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REVIEW

Epistaxis in people with hereditary haemorrhagic telangiectasia: surgical management and psychological impact

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Abstract

From the emergency management of acute epistaxis to the surgical procedures for chronic epistaxis, this article covers the options available to control the archetypal symptom of hereditary haemorrhagic telangiectasia while exploring the psychological effect such a disease has on the patient.

Key words: Epistaxis; Genetics; Psychology; Surgery

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Introduction

Epistaxis is defined as acute haemorrhage from the nostril, nasal cavity or nasopharynx (Osman and Swift, 2007). For patients with hereditary haemorrhagic telangiectasia (also known as Osler–Weber–Renu disease), spontaneous recurrent epistaxis is by far the most common clinical manifestation and affects 90–98% of patients (Plauchu et al, 1989; Chin et al, 2016). While some patients only experience occasional epistaxis, others have significant bleeds every day (Assar et al, 1991). Almost 50% of patients with hereditary haemorrhagic telangiectasia will have experienced epistaxis by the age of 20 years (Plauchu et al, 1989).

Since hereditary haemorrhagic telangiectasia is a genetic condition, inherited in an autosomal dominant fashion through defects in ENG, ACVRL1 or SMAD4 genes (Plauchu et al, 1989; Faughnan et al, 2011), patients potentially face a lifetime of recurrent epistaxis. This can take a significant psychological toll and demonstrably negatively impact quality of life. Management of epistaxis in people with hereditary haemorrhagic telangiectasia, whether during an acute severe bleed or following more recurrent chronic bleeding, can be challenging, especially as these patients may present to clinicians from a variety of different specialties.

The management of epistaxis in patients with hereditary haemorrhagic telangiectasia is best approached in a stepwise manner. Medical treatment strategies, including the use of antifibrinolytic agents, hormonal and antiangiogenic therapy are well described elsewhere in this issue (see linked paper https://doi.org/10.12968/hmed.2020.0560).

This article discusses the management of acute epistaxis in patients with hereditary haemorrhagic telangiectasia and provides a stepwise guide to the various surgical options available for chronic, recurrent epistaxis. Finally, it explores the psychological impact of both living with hereditary haemorrhagic telangiectasia and quality of life after such surgical interventions.

Management of acute epistaxis in hereditary haemorrhagic telangiectasia

Resuscitation

Clinical guidance for the management of acute epistaxis in the hospital setting is well documented elsewhere (Osman and Swift, 2007) and a structured systems-based approach that assesses airway, breathing and circulation should be performed on arrival, with corrective measures implemented to stabilise the patient in the first instance (as for any critically unwell patient). Once stabilised, a clinical history must be obtained and haemostasis attempted.

Hereditary haemorrhagic telangiectasia affects the structure of blood vessels, resulting in multiple mucocutaneous telangiectasia and arteriovenous malformations. Arteriovenous

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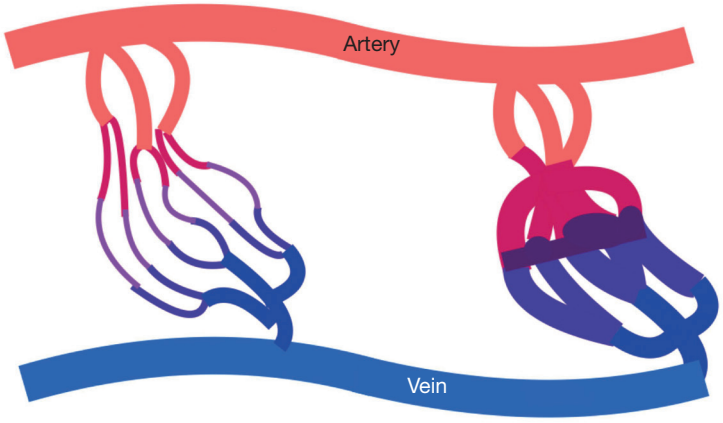

Figure 1. Normal capillary bed (left) vs arteriovenous malformation (right).

