

Introduction

Hereditary hemorrhagic telangiectasia (HHT) is an autosomal dominant, multisystem angiodysplastic disorder¹

Recurrent epistaxis occurs in nearly 100% of cases by age 40

Epistaxis related to HHT can be life-threatening and decreases QoL

Numerous topical, ablative, systemic, and surgical treatments have been utilized with varying degrees of success²

Septodermoplasty decreases the need for blood transfusions and improves quality of life (QoL)

Current medical and surgical options are limited, particularly in the advanced stages of the disease

Nasal resurfacing using radial forearm free flap through bilateral lateral rhinotomy incisions has been described³

Case

73-year-old man with HHT

History of HTN, BPH, Afib s/p ablation, left RCC s/p nephrectomy in 2017, lung AVMs embolized in 2017

Epistaxis started around age 25, up to 5x per day, choking on nightly bleeding, IV iron PRN

Declined nasal closure

Resurfacing endoscopic free flap (REFF)

1. Total septectomy
2. Bilateral endoscopic medial maxillectomies
3. Inferior and middle turbinate resections
4. Left Caldwell-Luc antrostomy
5. Left neck exploration for donor vessels
6. Right radial forearm fasciocutaneous free flap used to resurface nasal cavity
7. Nasal cavity packed with foley catheter balloon and NasoPore

