed epistaxis, one of whom has received four revisions. Three of the two-flap nasal closure patients required revision. The mean time to first revision was 21.5 months (range, 1.8-59.4 months). Of the thirteen patients who underwent nasal closure, six have developed pinpoint openings, two of which are bilateral. Two of the six patients report mild intermittent bleeding from these openings.

Figure 3 compares post-operative ESS, GBI and OR times of the two nasal closure methods. The most recent values for the three flap and two flap procedures are: mean ESS of 1.07 (range, 0-4.46) and 0.92 (range, 0-3.77) $p=0.87$, mean GBI scores of 73.3 (range, 55.6-100) and 56.3 (range, 8.3-83.3) $p=0.18$, and mean OR times of 3.12 hours (range 2.75-3.68 hours) and 1.44 hours (range, 0.93-2.32 hours) $p=0.0001$.

Discussion

In our population both the three-flap and two-flap closure methods appear equally efficacious in reducing epistaxis, with no difference in ESS and GBI. The two-flap technique, however, requires significantly less operative time with decreased technical difficulty compared to the traditional method. Our data also indicates a possible lower incidence of dehiscence requiring surgical revision with the outlined two-flap method when compared to three-flap nasal closure. Because dehiscence can occur long after the procedure, extended follow up will be needed to see if this difference continues long-term.

Based on previously shown efficacy¹⁰⁻¹⁵ of reduction in epistaxis severity and increased quality of life it is our belief that nasal closure is underutilized. A commonly heard phrase postoperatively from our patients is "I have my life back." This stems from the fact that these patients no longer have the uncertainty of severe, spontaneous, uncontrolled epistaxis. As such we advocate discussion of nasal closure as a treatment option in any HHT patient with chronic anemia due to severe, chronic epistaxis that has failed more conservative treatment strategies. From our experience patients with intranasal AVMs or large non-discrete telangiectasias are less likely to have satisfactory results with laser coagulation and cauterization. It is therefore our recommendation that these patients should also be informed of the potential benefits of nasal closure.

Careful selection of appropriate candidates is essential to achieving a high level of efficacy and patient satisfaction. None of the patients who have undergone nasal closure at our institution have requested reversal of the procedure, we believe, in part due to thorough discussion of expected outcomes and side effects. Side effects encountered may include wound breakdown with possible need for revision surgery, development or worsening of obstructive sleep apnea, anosmia, decreased taste, and obligate mouth breathing leading to mouth dryness. Currently six of our thirteen nasal closure patients presented here have developed pinpoint dehiscence along the line of closure. Four of these six have not experienced an increase in bleeding indicating that a small air leak may not produce sufficient desiccation to rupture the telangiectasias and can be managed expectantly. Posterior epistaxis can occur and may be difficult to treat due to loss of access via the nares as reported by the case of a 70 year old woman who required multiple transfusions, intubation, bilateral embolization, and reversal of nasal closure.¹⁹ Thankfully this is rare, and our experience has been nearly immediate cessation of bleeding after the procedure. In addition to discussion of these side effects we recommend a trial of functional closure using petroleum jelly-soaked cotton balls placed in the nares. This allows the patient to experience the effects of closure before undergoing the permanent procedure.

Shortcomings of this analysis include a small sample size, recall bias, short follow up, and lack of completeness in the medical record. Often our patients receive work up and treatments outside our hospital network, making it difficult to accurately track all laboratory values and procedures. Patients were contacted by telephone to obtain up-to-date follow up regarding epistaxis severity and quality of life data to decrease this bias.

Conclusion

Nasal closure is an excellent treatment for severe epistaxis in properly-selected HHT patients. In short-term follow-up, the two-flap procedure has shown comparable effectiveness with significantly reduced complexity and operative time compared to the three-flap procedure. We have found high patient satisfaction with the procedure as long as physicians provide clear expectations regarding the side effects and benefits. While there is no clear algorithm for performance of nasal closure, an average candidate patient is presented.

