

Double Balloon Enteroscopy in HHT Patients: A Large Case Series

Goldman, SA, BA¹, Conrad, M, MD, MPH², Langston, D, MSN², El-Nachef, N, MD³

¹Department of Medicine, University of California, San Francisco, San Francisco, California, USA; ²Department of Radiology, University of California, San Francisco, San Francisco, California, USA; ³Department of Gastroenterology, University of California, San Francisco, San Francisco, California, USA

INTRODUCTION

- Guidelines recommend esophagogastroduodenoscopy (EGD) and colonoscopy to diagnose and treat suspected HHT-related GI bleeding in the esophagus, stomach, first portion of the duodenum, and large bowel. Capsule endoscopy can diagnose lesions throughout the small bowel, but it cannot deliver treatment¹.
- DBE facilitates endoscopic access into the small intestine². It has been shown to have high diagnostic yield and therapeutic benefit in mid-GI bleeds³, but it has not been extensively studied in patients with HHT.

OBJECTIVES

- Evaluate double balloon enteroscopy (DBE) findings and outcomes in a large series of patients with HHT
- Assess safety of repeat DBE-directed therapy in patients with HHT

METHODS

- We reviewed medical records of all patients referred to UCSF HHT Center of Excellence who underwent DBE.
- DBE findings and complications were recorded, as were hemoglobin and blood transfusion requirements in the year prior and following DBE.
- Univariate analysis was used to analyze outcome predictors such as age, sex, smoking, and lesion burden.

RESULTS: DBE findings

- The study included 14 patients and 28 procedures. Median age was 60, range 43-78. 6 patients were female; 10 were on systemic therapy in the year preceding or following DBE.
- Arteriovenous malformations (AVMs) were identified in the stomach, duodenum, jejunum, ileum, and colon. Number of AVMs ranged from 9 to >100; 21 scopes revealed >20 AVMs.
- AVMs were treated with argon plasma coagulation (APC) in all but 4 procedures; gold cautery was used for 5 procedures; clips were placed in 2.
- No procedure was associated with any adverse event.

Sex	Genotype	Age	DBE #	AVMs				Total Lesions	Systemic Therapy	
				Stomach	Duodenum	Jejunum	Ileum		Pre-DBE	Post-DBE
M	ACVRL1	46	1	NR	NR	NR	-	12	0	0
F	ENG1	57	1	1	many	many	-	TNTC	1	1
		58	2	3	many	many	-	50	1	2
		59	3	2	many	many	-	TNTC	2	1, 2
M	ACVRL1	66	1	2	many	2	-	TNTC	1	0
		68	2	0	0	9	-	9	0	0
F	negative	68	1	0	0	many	-	TNTC	0	1
F	missing	55	1	3	many	many	-	50	1	2
M	ACVRL1	64	1	NR	NR	NR	-	36	0	0
		66	2	1	many	many	-	TNTC	0	0
		68	3	-	-	-	16	18	0	2
		68	4	1	few	many	-	TNTC	0	2
		68	5	6	many	many	-	TNTC	2	1
		70	6	0	0	8	-	8	1	1, 2, 3
		70	7	0	many	many	-	TNTC	1, 2	1, 3
F	ENG1	78	1	4	5	many	-	TNTC	1, 4, 5	1, 4
F	ENG1	58	1	0	0	10	-	9	1	0
M	missing	43	1	0	many	many	-	TNTC	0	0
		43	2	0	many	many	-	50	0	0
M	missing	61	1	0	many	many	-	100	0	0
M	ENG1	49	1	0	0	many	-	50	0	0
F	ENG1	44	1	NR	many	many	-	30	0	0
		45	2	0	many	many	-	60	1	1
M	missing	62	1	some	many	many	-	50	1	1
M	ACVRL1	55	1	many	many	many	-	36	0	1
		60	2	2	5	many	-	32	2	2

DBE findings extracted from procedure reports. Systemic therapy pre- and post-DBE = HHT pharmacotherapy in the year before and after DBE, respectively; M = male; F = female; NR = not reported; TNTC = too numerous to count; Systemic therapies: 0 = no systemic pharmacotherapy 1 = tranexamic acid, 2 = bevacizumab, 3 = pazopanib, 4 = estradiol, 5 = octreotide.



Column A: AVMs. Column B: AVMs following APC.

RESULTS: DBE outcomes

- In patients with <20 AVMs, hemoglobin dropped by a mean of 0.97 units (95%CI 0.26-1.68) 2 weeks following the procedure but returned to pre-procedure levels by 8 weeks. Other groups had no change in hemoglobin.
- Decreased transfusion requirements in the year following DBE were observed in the 13 procedures performed at UCSF (average -5.2 units/year 95%CI 1.4-8.9) and in those with 20-100 AVMs (-4.6 units/year 95%CI 0.7-8.5).

CONCLUSIONS

- HHT patients may have a heavy burden of AVMs in the small bowel resulting in blood losses requiring transfusions.
- In one of the largest series of HHT patients undergoing DBE, we found that DBE is safe, even when repeated.
- DBE may be associated with decreased blood transfusion requirements when performed in an HHT center, possibly due to improved interdisciplinary care.

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