ORIGINAL CONTRIBUTION

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A grading scale for epistaxis in hereditary haemorrhagic teleangectasia*

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SUMMARY

Background: *Epistaxis is the most common symptom in patients with Hereditary Haemorrhagic Telangiectasia (HHT). Different institutions are using different treatment modalities and different grading systems. The treatment options depend on the grade of epistaxis. It is important to have a common grading system to compare and evaluate the effectiveness of different treatment options. Furthermore, it is important to correlate quality of life with an epistaxis grading system. The aim of this work was to propose a new grading system for epistaxis in HHT.*

Methods: *A medical literature search was performed for grading systems of epistaxis in HHT. A questionnaire on five criteria's for a new grading system was sent to 22 internationally renowned medical experts, who have published results on epistaxis in HHT.*

Results: *Four different grading systems are currently in use for the grading of epistaxis in HHT. The response rate of the questionnaire was 43%. All the experts who answered the questionnaire agreed that the aimed grading system should be easy to understand for the patients. 90% of them wanted the system to focus on a definite time period. 70% answered that blood transfusion should be included in the grading system as an important factor. There was no clear consensus on whether the system should be a single multi-item scale or a composite scale consisting of more than one single scales, and similarly there was no clear consensus on whether is should be an absolute or a relative scale.*

Conclusion: *The proposed system should be easy to understand for the patients, focus on a definite time period of observation, and include blood transfusion as one of its parameters. For statistical reasons, an epistaxis grading scale with at least one absolute end point would be preferable.*

Key words: epistaxis, epistaxis classification, epistaxis grading, hereditary haemorrhagic teleangectasia, Osler disease

INTRODUCTION

Hereditary Haemorrhagic Teleangiectasia (HHT) also known as Rendu-Osler-Weber disease is an autosomal dominant non-sex linked disorder of the fibrovascular tissue, characterized by arterio-venous malformation and multi-systemic haemorrhage from multiple teleangiectasias throughout the skin and mucous membranes. Indeed epistaxis is the most common presenting symptom in 90% of the patients [1-4]. The condition is more common in women with a female to male ratio of 5:1. Serious nose bleeds occur in 80% of the patients by the age of 30 years [5,6]. The severity of epistaxis associated with HHT ranges from infrequent and mild epistaxis to frequent and severe bleeding requiring hospital admission, blood transfusions or multiple surgical interventions [7]. Management of epistaxis associated with HHT can vary depending on the grade of epistaxis [8]. Different therapies have been used. However, some of these methods are more successful than others while some methods have serious side effects and complications that need to be taken into consideration .

By reviewing the studies about epistaxis in HHT, it became obvious that different institutions are using different treatment modalities and different grading systems in order to assess the severity of epistaxis in HHT. This makes it difficult to compare and evaluate the effectiveness of different treatment modalities in treating epistaxis of different grades. Therefore it is necessary to aim for a common and internationally accepted grading system for epistaxis in HHT.

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METHODS

Literature search

To propose a new grading system for epistaxis associated with HHT, the literature has been searched in Pub-Med, using the following keywords: epistaxis, epistaxis AND grading system, epistaxis AND classification, epistaxis AND Osler disease, epistaxis AND hereditary haemorrhagic teleangiectasia.

Questionnaire

Expert opinions were collected using a questionnaire. Five questions were sent to 22 international medical experts, who have published results on epistaxis in HHT:

1) Should a grading system be divided up into two scales? For example: one scale for frequency and another scale for severity or should it be a "single multi-item scale".
2) Ideally, should a grading system consist of a relative scale or an absolute scale?
3) Is the need for blood transfusion an important parameter? Keep in mind that the patient may bleed simultaneously from other sites than the nose.
4) For research purposes, should a grading system be easy to understand for the patient? For example when the patient is using a diary.
5) Should a grading system focus on the duration of one single bleeding episode or should it focus on a definite time period (like for example one-month observation time).

In addition the questionnaire gave room for comments.

