xxx's Diary: A Novel Symptom Monitoring Strategy for Epistaxis

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May 20, 2022

Abstract

Objectives To assess the efficacy and implementation of an epistaxis symptom diary in the management of a child with de-novo HHT type 1. HHT is a cause of chronic, severe epistaxis which can lead to significant physical and psychogical morbidity. We propose that the use of symptom diaries can reduce associated patient morbidity and aid physicians in treatment planning. Participants We discuss an independently developed symptom diary for a 10-year-old girl with HHT, Patient x ("xxx"), and its subsequent impact. Including its uses for surgical and medical treatment planning. Design A patient developed epistaxis symptom diary was designed to including nosebleed timing and side of onset, estimated severity, exacerbating / trigger factors and use of medications. A phenomenological qualitative approach was taken to assessing the impact of the diary on "xxx's" physical and psychological wellbeing. Setting "xxx's" diary is a novel symptom diary for patients with of chronic epistaxis and HHT which can improve patient engagement with treatment, foster lifestyle changes, and aid clinicians in promoting tailored patient-centred care. Main outcome measures "xxx's" diary has proved to be an extremely useful tool for patient xxx, her parents and her ENT team, having improved both her physical and psychological wellbeing. Results and Conclusions We would recommend detailed symptom monitoring for all patients with severe epistaxis and HHT and suggest "xxx's" diary as a template for this.

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Results and Conclusions

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Keywords

Epistaxis, HHT, diary, symptom, monitoring

Key points

1. Chronic epistaxis can significantly impair quality of life for its sufferers
2. Hereditary haemorrhagic telangiectasia is a rare but serious form of chronic epistaxis
3. There is good evidence for the use of symptom diaries in chronic disease; improving patient outcomes and increasing sense of ownership over one's illness.
4. The use of a symptom diary for chronic epistaxis can improve patient outcomes and experiences, both physically and psychosocially.
5. We recommend all patients with chronic epistaxis and HHT keep a symptom diary with a format similar to xxx's

Introduction

HHT is a genetic vascular disorder characterised by disseminated vascular lesions in many organs including the lungs, liver and brain. For the ENT surgeon, the most relevant of these vascular malformations are telangiectasia of the nasal mucosa which are very friable and leave patients prone to recurrent and severe epistaxis.

HHT is an autosomal dominant inherited condition with an incidence of approximately 1 in 5000-8000 live births. Diagnosis is via the Curaçao criteria, an internationally recognised standard, with patients scoring from 0-4 based upon several categories *(figure 1)* . A score of 3-4 indicates '*definitive'* HHT, whilst 2 is '*suggestive'* and 0-1 is '*unlikely'* . These scores are both diagnostic and particularly useful to rule in a possible HHT diagnosis in younger adults and children. Genetic testing can be performed to identify the causative genetic defect in an index case, thereby allowing a diagnosis amongst family members who do not fit diagnostic criteria. Genetic screening for HHT focuses on two primary gene mutations: ENG (HHT type 1) and ACVRL1 (HHT type 2). Together these two genes are responsible for approximately 96% of HHT cases. Whilst these two forms of HHT are the most common, genetic testing is complex. Some families have 'private' HHT mutations with compelling evidence of HHT without any identifiable genetic mutation, further emphasising the importance of thorough history and examination.

Epistaxis affects up to 90% of patients with HHT and can significantly impact these patients' quality of life (QoL) both physically and psychologically. Most sufferers report epistaxis as the symptom that interferes most with their day-to-day activities, however, related symptoms such as fatigue from iron deficiency anaemia and nasal crusting or blockage cause additional distress. Frequency and duration of epistaxis are the most significant contributors to impaired QoL in patients with HHT.

Management of HHT is complex and multimodal; encompassing prophylactic, medical, and surgical methods. Importantly, treatment should be individually tailored and involve substantial collaboration between doctor and patient, considering physical and psychosocial aspects to improve patients' perception of themselves and their illness .

Individualising care of patients with HHT can be challenging, particularly as the main end-points of assessing successful treatment and patient satisfaction are the frequency, duration, and quantity of nose bleeds. The reporting of these symptoms is susceptible to recollection biases ; the under or over-reporting of symptomatology by patients. This makes tailored treatment more challenging. We report the case of a 10-year-old girl who developed a nosebleed diary to accurately and effectively track her symptoms over time and against the initiation of different treatments.