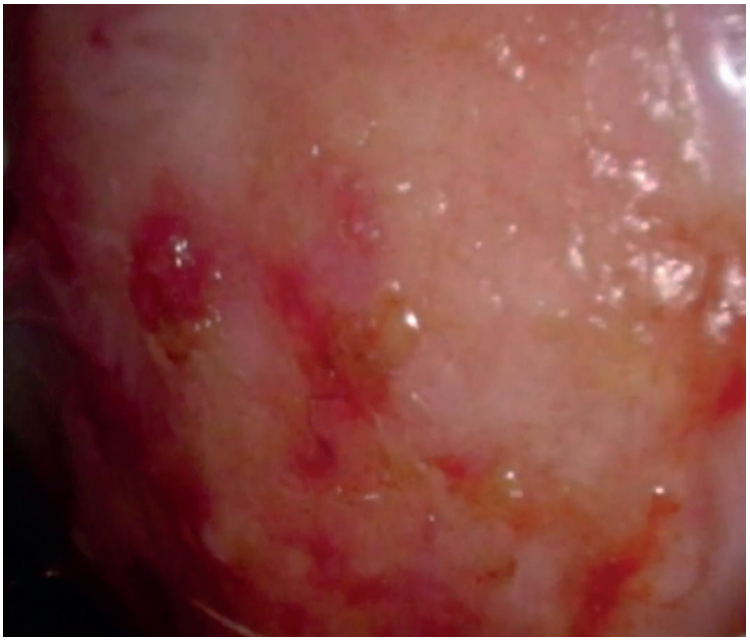

Figure 2. Endoscopic image of right nasal septum displaying characteristic telangiectasic vessels in an adult with hereditary haemorrhagic telangiectasia.

malformations essentially do away with capillary beds, and instead allow transfer directly from the arterial to the venous system (Figure 1). This creates areas with engorged blood vessel complexes within the mucosa which maintain relatively high intravascular pressures (Figure 2). These vessels are fragile and more prone to bleeding, which can be triggered by almost the most benign of activities, usually as a result of local changes in airflow or humidity.

Conservative measures

Broadly speaking, a patient with hereditary haemorrhagic telangiectasia will likely be familiar with basic management of epistaxis and have attempted conservative measures at home before presenting to hospital. This is centred around the Hippocratic method, which involves pinching the anterior cartilaginous part of the nose and bending forward, ensuring no blood is swallowed, for a period of at least 10 minutes continuously. This aims to allow tamponade by bringing the two opposing sides of nasal mucosa together with firm pressure and exerting gentle but continuous force onto the mucosae. Given the frequency with which patients with hereditary haemorrhagic telangiectasia often experience epistaxis, presentation to hospital suggests this is a more significant episode of bleeding and so early involvement of ear, nose and throat specialists is usually advised.

Figure 3. Floseal (Baxter, Deerfield, IL, USA), a biodegradable haemostatic sealant.

Clinical history

It is vital to take a focused clinical history. Table 1 outlines key questions that should be considered and documented at this point.

Source control for haemostasis

If conservative measures with the Hippocratic technique have failed to achieve haemostasis then involvement of ear, nose and throat specialists is generally recommended. Ensuring intravascular volume is maintained, via intravenous fluids and blood products where necessary, is vital at this stage.

When attempting source control in epistaxis in a patient who does not have hereditary haemorrhagic telangiectasia, management options are centred around the use of silver nitrate cautery and/or nasal packing (Melia and McGarry, 2008). The imprecise nature of silver nitrate cautery risks further mechanical damage to the nasal mucosa and can both exacerbate the severity of bleeds and cause septal perforation. As such, silver cautery should generally be avoided in patients with hereditary haemorrhagic telangiectasia (Sautter and Smith, 2016). For the same reason, nasal packing should be avoided wherever possible. Insertion of nasal packing damages the mucosa via frictional forces as it is projected into the nasal cavity alongside outwards circumferential pressure exerted by the cylindrical body when inflated. While in the nasal cavity, the packing exerts significant force onto the mucosa for hours, which causes damage and necrosis even in healthy tissue. The subsequent removal of nasal packing can also damage arteriovenous malformations and trigger further epistaxis.

