Outcome of Septal Dermoplasty in Patients With Hereditary Hemorrhagic Telangiectasia

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Objectives/Hypothesis: Septal dermoplasty has been recommended as the treatment of choice for life-threatening epistaxis in patients with hereditary hemorrhagic telangiectasia. The purpose of the study was to evaluate the effectiveness and outcomes of septal dermoplasty for management of transfusion-dependent epistaxis. Study Design: Retrospective study. Methods: Between 1994 and 2004, septal dermoplasty was performed on 67 consecutive patients with severe epistaxis attributable to hereditary hemorrhagic telangiectasia. The numbers of units of blood received 1 year before and 1 year after septal dermoplasty were obtained. A subjective appraisal of the results of the surgery as well as second procedures after septal dermoplasty was determined. Patients were screened for pulmonary and cerebral arteriovenous malformations, gastrointestinal tract bleeding, and symptomatic liver disease. Results: Data were obtained in 66 of 67 (98%) patients with a mean age of 61.5 years (mean follow-up, 3.9 y). Accurate transfusion requirements 1 year before and 1 year after septal dermoplasty were available in 32 of 66 (48%) patients. In these 32 patients, the mean units of blood received decreased from 21 units (range, 2–100 units) 1 year before septal dermoplasty to 1 unit (range, 0–10 units) in the year after septal dermoplasty (P < .001). Improved quality of life was claimed in 57 patients. Second therapies, ranging from cautery to repeat partial septal dermoplasty, were required in 15 patients during follow-up. Among the 67 patients, 31 (46%) had pulmonary arteriovenous malformation, 14 (21%) had gastrointestinal tract bleeding, 7 (10%) had symptomatic liver disease, and 5 (7%) had cerebral arteriovenous malformation. During the follow-up, 14 patients died of other complications of hereditary hemorrhagic telangiectasia (11 patients) and unrelated causes (3 patients). Conclusion: Septal dermoplasty remains an effective way of reducing transfusion requirements in patients with hereditary hemorrhagic telangiectasia and subjectively improves their quality of life. The otolaryngologist caring for patients with hereditary hemorrhagic telangiectasia should be familiar with other organ involvement by hereditary hemorrhagic telangiectasia to prevent complications during surgery. Key Words: Septal dermoplasty, epistaxis, hereditary hemorrhagic telangiectasia.

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INTRODUCTION

Hereditary hemorrhagic telangiectasia (HHT) is an uncommon systemic autosomal dominant disorder of angiogenesis characterized by the presence of vascular telangiectases in mucocutaneous tissues, visceral organs, and the central nervous system. Epistaxis, caused by spontaneous bleeding from telangiectases of the nasal mucosa, is the most common manifestation of HHT. It may be so severe as to require multiple transfusions and intravenous iron supplementation. Bleeding becomes more severe in later decades in approximately two-thirds of affected persons. Although nasal manifestations of HHT are well known to otolaryngologists, many of the other organ manifestations of this disorder are less well known and may pose issues for the physician treating the patient with HHT.

Mild and moderate epistaxis is treated by hormonal therapy with estrogens, chemical cautery and electrocautery, and laser coagulation. Life-threatening epistaxis, which is associated with HHT, has been treated with varying success using embolization, surgical ligation, Young’s surgical closure of the nostrils by a skin flap, and microvascular free flaps.

William Saunders pioneered a skin grafting technique for severe epistaxis in 1958; since then, this technique has undergone modification. The principle of this procedure is to replace the fragile respiratory mucosa of the nose with strong keratinized split-thickness skin.
grafts from the thigh that resist trauma and thereby pre-
vent bleeding. Since the early 1990s, the senior otolaryngologist (D.R.) has used the Saunders method of septal dermoplasty (SD), with his own modifications, to control transfusion-
dependent epistaxis. The purposes of the present study were to describe indications and new variations of SD, to characterize the phenotype of patients requiring this ther-
rapy, and to report the long-term outcome of SD.

