Embolisation in the management of epistaxis in patients with hereditary haemorrhagic telangiectasia: a systematic review

Nikhil Shah¹, Lawrence Nip¹, Maha Khan² & Katherine Conroy²

1. University College London
2. North Western Deanery

Introduction

Hereditary haemorrhagic telangiectasia (HHT), also referred to as Osler-Weber-Rendu disease, is an inherited disorder of the vasculature with consequent involvement and sequelae in a number of different organ systems. It is characterized by the appearance of telangiectasia, most prominently in the skin and mucosa of the respiratory and gastrointestinal tracts, and spontaneous bleeding from multiple sites. Epistaxis is a prominent feature of the condition, occurring multiple times per day in the most affected with potential life threatening consequences.

There remains much debate regarding the best treatment for these patients. As of now, no treatment offers a cure and there is lack of consensus regarding the most effective way to manage epistaxis in a patient with HHT.

Aims & Objectives

The aim of this systematic review was to evaluate the efficacy of embolisation therapy in the management of epistaxis in patients with HHT. Five papers were selected for critical appraisal, with the key findings shown in the table below.

Search Strategy

A standardised search methodology was performed using MEDLINE, EMBASE, Cumulative Index to Nursing and Allied Health Literature (CINAHL) , Web of Science and the Cochrane Library databases. Following removal of duplicates, 466 records were initially identified and after a two stage screening process, 5 articles were accepted for this review (Fig 1.).

Fig.1 Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) diagram for the review, mapping out the different phases of the record selection process

Author & Journal	Patient group	Outcomes	Key results	Comment
P Andersen et al., 2005. Acta Oto-laryngologica Case-control study	22 patients (16 male, 6 female) treated with embolization divided into 2 groups: Patients with HHT, n=9 Patients without HHT, n=13 Patients followed up by questionnaire and telephone interview.	Aims: Determine the efficacy of embolization therapy in HHT patients and patients with no pathology of the nasal cavity and identify the potential complications of embolization. End points: - Frequency of epistaxis - Duration of epistaxis - Number of major complications	1. Epistaxis recurred following 13/15 procedures on the 9 patients with HHT. 2. Frequency and duration of episodes of epistaxis were reduced following 12/15 procedures in patients with HHT. 3. One serious complication was observed – necrosis of the nasal tip.	This study demonstrates good efficacy of embolization in reducing the frequency and duration of episodes in those with HHT. However, epistaxis was not completely stopped and cannot therefore be seen as a long-term cure. Embolisation was shown to be relatively safe with only 1 documented major complication. The relatively small sample size and non RCT nature are limitations of this study. There may also be some recall bias and subjectivity due to the nature of the study design
S Braak et al., 2009. Rhinology Case-control study	Patients with HHT, n=12 Patients followed up by questionnaire and telephone interview and blood results gathered from hospital records.	Aims: Investigate and measure the results of embolization on epistaxis in patients with HHT End points: - Impact Factor (IF) defined as the daily frequency of epistaxis x severity - Hb levels following therapy - Impact on lifestyle (IoL)	1. IF of the entire group decreased from 10.9 to 3.4 (p = 0.0001) after 1 month and from 10.9 to 7.0 (p = 0.009) after 1 year. 2. The mean Hb improved from 6.4 mmol/l to 7.2 mmol/l (p = 0.045) after 1 year. 3. IoL after 1 year was positive following 12 procedures (63%) and equal in the remaining 7 (37%) (p=0.0001).	The result from this study confirms the effectiveness of embolization in reducing the frequency and severity of epistaxis in patients with HHT. However there may be some recall bias and subjectivity due to the nature of the study design. The relatively small sample size and non-RCT nature are also limitations of this study.
Layton et al., 2007. Am J Neuroradiol Case-control study	22 patients (13 male, 9 female) treated with embolization divided into 2 groups: Patients with HHT, n=12 Patients without HHT, n=10 Patient records and procedure notes were reviewed retrospectively.	Aims: Compare outcomes following endovascular embolization for epistaxis in patients with and without HHT. End points: - Angiographic findings - Requirement for multiple embolisations - Treatment efficacy - Complication rates	1. Angiographic abnormalities were noted in 11 patients with HHT (83%) compared to 3 patients without (30%) (p=0.0062). 2. 50% of patients with HHT required repeated embolization, compared to 10% of patients without HHT (p=0.0743). 3. Embolisation alone was able to control epistaxis in 50% of patients with HHT, compared to 90% of patients with idiopathic epistaxis. 4. 2/12 (17%) HHT patients continued to have significant symptoms despite embolisation and surgery.	This paper demonstrates the efficacy of embolisation in reducing severity and duration of epistaxis in patients with HHT, whilst maintaining a relatively low risk of complications. Embolisation alone does not represent a long term cure for epistaxis in these patients. Limitations relate to the rarity of HHT and include small sample size and the retrospective nature of the study.
Strach et al., 2011. Cardiovasc Intervent Radiol Case-control study	48 patients presenting with intractable epistaxis requiring embolisation divided into 3 groups: Patients with HHT, n=5 Idiopathic epistaxis, n=31 Traumatic/iatrogenic epistaxis, n=12 Patient records and procedure notes were reviewed retrospectively.	Aims: Evaluate outcomes following embolisation for life threatening intractable epistaxis. End points: - Continuing epistaxis - Reduced epistaxis frequency and duration - Cessation of epistaxis	1. 3/5 patients with HHT required 2 embolisations. These patients experienced a reduction in frequency and duration of epistaxis episodes for several years. 2. Severe complications were noted following 2/53 embolisations – necrosis of the nasal tip and transient left hemiplegia lasting 4 days. 3. Clinically indicated repeat embolisation was declined in 2/5 patients.	Reduced frequency and duration of epistaxis was noted in patients with HHT willing to undergo multiple embolisations. However, cessation of epistaxis was not recorded in any patient with HHT. However, the sample size used limits any conclusions that can be drawn from the study.
Trojanowski et al., 2011. Acta Radiologica Case-control study	14 patients with HHT (8 male, 6 female) admitted for endovascular management of epistaxis. Follow up over 24 month period – patients asked to report symptoms.	Aims: Evaluate the efficacy of endovascular treatment of epistaxis in patients with HHT and determine the risk of complications. End points: - Initial control of acute bleeding - Rate of recurrent nasal bleeding - Frequency of epistaxis - Severity of epistaxis - Number of complications	1. Immediate complete control of epistaxis was achieved in 12/14 patients. 2. 6/14 (43%) patients had recurrent nasal bleeding between 6-24 months post-embolisation. 3. 10 patients noted reduced frequency of epistaxis 24 months post-embolisation. 11 patients noted reduced severity of epistaxis. 2 patients were unavailable for follow up. 4. No major complications were noted.	Epistaxis offers a relatively low risk intervention with a high success rate in controlling acute bleeding, making it an attractive treatment option in the management of severe, intractable epistaxis. Furthermore, it can significantly improve quality of life in patients with HHT, in whom epistaxis can be a daily occurrence, by reducing both frequency and severity of bleeding.