Results/Discussion

Minimal epistaxis with significantly improved QoL

Epistaxis Severity Score improved from 8.9 to 3.5

Underwent revision surgery with debulking of flap and DCR for epiphora

Future work will revise inset technique using 3D printed skull to map placement

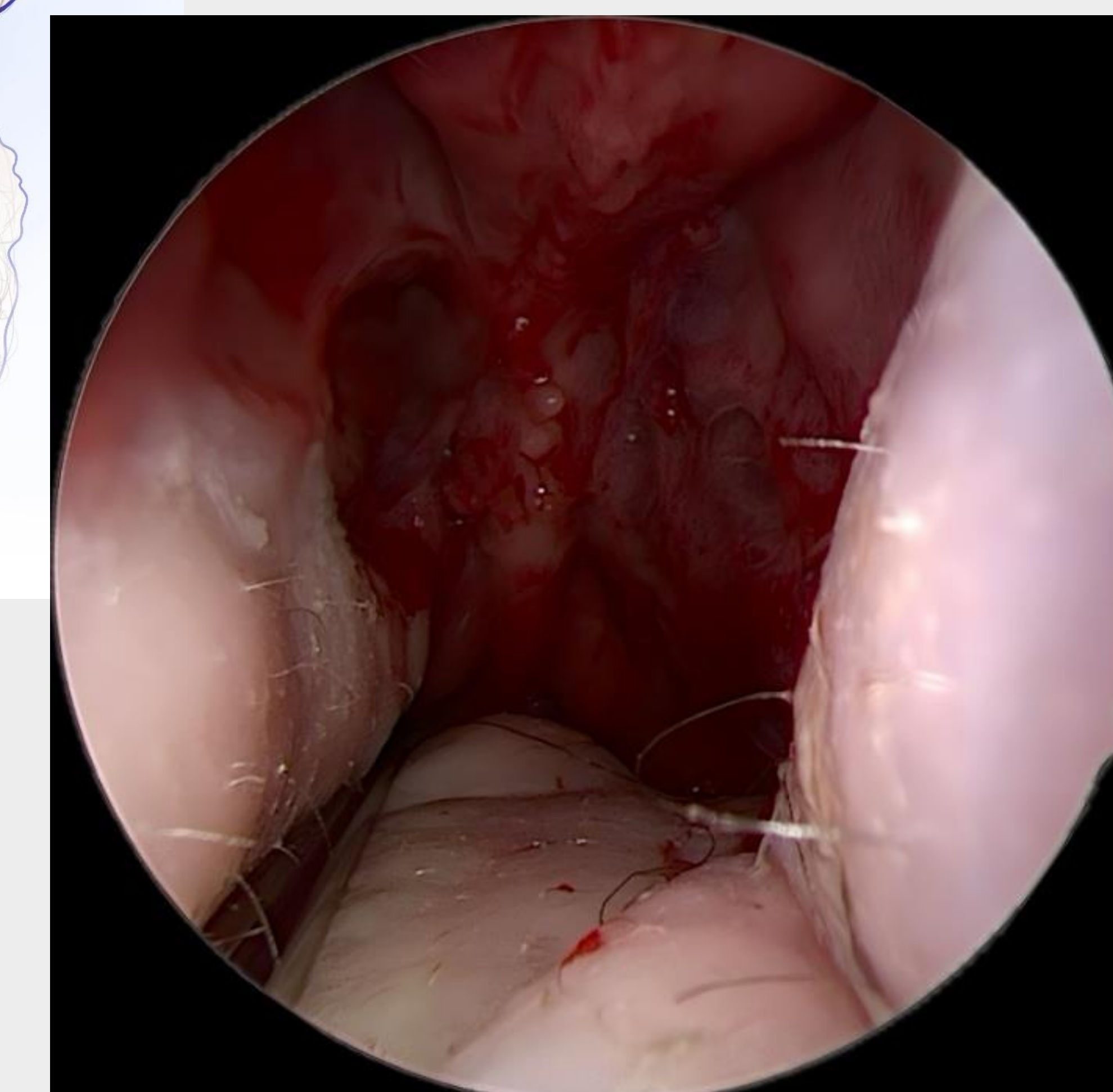
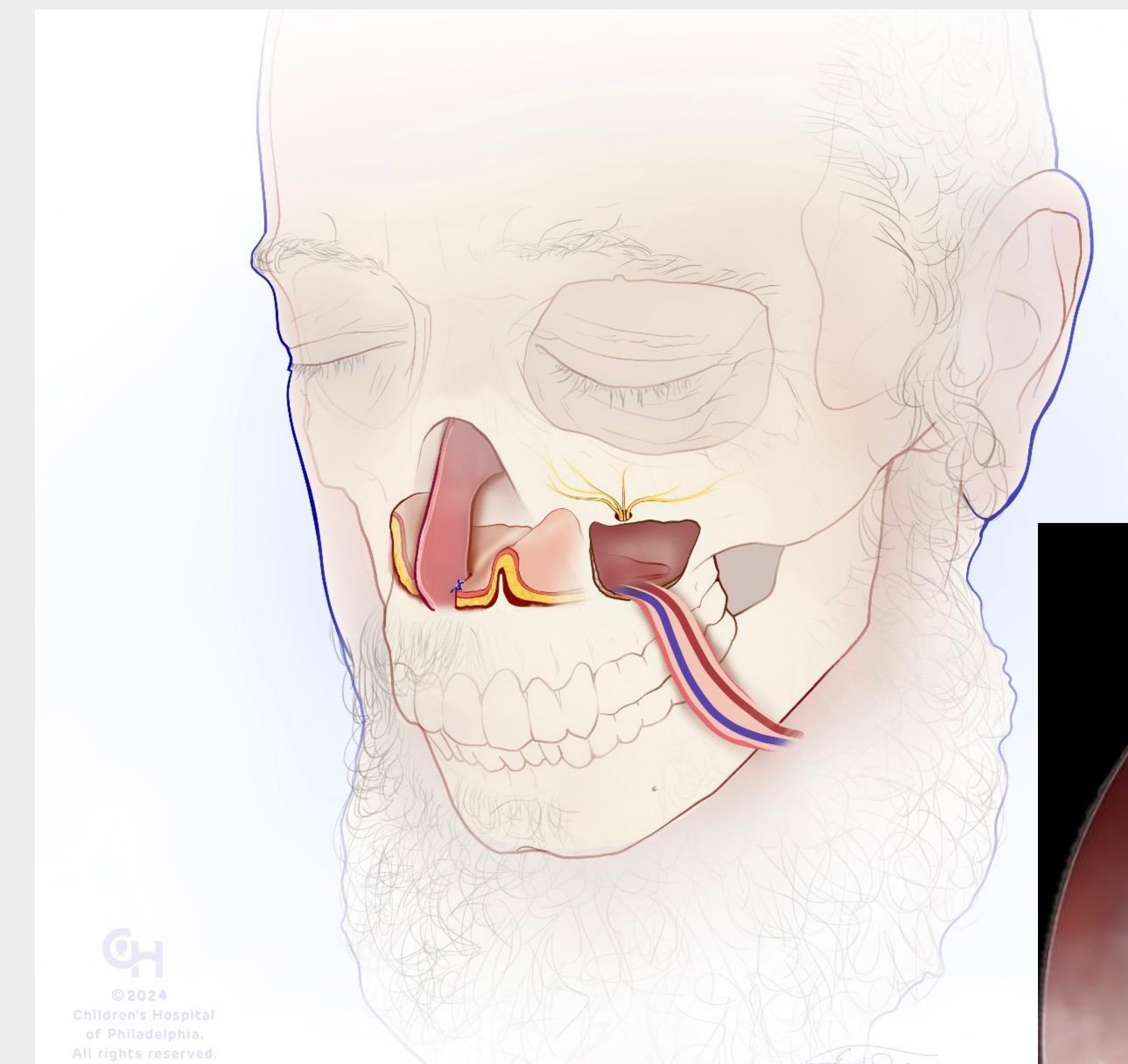
Treatment History

Medical:

- Topical moisturization
- Doxycycline trial, had initial improvement but benefit waned
- Avastin (bevacizumab) trial, did not have improvement
- PO TXA 1,300 mg TID with improvement
- Topical TXA PRN

Surgical:

- Numerous cauterizations
- Sclerosing agents
- Septodermoplasty



References

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