Acknowledgements

Medical Student Research Program at the University of Utah School of Medicine

Grant Title: Short-Term Training: Students in Health Professional Schools

Funding Source: NIH/NHLBI

Grant #: T35 HL007744

PI: Jerry Kaplan

This work was supported by grants from the National Institutes of Health: R01NS075168

References

1. Lesca G, Olivieri C, Burnichon N, et al. Genotype-phenotype correlations in hereditary hemorrhagic telangiectasia: Data from the French-Italian HHT network. *Genet. Med.* 2007; 9(1):14–22. [PubMed: 17224686]
2. Bayrak-Toydemir P, Mao R, Lewin S, McDonald J. Hereditary hemorrhagic telangiectasia: an overview of diagnosis and management in the molecular era for clinicians. *Genet. Med.* 2004; 6(4): 175–91. [PubMed: 15266205]
3. Jørgensen G, Lange B, Wanscher JH, Kjeldsen AD. Efficiency of laser treatment in patients with hereditary hemorrhagic telangiectasia. *Eur. Arch. Otorhinolaryngol.* 2011; 268(12):1765–70. [PubMed: 21706320]
4. Karapantzos I, Tsimpliris N, Goulis DG, et al. Management of epistaxis in hereditary hemorrhagic telangiectasia by Nd:YAG laser and quality of life assessment using the HRQoL questionnaire. *Eur. Arch. Otorhinolaryngol.* 2005; 262(10):830–3. [PubMed: 15739086]
5. Harvey RJ, Kanagalingam J, Lund VJ. The impact of septodermoplasty and potassium-titanyl-phosphate (KTP) laser therapy in the treatment of hereditary hemorrhagic telangiectasia-related epistaxis. *Am. J. Rhinol.* 22(2):182–7. [PubMed: 18416977]
6. Fiorella ML, Ross D, Henderson KJ, White RI. Outcome of septal dermoplasty in patients with hereditary hemorrhagic telangiectasia. *Laryngoscope.* 2005; 115(2):301–5. [PubMed: 15689755]
7. Levine CG, Ross DA, Henderson KJ, Leder SB, White RI. Long-term complications of septal dermoplasty in patients with hereditary hemorrhagic telangiectasia. *Otolaryngol. Head. Neck Surg.* 2008; 138(6):721–4. [PubMed: 18503842]

8. Karnezis TT, Davidson TM. Efficacy of intranasal Bevacizumab (Avastin) treatment in patients with hereditary hemorrhagic telangiectasia-associated epistaxis. *Laryngoscope*. 2011; 121(3):636–8. [PubMed: 21344445]
9. Karnezis TT, Davidson TM. Treatment of hereditary hemorrhagic telangiectasia with submucosal and topical bevacizumab therapy. *Laryngoscope*. 2012; 122(3):495–7. [PubMed: 22147664]
10. Brooker DS, Cinnamond MJ. Young's procedure in the treatment of epistaxis. *J. Laryngol. Otol.* 1991; 105(10):847–8. [PubMed: 1753198]
11. Gluckman JL, Portugal LG. Modified Young's procedure for refractory epistaxis due to hereditary hemorrhagic telangiectasia. *Laryngoscope*. 1994; 104(9):1174–7. [PubMed: 8072369]
12. Lund VJ, Howard DJ. Closure of the nasal cavities in the treatment of refractory hereditary haemorrhagic telangiectasia. *J. Laryngol. Otol.* 1997; 111(1):30–3. [PubMed: 9292127]
13. Richer SL, Geisthoff UW, Livada N, et al. The Young's procedure for severe epistaxis from hereditary hemorrhagic telangiectasia. *Am. J. Rhinol. Allergy*. 26(5):401–4. [PubMed: 23168156]
14. Hosni AA, Innes AJ. Hereditary Haemorrhagic telangiectasia: Young's procedure in the management of epistaxis. *J. Laryngol. Otol.* 1994; 108(9):754–7. [PubMed: 7964136]
15. Ichimura K, Kikuchi H, Imayoshi S, Yamauchi T, Ishikawa K. Are patients with severe epistaxis caused by hereditary hemorrhagic telangiectasia satisfied with nostril closure surgery? *Auris. Nasus. Larynx*. 2012; 39(1):59–64.
16. Young A. Closure of the nostrils in atrophic rhinitis. *J. Laryngol. Otol.* 1967; 81(5):515–24. [PubMed: 6024992]
17. Hoag JB, Terry P, Mitchell S, Reh D, Merlo CA. An epistaxis severity score for hereditary hemorrhagic telangiectasia. *Laryngoscope*. 2010; 120(4):838–43. [PubMed: 20087969]
18. Robinson K, Gatehouse S, Browning GG. Measuring patient benefit from otorhinolaryngological surgery and therapy. *Ann. Otol. Rhinol. Laryngol.* 1996; 105(6):415–22. [PubMed: 8638891]
19. Ting JY, Remenschneider A, Holbrook EH. Management of severe epistaxis after Young's procedure: a case report. *Int. Forum Allergy Rhinol.* 2013; 3(4):334–7. [PubMed: 23109496]

