



The Effects of Nasal Closure on Quality of Life in Patients with Hereditary Hemorrhagic Telangiectasia

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Introduction: Epistaxis is the most common symptom of hereditary hemorrhagic telangiectasia (HHT). Complete nasal closure is one of the treatment options for patients with severe, intractable epistaxis. In our experience, this surgery can be life changing in a positive sense; but many patients as well as their physicians understandably fear that such a procedure will diminish certain aspects of quality of life (QOL).

Methods: Case-control study of HHT patients treated at the University of Utah HHT Center of Excellence with and without nasal closure from January 2005 to January 2016. Patients were matched according to epistaxis severity. Each included patient was issued three surveys: Epistaxis Severity Score (ESS), the Pittsburg Sleep Quality Index (PSQI), and the Nasal Obstruction Symptom Evaluation (NOSE).

Results: After treatment, the mean PSQI and NOSE scores were not significantly different between the two groups. However, the mean ESS score in the nasal closure group was significantly lower at 1.10 compared to the severe epistaxis group with a mean score of 3.99 ($P = .027$).

Conclusion: The results of this study demonstrate that nasal closure significantly improves epistaxis severity without having a significant effect on sleep or nasal obstruction as they relate to QOL. These findings suggest that nasal closure should be considered for HHT patients with chronic severe epistaxis.

Key Words: Hereditary hemorrhagic telangiectasia, epistaxis, quality of life.

Level of Evidence: 4.

INTRODUCTION

Hereditary hemorrhagic telangiectasia (HHT) is a hereditary blood vessel disorder with multisystem manifestations, including cerebral, pulmonary and hepatic arteriovenous malformations.^{1,2} However, the most common symptom is epistaxis related to intranasal telangiectasias. Oral and cutaneous telangiectasias are also characteristic, but rarely bleed. It is thought that the nose bleeds preferentially due to the drying effects of the air passing over the telangiectasias, causing them to rupture and bleed, sometimes profoundly. The frequency of episodes of epistaxis, as well as their duration and quantity of blood lost, vary widely. Chronic and severe epistaxis can be exceedingly detrimental to the patient's quality of life (QOL).^{3,4}

HHT patients with severe epistaxis often seek medical attention to help control this debilitating and unpredictable issue. A range of interventions can be employed including nasal packing, hormone therapies, monoclonal antibodies, local sclerotherapy, diathermy, and various coagulation and cautery techniques that often demonstrate at least short-term effectiveness.⁵⁻⁸ Coagulation and cautery techniques have consistently shown benefit, but most patients experience a recurrence of their symptoms within 1–2 years.^{5,7,8} Systemic pharmacological therapies, such as low-dose bevacizumab, have been explored in recent years with evidence of benefit for patients seeking a non-invasive treatment of severe epistaxis prior to considering surgical intervention.^{9,10}

Surgical closure (modified Young's procedure) of the nares has demonstrated effectiveness in preventing epistaxis in patients who do not respond to other treatments or fail to achieve long-term resolution.^{7,11,12} The procedure begins at the mucocutaneous junction within the nasal vestibule by creating a circumferential incision in the skin. Three-flap and two-flap techniques have been described with equal efficacy.¹³ Permanently closing the nares is thought to be effective because it protects the fragile intranasal telangiectasias from the drying effects of air flow. The utility of the Young's procedure for treating epistaxis can be evaluated using the Epistaxis Severity Score (ESS).¹⁴ Although complete or near-complete cessation of nasal bleeding has been shown to improve QOL in patients,¹⁵ the Young's procedure has notable drawbacks,

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Editor's Note: This Manuscript was accepted for publication 12 March 2018.

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DOI: 10.1002/liv.2.157

TABLE I.

Demographics of Nasal Closure and No Nasal Closure Groups.
[Color figure can be viewed at wileyonlinelibrary.com]

Demographic	Nasal Closure (treatment)	No Nasal Closure (control)	<i>P</i> value
Total patients	5	8	
Male	1	3	
Female	4	5	
Age (mean \pm SD)	64.6 \pm 16.44	67.4 \pm 15.8	.767

including loss of smell, dry mouth, and nasal obstruction, which can potentially interfere with sleep.¹⁶ Currently, there is no evidence to suggest that these patients experience increased nasal obstruction or diminished sleep quality following nasal closure.