Clark et. al recommends that the following should be recorded in a nosebleed diary: date and time, duration, intensity, severity, and the need for medical attention. Whist the authors discuss the development of an e-diary for patients with HHT, we have found no reports of a symptom diary being used to monitor care and adjust management in these patients.

Background

"xxx" presented with epistaxis in 2019 to the Royal Victoria Infirmary paediatric ENT department, Newcastle Upon Tyne NHS Foundation Trust. She had been experiencing recurrent bilateral epistaxis since the age of 4 and first presented to our department at the age of 7. General examination was significant for cutaneous telangiectasia on her left cheek and both hands. Family history identified her maternal grandmother and maternal granduncle suffering from epistaxis but no family history of diagnosed HHT. Examination under anaesthesia of her nose identified multiple mucosal telangiectasias which were subsequently cauterised. Given her symptoms of recurrent epistaxis and the presence of telangiectasia (Curaçao score 2), she was referred for genetic testing which established a de-novo ENG mutation consistent with HHT type 1.

Treatment was initiated with prophylactic twice-daily applications of nasal petroleum jelly and daily Sterimar spray. Tranexamic acid was provided for "xxx" and her parents to administer as a rescue medication in the case of particularly severe episodes.

Methods

The COREQ guidelines for qualitative research have been used to present our methodology.

Two interviews were conducted with "xxx" and her Mother across a two month time period. Initially an interview was conducted electronically by email and then a follow-up telephone interview was used to clarify any ongoing questions and to enable a more nuanced emotional assessment. The interviews were conducted by the primary author, a male junior doctor working in otolaryngology who has received training in qualitative research through the completion of a postgraduate diploma in medical education. The interviewees were knowlageable about the goals of the study and participated out of a willingness to help others with HHT and informed consent was provided.

A phenomenological approach was taken to interviewing and data analysis given the experiential nature of the questions to be answered. Sampling was purposive with a sample size of one. Data were collected at the time of the interviews via field notes. The telephone interview lasted approximately 45 minutes. Data analysis was performed by the primary author and a third party using thematic analysis; themes were compared for consistency and congruency.

Findings

Shortly after her diagnosis, "xxx" began to record her nose bleeds in her homework diary, enabling her and her parents to keep a track of her symptoms. Initially, she recorded the times of each nose bleed in addition to an estimated severity. She also included any possible exacerbating or trigger factors such as occurring

during activity or in a certain location. As medications were introduced "xxx" incorporated their use into her symptom diary, an example of this can be seen in appendix 1. Although "xxx's" diary was developed independently, the recording of her symptoms was in keeping with recommendations from Clark et al. with some minor variations.

"xxx" developed a coding system to easily document events. Blue for right nostril bleeds, green for left nostril bleeds, a yellow highlight for oral tranexamic acid and a red highlight for topical tranexamic acid. Through the use of different colours and highlights, it is easy to see at a glance not only the number of nose bleeds in each month but the more affected side and the impact of medications on future bleeds. By enabling retrospective review of medication efficacy "xxx's" diary has indicated that although tranexamic acid can help control an active nose bleed, its use has little effect on her future epistaxis frequency.

A particularly notable use of "xxx's" diary was to time and monitor the effect of nasal coblation therapy. At the end of 2020 nosebleed frequency was seen to increase from a baseline of around 9-11 per month to 16-17 per month (see appendix 2). Noticing this increase, "xxx's" parents were able to initiate early follow-up with both her GP to arrange blood tests to check haemoglobin levels and with our unit for an expedited appointment. Coblation therapy was subsequently arranged for 16/02/2021. Appendix 2 demonstrates the efficacy of the coblation therapy with an immediate reduction of nose bleeds to 3 per month with none heavier than 'small'. Not only was it useful for our team to have objective evidence of a significant improvement following coblation but it has been helpful for "xxx" and her parents to look back at this time and recall how effective coblation treatment can be. In this way, her diary has helped "xxx" to view future treatments and their likely necessity in a more positive light.

Discussion

Symptom diaries have a long history in the medical literature with benefits including patient engagement, trigger identification, and information gathering. Severe nosebleeds have always been a feature of life for "xxx" and something that for the most part has been out of her control. The creation of her symptom diary allowed her to form a sense of ownership and understanding over her condition and accept it as a part of herself. Chronic disease can strip suffers (and particularly children) of a sense of self and autonomy as decisions are gradually taken away from them. "xxx's" diary has returned some of this control back to her and has allowed her to finesse the use of her rescue medications.