RESULTS

Four different grading systems have been applied for the grading of epistaxis in HHT.

The first one depends on a disease-specific questionnaire assessing the severity and frequency of bleeding using a visual analogue scale and information regarding other treatments such as blood transfusion. In addition the quality of life is taken in consideration. According to this system, epistaxis secondary to HHT is graded into: mild, moderate and severe (Figure 1) [1,6,7].

The second grading system categorize the degree of epistaxis in HHT according to the criteria developed by Reibez et al., into mild, moderate and severe (Table 1). In this system the grading depends on the frequency and duration of bleeding as one item, and the need of blood transfusion as another item and put these 2 items in one scale (single multi-item scale). A fourth grade (intractable bleeding) has been used in this system.

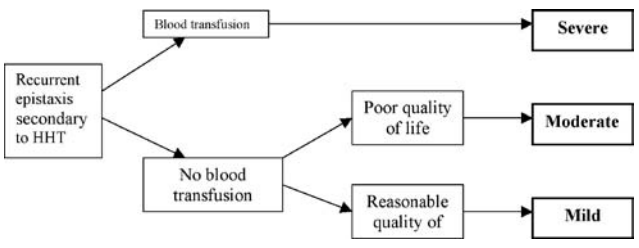

Figure 1. Grading epistaxis according to Lund et al.

Table 1. Grading epistaxis according to Reibez et al.

Severity of epistaxis	Frequency of epistaxis	No. of transfusions
Mild	Few episodes / week	None
Moderate	1-2 time / day	< 10/ lifetime
Severe	Daily epistaxis lasting greater than 30 min	> 10/ lifetime

Table 2. Grading according to Bergler et al.

Intensity of bleeding	**Freguency of bleeding**
Grade 1: slight stains on the handkerchief	Grade 1: less than once a week
Grade 2: soaked handkerchief	Grade 2: a few times a week
Grade 3: bowl or similar utensil necessary	Grade 3: more than once a day

The third system is proposed by Bergler et al. This is a two-scale system in which the severity (intensity) of bleedings is taken as one scale, graded into three grades from 1 to 3 while frequency of bleeding is taken as another scale, graded into other three grades from 1 to 3 (Table 2) .

The fourth system was published by Pagella et al. According to their system, the severity of epistaxis is established by the history of blood transfusion, the frequency and the duration of bleeding episodes [9] (Table 3).

The response rate of the questionnaire was 43%.

Table 3. Grading according to Pagella et al.

Necessity of blood transfusion	
1	None
2	< 10 in lifetime
3	> 10 in lifetime
Frequency of epistaxis	
1	Less than once a week
2	Several times a week
3	Several times a day
Length of epistaxis	
1	< 10 min
2	Between 10 min and 30 min
3	> 30 min

Question 1): 50% preferred a "single multi-item scale", while the other 50% preferred more than one scale.

Question 2): 60% chose the absolute scale, and 40% chose the relative scale.

Question 3): 70% answered that blood transfusion is an important parameter, while 30% answered that blood transfusion is not an important factor.

Question 4): 100% decided that the system should be easy to understand for the patient.

Question 5): 90% wanted the system to focus on a definite time period, while 10% wanted it to focus on a single bleeding episode.

Table 4. Epistaxis grading scale.

Observation of intensity, frequency and blood transfusion during a period of 4 weeks.		
Intensity of the bleedings (I)	**Frequency of the bleedings (F)**	**Blood transfusion (T)**
0 None	**0** None	**0** None
1 Slight stains on the handkerchief	**1** 1-5 times	**1** Once
2 Soaked handkerchief	**2** 6-10 times.	**2** More than once
3 Soaked towel	**3** 11-29 times	
4 Bowl or similar vessel is necessary	**4** Daily bleeding	

Comments

One of the comments received, suggested the use of haemoglobin level instead of number of blood transfusions. Another suggestion was frequency of hospital attendance because of epistaxis can be taken in consideration, and this can be taken as a third scale as in TNM classification of malignancies.