An alternative strategy is to apply Floseal (Baxter, Deerfield, IL, USA), a biodegradable haemostatic sealant primarily made up of gelatine granules, which exert a degree of

Table 1. Key points when taking a clinical history for acute epistaxis from a patient with hereditary haemorrhagic telangiectasia

Laterality	Which side did the bleeding start from?
Duration	When did the bleeding start? Has it been continuous?
Amount	Can they estimate how much blood has been lost? What has been tried so far to achieve haemostasis?
Recent events	Have they experienced frequent episodes of bleeding in the preceding days? Could there be pre-existing anaemia?
Previous attendances	Have they come into hospital before? What was done to control the bleeding? Did they require a blood transfusion?
Previous medical history and medications	Consider that co-existing conditions or medication may also limit bleeding or the individual's capacity to manage blood loss

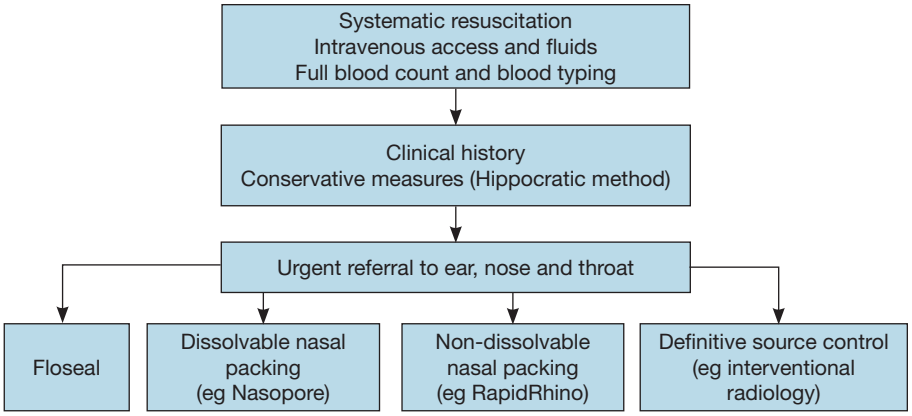

Figure 4. Management of acute epistaxis in hereditary haemorrhagic telangiectasia.

tamponade, and human thrombin, accelerating clot formation. Floseal is effective in the management of epistaxis in patients who do not have hereditary haemorrhagic telangiectasia (Lau et al, 2016) and results suggest it is also effective in those with hereditary haemorrhagic telangiectasia (Lee et al, 2019). Its relative simple administration makes it suitable for self-application and patients with hereditary haemorrhagic telangiectasia may find it useful to have their own supply of Floseal to administer at home in event of epistaxis (Warner et al, 2014).

If bleeding continues and packing is required, the use of dissolvable packing such as Nasopore (Kalamazoo, MI, USA) is preferable to non-dissolvable nasal packs if available (Figure 4). However, if none of the above methods are successful then haemostasis must be prioritised over the risk of potential exsanguination. In this instance, RapidRhino (Smith and Nephew, UK) packing is preferred as it is relatively atraumatic to the nasal mucosa and better tolerated than other non-dissolvable packing options (Iqbal et al, 2017). Once packed, ensure the patient sits upright and forward to prevent aspiration of any residual blood as this may risk obstructing the airway. Monitor closely following packing to ensure adequate haemostasis has been achieved.

There are occasions when severe bleeding may persist despite well-placed nasal packing. In cases of such recalcitrant severe epistaxis, then more definitive source control may be required. Much as with epistaxis in a patient who does not have hereditary haemorrhagic telangiectasia, this can involve endoscopic ligation of the sphenopalatine, internal maxillary or anterior ethmoidal artery or even open external carotid ligation (Osman and Swift, 2007).

If interventional radiological services are available, endovascular embolisation is effective in treating acute intractable epistaxis in the patient with hereditary haemorrhagic telangiectasia (Trojanowski et al, 2011). This spares further instrumentation of the nasal cavity and aims to accurately identify the site of bleeding and superselective embolisation of the contributory vessels.

A pragmatic strategy for the management of acute epistaxis in hereditary haemorrhagic telangiectasia is shown in Figure 4.

Surgical management of chronic epistaxis in hereditary haemorrhagic telangiectasia

As mentioned above, there is a variety of treatment options for patients with hereditary haemorrhagic telangiectasia suffering from chronic recurrent epistaxis, which aims to reduce both the frequency and severity of episodes. The medical management of chronic epistaxis in patients with hereditary haemorrhagic telangiectasia has been covered in other articles in this issue (see linked paper https://doi.org/10.12968/hmed.2020.0560).