PATIENTS AND METHODS

The Yale Arteriovenous Malformation Center is an interdis-
ciplinary center of physicians caring for patients with HHT. From a population of approximately 250 patients seen yearly, 67 con-
secutive patients with severe, debilitating epistaxis were referred to the otolaryngologist and underwent SD. Human Investigations Committee approval was obtained for a retrospective study that included contacting all patients, as well as their primary care physicians. Detailed information about total transfusions (units of blood) 1 year before and 1 year after SD was requested. A subjective appraisal of the results of the surgery was asked of the patients and their families. Second therapies for control of nose-
bleeds after SD were also tabulated. Patients in the present study were seen by the genetic counselor and senior physician in the center and screened for pulmonary and cerebral arteriovenous malformations and symptomatic liver disease.

Surgical Technique

Surgical technique is shown in Figures 1 and 2. With the patient under general endotracheal anesthesia, a 3 × 7-inch split-thickness graft is taken from the lateral aspect of the right-
side thigh, using a Zimmer air dermatome on a setting of 141000of an inch. Xeroform and Telfa with Kerlex-wrapped dressing are applied on completion of harvesting of the skin and the graft, which is preserved in a saline-soaked sponge.

Bilateral inferior turbinectomies are performed in the fol-
lowing manner. Neo-Synephrine is placed into the nose for ade-
quate vasoconstriction; then the inferior turbinates are infiltrated with 1% lidocaine and 1:100,000 epinephrine. The turbinate is medialized and removed at its base using an anterior straight scissors under endoscopic view, cutting anteroposteriorly, as much as possible toward the lateral nasal wall. The free edge of the remnant of the turbinate is then cut down using microderider. Meticulous hemostasis is achieved using Bovie electrocautery.

The septal dermoplasty is performed. The nasal vestibule is injected with 1% lidocaine and 1:100,000 epinephrine. After 7 minutes a narrow circumferential strip of vestibular skin is excised from the septum and the floor and from the lateral wall of both nostrils at the mucocutaneous junction. As much mucosa as possible is removed using a Faulkner curette, taking care to preserve the perichondrium of the septum that must nourish the new grafts and the septal cartilage.

Bleeding is controlled by the topical application of cotton soaked in epinephrine in one nostril and continuing the operation in the opposite side. Once the bleeding has slowed, the curetting is continued until one is assured of adequate removal of mucosa.

The skin graft is sutured in a U shape. A stitch is placed at the inferior aspect of the rim incision with interrupted 3-0 chromic suture, and the skin graft is sutured circumferentially around the rim incision of the nasal vestibule. The skin graft is then inserted into the nose with a No. 2 bayonet forceps, and a long nasal speculum is used to hold it in place for packing.

Packing and Removal

An important aspect is careful nose packing, to avoid the displacement of the skin graft while healing takes place. Multiple 0.5 × 3-cm Neuropatts treated with mupiurocin calcium oint-
ment (Bactroban, Galaxo Smith Kline, Philadelphia, PA) are used for this task. The first pack is placed along the floor of the nose to anchor the graft inferiorly; then a second piece is placed superi-
orly, continuing in this alternate fashion until the nose is packed firmly. Packing is removed only after 5 days, when the grafts are well attached.

There is often “temporary” nasal obstruction after the packs are removed, because of excess skin at the posterior part of the nose. This acts as an avascular small flap that eventually disinteg-
rates. When this occurs, the nasal airways improve.

Follow-Up Care

Nothing more is performed for 10 to 14 days after surgery so that the skin graft can become firmly attached to the nasal walls. During follow-up, cautious suctioning of crusts is performed to facilitate breathing and success of graft take. Skin is not meant to be in the nose, and crusting from desquamation frequently devel-
ops, requiring that the patient provide vigorous nasal hygiene. Crusting may lead to nasal obstruction and odor; therefore, the nose must be cleaned once or twice daily. Cleaning is facilitated using cotton-tipped applicators and moisturizing substances such as mineral oil, Borofax ointment, half hydrogen peroxide and half water, or soap and water. Saline irrigations can also be used. This cleaning cannot injure the skin, and it cannot be scraped off.