DISCUSSION

All the four systems in use nowadays were easy to understand for the patients. Two of the four systems were *multi-scale* systems (Table 2 and Table 3), while the other 2 used a *single multi-item scale* (Figure 1 and Table 1). All the existing systems were relative systems and did not take at least one absolute end point into consideration. Blood transfusion was taken as a factor in the grading in 3 systems (Figure 1, Table 1, and Table 3). The systems that are in use now either did not focus on a definite observation period (Figure 1) or the observation period was not a fixed period but varied (week, day or lifetime) from one grade to the other in the same system. At least three systems took both the intensity of the bleeding and the frequency as 2 separate parameters. One of the existing systems took QoL as a dimension in the grading.

All the medical professionals, who have answered the questionnaire, agreed that the grading system should be easy to understand for the patients. The majority wanted the system to focus on a definite period of time not on a single bleeding episode. There was tendency to regard blood transfusion as an important factor. There was no clear tendency as to whether the system should be a one multi-item scale or a scale consisting of more than one scale, and similarly there was no clear tendency towards an absolute scale or a relative one.

By studying the existing systems and the presented questionnaire, we concluded that the ideal system should be easy to understand for the patients, focus on a definite time period of observation, and blood transfusion should be included as a parameter in the grading. It is obviously that the intensity and frequency of bleeding are important to consider in a grading system. Since the frequency, intensity and blood transfusion should be considered, the proposed system should be multi-scale. We are convinced that this is an important aspect, although there were no clear tendencies, neither in the questionnaire nor in the existing systems towards an absolute scale. As a measure of prognosis, a grading system with at least one absolute end point is advantageous. In addition, patients who stop bleeding completely after treatment would be lost for statistical calculation when using a grading scale without a zero grade.

As a consequence, a grading system is proposed (Table 4) which is a multi-scale grading system, composed of 3 scales. There is one absolute end point of the scale, which is zero. It focuses on a definite time period of observation which is one month. The system is meant to be used in a similar way as the TNM classification system of tumours by using the abbreviation (I) for the intensity of bleeding, (F) for the frequency and (T) for blood transfusion. The digits corresponding to the appropriate intensities or frequencies are added.

For example, a patient has been bleeding 16 times during the last month, with some slight stains on the handkerchief. In addition, he had 2 episodes of severe bleeding, filling a bowl and he needed a blood transfusion on one occasion. His epistaxis grading would be I_5 F_3 T_1. During the first month after treatment this patient had bloodstains on a handkerchief just ten times, there was no need for blood transfusions, so the epistaxis grading is I_1 F_2 T_0.

Correlation with QoL

The proposed system concentrates on epistaxis as the main symptom in patients with HHT and focuses on grading an objective finding. It does not take into consideration the patient's personal subjective evaluation of the effect of epistaxis on their QoL. In research, the personal patient perception of the QoL is an important parameter in their evaluation and in comparing different treatment outcomes. Some patients with mild bleeding may have high alteration of their QoL and might require more aggressive therapy than others with normal QoL. The QoL has already been used in one of the existing grading systems (Figure 1) [6]. The ability of SF-36 to measure the QoL of the patients with epistaxis due to HHT has been evaluated in other studies and has proven to be valuable [10,11].

The aim of this work was to find a proposal for an objective grading system of epistaxis in HHT. The question on how the proposed epistaxis grading will correlate with a QoL system needs further research.

CONCLUSION

Epistaxis is the most frequent symptom in HHT, but it varies in intensity and frequency. Several therapy modalities have been used in this field. The indication and effectiveness of these modalities depends on the severity of epistaxis. Different institutions are using different treatment modalities and different grading systems and there are at least 4 different grading systems for epistaxis in HHT. This makes it difficult to compare and evaluate the effectiveness of different treatment modalities in treating different grades of epistaxis from different institutions. A common internationally accepted grading

system would help to choose the type of treatment and to measure the response and realize cost-effectiveness value. It is important both for research purposes and daily clinical routines in this field.

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