Broadly speaking, surgical procedures can be divided into those that use topical therapy and those that are more interventional. In keeping with this, algorithms often suggest a stepwise progression through treatment options based on a combination of disease severity and/or efficacy of previous therapy (Figure 5).

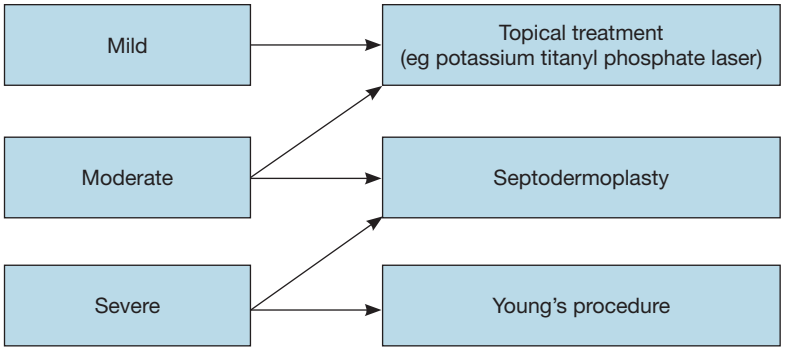

Figure 5. Suggested treatment algorithm for surgical options (as per Rimmer and Lund, 2015).

In essence, topical procedures involve the application of external media onto the target site, such as the use of laser coagulation and coblation. More interventional procedures include septodermoplasty and Young's procedure. Given the number and specialised nature of the treatment options that can be required, management is best provided in a specialist centre with a multidisciplinary approach in which all medical and surgical treatment options are discussed in full. Surgical treatment strategies are outlined further below.

Bipolar diathermy

While bipolar diathermy has a role in the treatment of conventional epistaxis, its use in patients with hereditary haemorrhagic telangiectasia is less clear. While there have been reports of its use to treat larger telangiectasiae (in conjunction with targeted laser therapy; Ghaheri et al, 2006), it may generate deeper thermal injury to surrounding tissues. Such injury carries a risk of further bleeding and possible perforation, leading many groups to avoid diathermy if possible. Monopolar and bipolar diathermy can both be used if a laser is not available. However, caution must be applied to limit collateral damage from the thermal injury of diathermy. When applied with caution, diathermy is low risk and effective, but failure to observe these standards leads to septal perforation.

Laser coagulation

Most case series that report the use of topical treatment concern the use of laser coagulation. Lasers work on the principle of photothermolysis and use light to heat the targeted structure to break open (lyse) the cells. Laser therapy is applied to lesions under endoscopic guidance. Multiple types of laser have been used for patients with hereditary haemorrhagic telangiectasia, although the most commonly used is the KTP laser, which uses a potassium titanyl phosphate (KTP) crystal to generate a beam visible in the green spectrum with a wavelength of 532 nm. This beam is then projected onto the target structure and is either reflected, transmitted or absorbed. The targeted structures (in this case the vascular malformations within the nose) absorb the vast majority of the light energy and this is subsequently converted to thermal energy, ultimately causing destruction at a cellular level (Figure 6). Compared to bipolar diathermy, KTP laser can target smaller lesions and generally offers a greater degree of precision with lower risk of deeper mucosal injury. As such, it can be repeated frequently for these patients as and when new lesions form.

An Nd:YAG (neodymium:yttrium aluminium garnet) laser can also be used although it transfers more heat to surrounding tissues than KTP. The effect of any laser coagulation is not permanent, but such treatment is effective at symptom control in patients with a mild to moderate severity of bleeding (Jorgens et al, 2011).

Coblation plasma coagulation

Coblation is a relatively novel modality which uses radiofrequency energy (usually 20 kHz–300 GHz on the electromagnetic spectrum) to generate a plasma field allowing tissue coagulation. Coblation uses much lower temperatures (60–70°C) than bipolar diathermy (400–600°C), suggesting it could also be of benefit for laser coagulation in hereditary haemorrhagic telangiectasia patients (Luk et al, 2014) although reports of its use are more limited to date.