Figure 1.

An incision is made at the mucocutaneous junction and two flaps are back elevated within the anterior nasal vestibule. The flaps are sewn together with a single-layer closure.

Figure 2.

Intraoperative photographs of two-flap nasal closure: A - Marking for incision at mucocutaneous junction within the nasal vestibule, B - Elevation of the lateral nasal flap, C - Elevation of the medial nasal flap, D - Suture closure of the medial and lateral nasal flap, D - Completed nasal closure close up view, F – Completed nasal closure.

Figure 3.

Comparison of most recent ESS, GBI score and operative time. P values are calculated using a two-sample, two tailed, t-test with pooled variance.

Table 1

Summary of nasal closure patients showing follow-up time after initial nasal closure, time to closure revisions from initial closure, pre-operative hemoglobin (Pre-OP HgB), pre-operative ESS (Pre-OP ESS), most recent ESS (Current ESS), most recent GBI score (GBI), and time spent in the operating room (t_{or}) for the initial nasal closure procedure.

Three-Flap Young's Procedure							
Patient	Follow-up (months)	Revision/s (months)	Pre-OP HgB	Pre-OP ESS	Current ESS	GBI	t_{or} (hours)
1	110.2	59.4	9.9	n/a	0.00	66.7	3.68
2	106.8	22.6	7.3	n/a	0.00	100.0	n/a
3	97.3	45.6	10.9	n/a	0.00	75.0	3.15
4	91.8	20.5	7.2	n/a	4.46	69.4	2.90
5	78.9	1.8, 12.4, 18.4, 56.8	n/a	n/a	0.89	55.6	2.75

Two-Flap Young's Procedure							
Patient	Follow-up (months)	Revision/s (months)	Pre-OP HgB	Pre-OP ESS	Current ESS	GBI	t_{or} (hours)
6	32.2	n/a	13.2	n/a	0.00	72.2	1.00
7	24.2	n/a	7.9	10.0	1.63	58.3	1.20
8	21.7	3.2	6.5	9.09	0.00	69.4	1.45
9	16.1	15.4	7.0	8.99	0.00	83.3	1.40
10	15.7	3.7	7.7	5.25	1.92	38.9	1.40
11	14.4	n/a	6.7	9.09	0.00	55.6	2.32
12	13.9	n/a	6.8	8.19	3.77	8.3	1.85
13	13.2	n/a	8.9	4.53	0.00	63.9	0.93

ESS: Epistaxis Severity Score, GBI: Glasgow Benefit Inventory, t_{or} : Initial nasal closure procedure duration.