By removing cognitive and recollection biases, epistaxis diaries are a useful tool to review in clinic. Objectively measuring the efficacy of treatment and symptom severity can enable more personalised treatment and allow surgery to be booked for a time when its efficacy will be maximal.

Surgery, particularly surgery under a general anaesthetic is not completely benign. It is accompanied by not only physical risks but additional stress and anxiety for patients and/or their carers. This is also true on a lesser scale for minimally invasive procedures such as blood tests, especially in the case of children. A symptom diary can help to minimise exposure to these procedures by only performing them at a time when their impact will be maximised or when symptoms have become unmanageable. Diaries can help to empower patients to engage in shared decision making regarding these choices; the bidirectional sharing of key information between patient and doctor will help to guide ongoing management in colaberative way that improves the satisfaction of both parties.

The important role of patient self-care in chronic conditions such as diabetes and asthma is well understood, however, there is little research as to the benefit of self-care and disease engagement in patients who suffer from chronic epistaxis. Hayama et al. found that although HHT can significantly impact QoL proportionally to epistaxis severity, less than 50% of surveyed participants used prophylactic medications or took lifestyle precautions. We suggest that by facilitating the use of symptom diaries, patients will be encouraged to take ownership of their disease, identify symptom triggers and therefore take additional precautionary measures.

Conclusion

We would recommend that all patients with HHT or severe epistaxis keep a symptom diary similar to "xxx's". We suggest that patients should record a minimum of date and time of nosebleed onset, side of bleed, estimated severity, any potential influencing factors, and use of treatments or medications.

An effective symptom diary would enable clinicians to quickly identify patterns over time and for patients to gain a more objective understanding of their condition. We suggest that "xxx's" diary could be such a standard.

Bibliography

Criteria	Description
Epistaxis	Spontaneous and recurrent
Telangiectasias	Multiple, at characteristic sites: lips, oral cavity, fingers, nose
Visceral lesions	Gastrointestinal telangiectasia, and pulmonary, hepatic, cerebral or spinal arteriovenous malformations
Family history	A first-degree relative with HHT according to these criteria

Appendix 1

Example month from "xxx's" epistaxis diary. Times and approximate nose bleed quantity are recorded. Yellow boxes indicate the use of rescue oral tranexamic acid. Blue for the right nostril and green for the left nostril.

Mon	Tue	Wed	Thu	Fri	Sat	Sun
					1	2 AM - small (bath)
3 AM - medium	4 AM - medium Night - small	5 AM - small	6 Night - small	7	8 AM - small	9
10	11 PM - big	12	13 PM - medium	14	15	16 AM - small
17	18	19	20	21 AM - small	22 Night - small	23
24 AM - small	25	26 PM - medium (at school)	27	28	29 PM - big	30
31						

Appendix 2

Impact of coblation therapy on nose bleeds as recorded in “xxx’s” diary

January 2021

Mon	Tue	Wed	Thu	Fri	Sat	Sun
				1 AM - small	2 AM - small	3 AM - small
4	5 PM - small	6	7	8 Night - medium	9 PM - small	10
11	12 AM - small	13	14	15 Night - medium	16	17 Night - medium
18	19 Evening - medium (shower)	20 AM - small	21	22	23 Night - small	24 Night - medium
25	26 Night - small	27	28 AM - small	29	30 AM - small	31 Night - medium

Right = 11
Left = 5
Total = 16

February 2021

Mon	Tue	Wed	Thu	Fri	Sat	Sun
1	2 AM - small PM - medium	3 Night - small	4	5	6 Night - medium	7
8	9 AM - small	10 Night - medium	11	12	13 AM - small	14 AM - small
15	16 AM - small Coblation	17	18	19	20	21
22	23	24	25	26	27	28

Right = 9
Left = 0
Total = 9

March 2021						
Mon	Tue	Wed	Thu	Fri	Sat	Sun
1	2	3	4	5	6	7
8	9	10	11	12	13	14
15	16 Night - medium (sneeze)	17	18	19	20	21
22	23	24	25	26 AM - small	27	28 AM - small
29	30	31				

Right = 2
Left = 1
Total = 3

April 2021						
Mon	Tue	Wed	Thu	Fri	Sat	Sun
			1	2	3	4
5 AM - small (COVID test)	6	7	8	9	10	11
12	13	14	15	16	17 AM - small	18
19	20	21 AM - small (COVID test)	22	23	24	25
26	27	28	29	30		

Right = 2
Left = 1
Total = 3