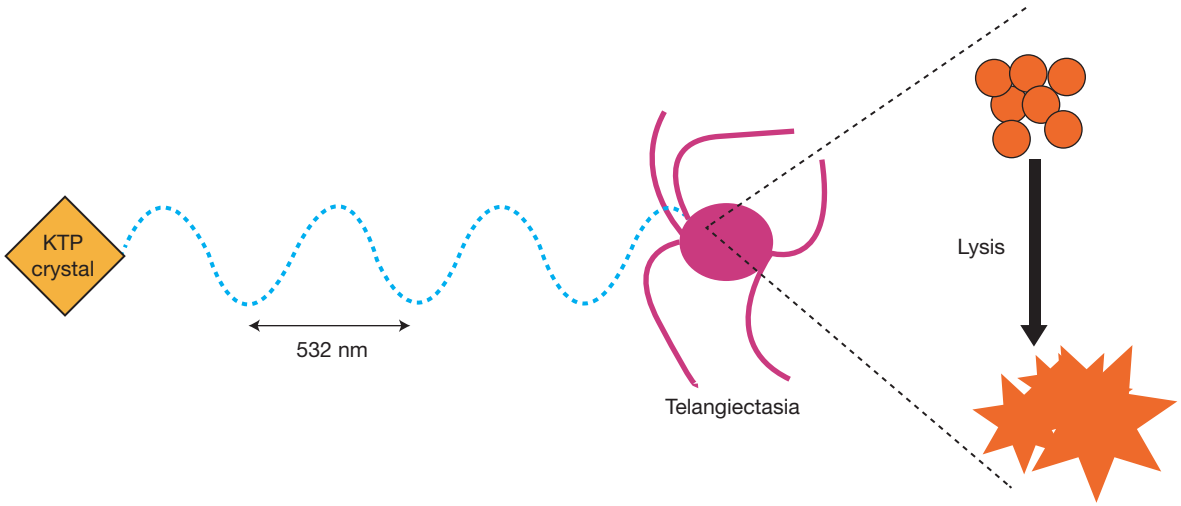

Figure 6. KTP laser principles. The KTP crystal generates a green laser beam with a wavelength of 532 nm. This is targeted at telangiectasia whose cells absorbs the energy of the laser beam. This is transformed into heat energy and the cells undergo lysis (photothermolysis).

Septodermoplasty

Septodermoplasty is a procedure in which mucosa from the anterior portion of the nasal septum, floor and lateral wall is excised and replaced with a split thickness skin graft. This procedure would usually only be considered in patients with hereditary haemorrhagic telangiectasia who have more severe epistaxis or those who have failed to benefit from more conservative treatment strategies. Indeed, it is often used alongside repeated laser treatment. Although the procedure itself is relatively destructive, and the loss of native mucosa can cause both increased nasal crusting and loss of olfaction, it reduces epistaxis severity and the frequency of laser treatment required (Harvey et al, 2008). However, results are not uniform for all patients and there are risks of both graft failure and symptom recurrence with time as lesions recur in the nasal mucosa.

Young's procedure

Young's procedure is typically performed for patients with hereditary haemorrhagic telangiectasia with severe epistaxis refractory to other treatments. In these patients, bleeds can be potentially life threatening and this surgical procedure closes the nostril. As shown in Figure 7, it involves a circumferential incision at the mucocutaneous junction before three radial flaps are brought down to the alar rim and sutured together at the entry point into the nose to achieve complete nasal closure.

Ultimately, this prevents air from entering the nostril, preventing turbulent airflow over the delicate, friable arteriovenous malformations, drying them out and causing them to bleed. This is best considered as a definitive procedure as there are reports of severe bleeding following reversal. Although beneficial results have been shown following the use of a nasal obturator (Soni-Jaiswal and Woolford, 2009), insertion and removal of such a device can induce bleeding and when removed from the nose, an obturator is of no benefit as nasal airflow will obviously return. While Young's procedure may seem a radical step, it is extremely effective in such patients, with complete cessation of bleeding reported in the majority of patients (Lund et al, 2017). Patients need to be adequately counselled on the side effects of the procedure, given the effects on smell and taste and the mouth dryness which will follow with life as an obligate mouth-breather.