RESULTS

Between 1994 and 2004, SD was performed in 67 patients (30 men and 37 women) (Table I). Data were obtained in 66 of 67 patients (98%). The mean age at the time of SD was 61.5 years (age range, 35–80 y). Transfusion requirements either before or after SD were available in 50 of 67 patients (75%). Complete data before and after SD were available in 32 of 66 patients (48%) of this group. Table II In 66 of 67 patients (98%), the mean follow-up period was 3.9 years (range, 2 mo–9.8 y). Quality-of-life information was available for all living patients and from their families for patients who had died.

TABLE I.
Clinical Phenotypes in 67 Patients Undergoing Septal Dermoplasty.

<table>
<thead>
<tr>
<th>Variable</th>
<th>n</th>
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<tbody>
<tr>
<td>Male</td>
<td>30</td>
</tr>
<tr>
<td>Female</td>
<td>37</td>
</tr>
<tr>
<td>Age (y)</td>
<td></td>
</tr>
<tr>
<td>Mean</td>
<td>61</td>
</tr>
<tr>
<td>Range</td>
<td>35–80</td>
</tr>
<tr>
<td>Other organ: n (%)</td>
<td></td>
</tr>
<tr>
<td>Pulmonary arteriovenous malformation</td>
<td>31 (46%)</td>
</tr>
<tr>
<td>Cerebral arteriovenous malformation</td>
<td>5 (7%)</td>
</tr>
<tr>
<td>Gastrointestinal tract bleeding</td>
<td>14 (21%)</td>
</tr>
<tr>
<td>Symptomatic liver disease</td>
<td>7 (10%)</td>
</tr>
</tbody>
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Transfusion Requirements and Quality-of-Life Outcomes

Thirty-eight of the 50 patients required blood transfusions before SD, and 11 required them after SD (Table II). In 32 patients we were able to determine the exact number of units of blood received before and after SD. In these 32 patients, the mean units of blood received decreased from 21 units in the year before SD (range, 2–100 units) to 1 unit (range, 0–10 units) during the year after SD. The Wilcoxon signed rank test demonstrated a significant decrease in the number of transfusions in these patients (P < .001).

We were able to question 57 patients or families of patients about quality of life: 53 reported that their quality of life had improved after SD, and 4 reported no improvement. Twenty-four of the 57 patients (42%) reported no epistaxis (mean follow-up, 2 y [range, 2 mo–8 y]); 29 patients (51%) reported decreased frequency and duration of epistaxis (mean follow-up, 2 y, 9 mo [range, 1 mo–8 y]), and 4 patients (7%) had not improved since surgery (Table II).

Clinical Phenotype

Other organ involvement. Pulmonary arteriovenous malformations were present in 31 of 66 patients (47%), and cerebral arteriovenous malformations were found in 5 of 66 patients (7%). Symptomatic liver disease was present in 7 of 66 patients (10%) (Table I). In addition, 14 patients (21%) had gastrointestinal tract bleeding before or after SD. Two patients had a prosthetic heart valve and tolerated coumadin well after SD.

Deaths during follow-up. Fourteen of 66 patients (21%) died during the follow-up after SD. Five of these patients had gastrointestinal tract bleeding but died of heart disease.

In 3 of 14 patients, death was attributed to pulmonary hypertension associated with HHT. Liver disease and intractable congestive heart failure were the causes of death in 2 of 14 patients, respectively, and bleeding from esophageal varices (1 of 14 patients) accounted for the additional death in this group. Three of 14 patients died of colon cancer, pancreatitis and pneumonia, and lung cancer, respectively.
Second therapies after septal dermoplasty.

Multiple additional therapies have been performed in 15 patients (laser therapy in 10 patients, a second SD in 6, embolization in 2, and maxillary artery ligation in 1 patient). Many of these therapies were performed at other institutions because some of the patients did not return to our center for follow-up.

DISCUSSION

Technique History and Modifications

Since the early 1960s, surgical techniques for managing debilitating nosebleeds in HHT patients have evolved, but epistaxis of this magnitude is still problematic. Many authors have proposed alternative procedures including laser cauterization, embolization, maxillary artery ligation, Young’s procedure, and microvascular free flaps. Although each method has advocates in these patients, no method has provided a complete cessation of epistaxis, and reporting of long-term outcomes in these patients has been limited.