Nasal closure has been shown to improve overall QOL,⁷ however, specific aspects of QOL that may be affected by the procedure remain unclear. The objective of the present study is to examine the effects of nasal closure on specific QOL parameters, including sleep.

MATERIALS AND METHODS

This is an Institutional Review Board (IRB)-approved study (86476). Patients with severe chronic epistaxis who have been treated at the University of Utah's HHT center were recruited for this study. Cases are those patients who had nasal closure. Controls were HHT patients who did not have nasal closure but had severe epistaxis, defined as an ESS greater than or equal to 7. A total of 8 controls and 5 cases were included in this study. Cases and controls were matched with regards to severity of epistaxis based primarily on their ESS. The ESS is a standardized, validated tool for the assessment of epistaxis severity. Patients are assigned a score from 1 to 10 based on their answers to six questions; mild (0–4), moderate (4–7), or severe epistaxis (7–10).¹⁴ ESS scores for controls prior to administration of the surveys was found through retrospective chart review. This procedure was offered to patients who had a recurrence of symptoms after cauterization techniques. All patients in case and control groups sought repeated medical attention and had undergone at least one cauterization procedure prior to obtaining baseline ESS.

After consent, each patient completed the ESS, Pittsburgh Sleep Quality Index (PSQI), and the Nasal Obstruction Symptom Evaluation (NOSE) questionnaires, all of which are standardized and validated tools used to evaluate varying aspects of QOL. For the PSQI, a score of 0 to 20 is determined. The lower the PSQI score, the better the sleep quality.¹⁷ The NOSE assigns a patient a score of 0 to 100, with higher scores correlating with greater nasal obstruction symptoms.¹⁸ These surveys were sent to each patient through the mail, providing them with instructions to return their completed survey. Per protocol, those who did not fill out the survey after two weeks were contacted by phone to complete the survey verbally. The results of the two groups were then compared. Statistical analysis was performed using two-tailed independent samples and t-test for equality of means using the SPSS software.

RESULTS

In total, 15 surveys were sent to cases and 17 were sent to controls; of which 9 were returned by mail and 4 were completed over the phone. Of the 13 completed surveys, 5 were from nasal closure patients (cases) and 8 were

from severe epistaxis patients who had not had nasal closure (controls) (Table I). Individuals were excluded if contact could not be made to conduct the survey. No patients voluntarily withdrew from the study. The nasal closure group were an average of 5.82 (\pm SD 3.65) years postnasal closure. The average age of the cases (64.6 \pm SD 16.44) and controls (67.4 \pm SD 15.80) was comparable (Table I).

Prior to this study, the controls had an average ESS of 7.54 (SD \pm 0.35). Two of the nasal closure cases had a preoperative ESS available which averaged 8.38 (SD \pm 0.64). There were no incomplete closures and no reported complications.

Table II displays the means and standard deviations of the ESS, PSQI, and NOSE scores for both groups at the completion of this study. Results show a statistically significant improvement of ESS in the nasal closure patients ($P = .027$). There was no significant difference between the PSQI scores of the two groups ($P = .411$). There was also not a statistically significant difference between the groups with regards to scores based on the NOSE questionnaire ($P = .467$).

DISCUSSION

The ESS tool measures epistaxis severity, and previous studies have demonstrated a strong, reliable correlation between ESS and Health-related Quality of Life (HR-QOL).³ The questionnaire contains six questions and focuses primarily on the three months prior to the administration of the survey. Specifically, the ESS identifies the frequency, duration, and intensity of bleeding episodes as well as whether or not the patient is currently anemic, has sought medical attention or required red blood cell transfusion as a direct result of nasal blood loss. A patient with severe epistaxis typically has a diminished overall QOL. Comparing the ESS for nasal closure patients to that of patients with epistaxis of similar severity, but without nasal closure, provides insight into their QOL. The nasal closure patients in this study had a significantly lower ESS (1.10) compared to the non-nasal closure group (3.99), indicating better epistaxis control. It is intuitive that the nasal closure group experienced better control of epistaxis than their non-nasal closure counterparts. However, even the non-nasal closure group experienced a decrease in ESS from their reported score prior to the study, from 7.54 to 3.99. The reason for the improvement of ESS in these patients is unclear, but could possibly result from recent interventions the patients underwent that were performed outside of our facility. For example, one patient reported on the phone that they had recently started a medication that had been helping their symptoms. This adds further evidence that patients with high ESS scores will continue to seek treatment to improve their symptoms. Even with the decrease of ESS in the control group, results demonstrate that epistaxis severity was lowest in the cases that had undergone nasal closure. Interestingly, among the nasal closure patients, an average of 5.82 years lapsed prior to ESS administration for this study yet epistaxis remained mild with an average of 1.10. This supports prior evidence that nasal closure provides long-term benefit for epistaxis, although previous

TABLE II.