Psychological impact of living with hereditary haemorrhagic telangiectasia

Compared to the work proposing treatment strategies for patients with hereditary haemorrhagic telangiectasia, relatively few studies have considered the psychological impact of the condition. Studies have used quality of life measures and qualitative approaches to explore the everyday impact and lived experience of people who have hereditary

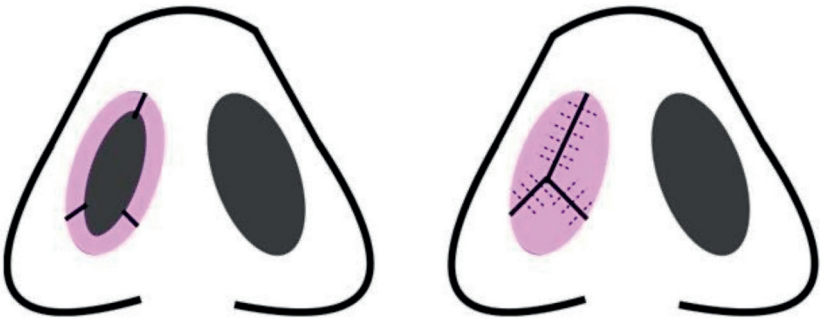

Figure 7. Young's procedure. Three radial flaps are created and are sutured together at the alar rim. This closes the nostril and blocks airflow into the nostril, ultimately stopping turbulent air from flowing over the delicate telangiectasiae and triggering epistaxis.

haemorrhagic telangiectasia. Reductions in quality of life measurements in patients with hereditary haemorrhagic telangiectasia have been reported (obtained using the commonly used Short Form-36 Health Survey; SF-36), with the severity of epistaxis correlating with such scores (Geisthoff et al, 2007; Ingrand et al, 2011). Qualitative studies highlight that, while there is no single typical experience of hereditary haemorrhagic telangiectasia (Sexton et al, 2019), there are aspects of the symptoms, patient journey, coping and interaction with healthcare which contribute to the psychological impact of the condition.

Visible symptoms, including epistaxis, have led to experiences of social stigma for some people living with hereditary haemorrhagic telangiectasia. For example, other people reacting to the sight of blood or fearing that the blood could carry the risk of contracting infectious diseases have led to feelings of embarrassment and anxiety, and reduced confidence. Such experiences can impact more on mental health than the severity of the physical symptoms themselves (Sexton et al, 2019). People living with hereditary haemorrhagic telangiectasia have described that experiences and emotions connected with epistaxis (including frustration and embarrassment) led them to restrict their activities and worry about their future health (Higa et al, 2016).

Before diagnosis of hereditary haemorrhagic telangiectasia, people living with the condition have reported feeling trivialised when presenting for medical opinion, contrasting with later feelings of shock, uncertainty and anxiety upon diagnosis (Geerts et al, 2017). Others have experienced diagnosis as a psychological relief, having previously felt like a 'hypochrondriac', with the diagnosis explaining symptoms (Sexton et al, 2019).

Diagnosis of hereditary haemorrhagic telangiectasia as a genetic disease has implications for the individual and their existing and future family (Geerts et al, 2017). Some patients have experienced genetic testing as merely confirming the clinical diagnosis, and therefore not a significant event, while others have valued the genetic diagnosis for information regarding reproductive risk and options (Sexton et al, 2019).

In efforts to respond and cope with the unpredictability of symptoms, people living with hereditary haemorrhagic telangiectasia have described trying to identify triggers (eg certain foods, temperature and hormonal cycles) to reduce the uncertainty and increase their sense of control. Some patients have also coped with their symptoms by minimising them and considering themselves fortunate to others with hereditary haemorrhagic telangiectasia who have more severe difficulties. People have also spoken about hereditary haemorrhagic telangiectasia not necessarily being a focus of their lives, despite it always being there, and trying to remain positive alongside anxiety about future symptoms (Sexton et al, 2019).