Conclusion

Our systematic review has shown that embolisation is a safe and effective means of reducing the frequency and duration of epistaxis in patients with HHT. Complications are few and some authors document a significant improvement in quality of life. However, embolisation alone does not represent a long-term cure with some form of relapse occurring in the majority of patients. Nonetheless, we recommend that embolisation is a valid treatment option and clinicians may continue to opt for it in light of the evidence in favour of it. Further large scale studies with level I evidence are needed to confirm our findings and would be useful for clarification. In its current form, embolisation is not a definitive treatment option but selected patients with recurrent severe epistaxis may benefit.

1. Andersen, P., Kjeldsen, A. and Nepper-Rasmussen, J. (2005). Selective embolization in the treatment of intractable epistaxis. Acta Oto-Laryngologica, 125(3), pp.293-297.

2. Braak S., de Witt C., Disch F. et al. (2009). Percutaneous embolization on hereditary hemorrhagic telangiectasia patients with severe epistaxis. Rhinology. 47(2), pp. 166-171.

3. Layton KF. (2007) Endovascular treatment of epistaxis in patients with hereditary hemorrhagic telangiectasia. American Journal of Neuroradiology. 28(5), pp. 885-888.

4. Strach K., Andreas S, Wilhelm K, et al. (2011). Endovascular treatment of epistaxis: indications, management, and outcome. Cardiovascular and International Radiology. 34(6), pp. 1190-1198

5. Trojanowski, P., Jargiełło, T., Trojanowska, A. and Klatka, J. (2011). Epistaxis in patients with hereditary hemorrhagic telangiectasia treated with selective arterial embolization. Acta Radiologica, 52(8), pp.846-849.