Currently, the most credible technique is septal dermoplasty, first described by Saunders9 in 1958. Replacement of the fragile nasal mucosa with a skin-thickness graft from the thigh renders the nose more resistant to trauma without altering the nasal respiratory function. However, good result of SD depends not only on quantity of mucosa covered by the skin graft but also on the follow-up care. Skin graft desquamation produces odor and crusting; therefore, daily cleaning is necessary to avoid nasal obstruction and nose bleedings.

Presence of telangiectases in nasal mucosa not covered by the graft, as well as graft contracture, can cause postoperative epistaxis, which is treatable by laser technique. Septal dermoplasty may be repeated in patients with recurrent severe epistaxis. Modification of the technique has been performed recently to facilitate the skin graft positioning, cleaning, and maintenance of airway by inferior bilateral turbinectomies.11 Septectomy has been useful for one patient with profuse nosebleeds and septal perforation.

Outcomes

Our study is a large retrospective review of patients with severe epistaxis attributable to HHT that was treated by SD. To date, the data on SD reported in the literature have been based on a small number of patients and the duration of follow-up has been short. Our outcomes are longer than previously reported and are based on transfusion requirements before and after SD, as well as subjective questioning of the patient or family. Cessation or marked reduction of epistaxis occurs in 90% of patients with HHT during the first 2 years after SD. Four patients did not benefit from SD. Our clinical experience has demonstrated that, beyond the obvious health damage, epistaxis of this magnitude seriously affects the quality of life of the patient with HHT.

Limitations of our study include the retrospective design and the recall bias associated with the quality-of-life assessment. Because some of our patients developed repeat bleeding attributable to new telangiectases or partial graft take and have received adjunctive therapy at 1 year, we currently recommend that all patients be seen 1 year after surgery. Management of recurrent epistaxis by laser or second partial SD is able to extend the time of improvement and emphasizes the importance of follow-up by the otolaryngologist performing the SD.

Importance of Phenotype

Many patients in the present series have had other serious organ involvement, which ultimately contributes to their disability and mortality. Symptoms such as dyspnea, fatigue, and congestive heart failure can be manifestations of pulmonary arteriovenous malformations, hepatic arteriovenous malformations, gastrointestinal tract bleeding, and iron deficiency anemia or some combination of all of these.15 In addition, in patients with pulmonary arteriovenous malformations, the lung no longer has a capillary filter, and small blood clots, bacteria, and, occasionally, air can pass directly through the pulmonary arteriovenous malformation into the systemic circulation, causing ischemic events in various organs.1 Intracerebral hemorrhages may also occur as a result of cerebral or spinal arteriovenous malformations.1 In our study, three patients had primary pulmonary hypertension, which is a syndrome newly recognized in HHT.16

Patients with a history of pulmonary arteriovenous malformations are prone to air or clot emboli during intravenous infusions. Antibiotics should be given prophy-lactically for procedures. In most instances, the otolaryngologist is the first physician to see a patient with HHT because epistaxis is the most common symptom.5 First-visit patients should be screened for other organ involvement.

Although imperfect, the modified Saunders dermoplasty, coupled with follow-up care at 1 year and in subsequent years, is the best therapy for this subset of patients. In the near future, many new pharmacological agents, with anti-angiogenesis properties will be developed and, perhaps, may offer a possible nonsurgical solution; However, until that time, SD that is specific for HHT should be the mainstay for transfusion-dependent epistaxis.

CONCLUSION

The modified SD for transfusion-dependent epistaxis in patients with HHT provides excellent palliation and improves quality of life. The need for transfusions was significantly reduced in the subset of 32 patients with accurate blood transfusion records 1 year before and 1 year after SD. All patients with HHT who are undergoing SD should be seen 1 year after SD because in as many as 25% of patients, laser or other secondary procedures will be necessary. The otolaryngologist treating patients with HHT should be aware of other organ manifestations, which may influence management of the patient during and after SD.

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BIBLIOGRAPHY