As already discussed, treatment for hereditary haemorrhagic telangiectasia is best delivered in a specialist setting. Negative experiences of suboptimal treatment have instilled the importance of self-advocacy in many patients with hereditary haemorrhagic telangiectasia (Sexton et al, 2019). Assuming a role as an expert patient enables patients to exert more control within clinical interactions and is a beneficial coping strategy used by many people with other complex chronic conditions (Boulet, 2016). As most patients have family members with the condition, patients with hereditary haemorrhagic telangiectasia

are generally very well informed and often request specific treatments that they know have proven effective for others close to them (Lund et al, 2017).

While self-advocacy was described as important, coordinated care has been frequently mentioned by those who felt fortunate to have accessed it, or by those who wished they could have help with coordination of various specialist services. Where there had been experiences of supportive partnerships with doctors, people described their doctors listening to them and respecting their experiential knowledge of hereditary haemorrhagic telangiectasia. Those who had not experienced effective partnerships described experiences of feeling not heard or dismissed (Sexton et al, 2019). This can lead to difficult medical consultations, especially if the examining doctor has not encountered this situation before and lacks insight or empathy into the situation.

As already discussed, these procedures can have a major impact, with both septodermoplasty and Young's procedure aiming to reduce the severity of epistaxis usually at the expense of considerable losses in both olfaction and taste. While significant improvements are seen in SF-36 quality of life measurements in patients undergoing Young's procedure (Hitchings et al, 2005), it should be noted that patients in this series who underwent nasal closure had previously had multiple procedures performed at a number of other institutions, which will undoubtedly frame their mindset and views on this treatment. The views of a patient with hereditary haemorrhagic telangiectasia undergoing Young's procedure as their first treatment modality may well differ. While there is good evidence that complete closure of both nostrils is effective at controlling bleeding, many patients refuse this option. Loss of the nasal airway is a huge disability to most people and is only accepted when the bleeding becomes so severe that the patient is happy to accept this deficit. It is paramount that the severity of a patient's experience of hereditary haemorrhagic telangiectasia be matched to the treatment options used and the level of support provided to allow patients to make informed decisions in their care.

Ultimately, hereditary haemorrhagic telangiectasia can pervade every aspect of a patient's life, including work, home life, family life, activities, family relationships and intimate relationships. This leads to concern regarding the future and the worsening of symptoms, often exacerbated when family members die as a result of the condition. These factors significantly influence a patient's decision making when discussing surgical management options with their clinician.

Given the psychological impact of the patient journey and interactions with healthcare systems, it is likely that this area will evolve with increased awareness of hereditary haemorrhagic telangiectasia among professionals and the development of specialist centres. It is also likely that experiences will vary around the world in different healthcare contexts. Additionally, education and raising awareness in the general population may reduce fear of, and stigmatising responses to, symptoms such as epistaxis. Alongside promoting positive experiences of healthcare and responses from others, approaches that may be useful in responding to the psychological impact of living with hereditary haemorrhagic telangiectasia include individual psychological therapy, family therapy and opportunities for peer support.

Conclusions

As a multisystem disease, hereditary haemorrhagic telangiectasia can present to different specialities, so it is important for all clinicians to understand this condition. Whether treating patients with hereditary haemorrhagic telangiectasia acutely, with severe epistaxis, or in a more elective setting with chronic recurrent epistaxis, it is key to recognise the differences in management in comparison with more conventional epistaxis. Crucially, a stepwise management plan should be taken with involvement of those with specialist interest to provide optimal treatment and establish appropriate multidisciplinary support for patients living with this chronic condition.

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Key points

- Epistaxis resulting from hereditary haemorrhagic telangiectasia must be managed differently from conventional epistaxis.
- Surgical management of hereditary haemorrhagic telangiectasia is best provided in a specialist centre with a multidisciplinary approach in which all medical and surgical treatment options are discussed in full.
- Hereditary haemorrhagic telangiectasia can have a negative impact on the quality of life of patients with the disease. Clinicians in any speciality who encounter patients with hereditary haemorrhagic telangiectasia should be aware of how pervasive this disease is and encourage patients to assume the role of the expert patient.
- Identification of patients with hereditary haemorrhagic telangiectasia is important to allow a coordinated, step-wise management plan from various medical and surgical specialities.

Conflicts of interest
The authors declare that there are no conflicts of interest.

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