Average Survey Scores for Nasal Closure Group and Group Without Nasal Closure with Corresponding *P* Values. [Color figure can be viewed at [wileyonlinelibrary.com](#)]

Survey	Nasal Closure (treatment)	No Nasal Closure (control)	<i>P</i> value between groups
ESS			
Mean	1.10	3.985	.027*
SD	1.33	2.275314735	
<i>n</i>	5	8	
PSQI			
Mean	11.60	14.875	.411
SD	6.11	7.059694449	
<i>n</i>	5	8	
NOSE			
Mean	53	43.125	.467
SD	21.10	24.04423008	
<i>n</i>	5	8	

ESS = Epistaxis Severity Score; PSQI = Pittsburgh Sleep Quality Index; NOSE = Nasal Obstruction Symptom Evaluation Questionnaire.

research has not demonstrated precise pre- and post-op ESS.^{7,11,19,20}

Decreasing epistaxis severity will significantly improve health-related QOL, as demonstrated by several former studies.^{3,21,22} As an example, Merlo et al. studied 604 HHT patients and found that severe epistaxis, measured using ESS, was associated with significantly lower scores in both mental and physical domains in Health-related QOL. In the present study, the decreased epistaxis severity following nasal closure implies improved QOL. However, epistaxis severity scores alone are insufficient at determining if nasal closure is worth the drawbacks associated with not being able to breathe through one's nose after complete closure of the nose. Previous research has identified patient-reported disadvantages of nasal closure, relating to respiratory, olfactory, and phonation difficulties.¹⁶ No research has yet elucidated the impact that nasal closure has on sleep and nasal obstruction using objective and reproducible metrics. Evaluating the PSQI and the NOSE results of cases versus controls further examine the impact nasal closure has on specific aspects of one's life.

In this study, the PSQI results indicate no difference between the groups in terms of sleep quality. However, a good PSQI score is considered to be 5.0 or less, an ideal score that neither group reached. This indicates that severe epistaxis may have a negative impact on sleep regardless of whether the nose has been closed. One nasal closure patient stated, "Before the Young's procedure I would wake up with horrible nose bleeds...now I just get dry mouth which wakes me up. I just keep a glass of water next to my bed and I go right back to sleep." Comments such as this from our patients, combined with the PSQI data indicate that nasal closure did not significantly alter sleep quality in these patients with a history of severe epistaxis.

The results of the NOSE questionnaire were also striking. Both cases and controls reported similar moderate-severe nasal obstruction. It is interesting that the nasal closure patients did not report more severe nasal obstruction, given that they no longer have airflow through their nose at all. It is noteworthy that the non-nasal closure patients had such significant nasal obstruction, presumably from the chronic nasal crusting and swelling that is present in these patients. These results indicate that even completely closing off the nares does not significantly alter nasal obstruction in patients with severe epistaxis.

The main limitation of the study is the fact that there was no QOL questionnaire administered to patients at the time they were counseled about the option for nasal closure or preoperatively for those who underwent nasal closure. In addition, although QOL was measured as it relates to epistaxis severity, nasal obstruction and sleep quality, an overall QOL assessment was not administered as part of this study. However, previous studies provide evidence that epistaxis severity per se is significantly correlated to overall HR-QOL. Lastly, since this study was a retrospective review, some information was not found including, the exact number of interventions performed for epistaxis and the exact age of onset of epistaxis. However, both cases and controls had similar epistaxis severity and underwent at least one laser surgery prior to this study, making their comparison appropriate and informative.

CONCLUSION

Nasal closure significantly decreases epistaxis severity with no significant effect on sleep quality or nasal obstruction as compared to patients without nasal closure. Nasal closure should be considered for HHT patients with chronic severe epistaxis that have failed more conservative measures.

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