ORIGINAL CONTRIBUTION

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Psychosocial quality of life in hereditary haemorrhagic telangiectasia patients

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SUMMARY

Objectives: *The aim of this study was to evaluate psychosocial quality of life (PQoL) in patients with Hereditary Haemorrhagic Telangiectasia (HHT).*

Study Design and Setting: *A retrospective study was performed on PQoL in HHT patients presenting with epistaxis. One hundred fifteen patients were interviewed using a questionnaire designed by two sociologists and a head and neck surgeon. Changes over time were assessed according to information on psychosocial well-being, social life, family support, occupation, and medical and demographic data regarding age, gender and patient appearance.*

Results: *Analysis of Psychosocial Quality of Life (PQoL) revealed no statistical difference in relation to gender, marital status, household income or place of residence (rural or urban); however, a significant difference was observed with age. Elderly patients had a poorer PQoL than younger patients. Workers had a better PQoL than unemployed patients. Epistaxis and professional duties were correlated: workers with less than one episode of epistaxis per month were more active. Frequent episodes of epistaxis and abundant bleeding decreased PQoL. These patients felt different and often experienced a desire to withdraw compared to others.*

Conclusion: *Epistaxis in hereditary haemorrhagic telangiectasia patients was associated with the impairment of many PQoL criteria, together with relationship modifications.*

Keywords: *Hereditary Haemorrhagic Telangiectasia, Osler-Weber-Rendu Disease, epistaxis, Quality of life*

INTRODUCTION

Hereditary Haemorrhagic Telangiectasia (HHT) is a hereditary autosomal dominant vascular disease involving epistaxis, telangiectasia, visceral lesions (lung, liver, brain) and family history. Three criteria are necessary to confirm diagnosis (1). Approximately 90% of patients with HHT suffer from nose bleeding (2). Life and quality of life (QoL) are also modified by recurrent epistaxis and its associated treatment.

PQoL is a complex and subjective concept, which attempts to evaluate the well-being of individuals. PQoL has been incorporated and adapted to modern medicine to provide an analysis tool capable of quantifying health and illness. The term Health-Related Quality of Life (HRQoL) was born from this incorporation, its accurate definition being: *'the subjective individual perception of one's physical state (organic function), emotional state (mental and psychical state), and social state (capacity to create and maintain normal relationships with others), after having taken into account the effect of illness (symptoms) and of its treatment (sequelae, handicap) (3).'*

Our work concerned the quality of life of patients with HHT. We analysed the influence of epistaxis on relationships and psychosocial quality of life (PQoL) in HHT patients.

PATIENTS AND METHODS

Retrospective study

All living, adult HHT disease patients resident in France were considered eligible to enter the present study. All patients were able to write in French. Entry into the study began in September 2003 and ended in June 2005.

Questionnaire

The questionnaire was created by two sociologists and a Head and Neck surgeon. It was broad-based, multidimensional and reliable (questionnaire available upon e-mail request

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to the corresponding author). The 71 items in the study included information on psychological well-being, social life, family support, occupation, and medical and demographic data regarding age, gender, and patient appearance. Poor or good PQoL was determined by the response to the following question: 'according to you, your quality of life is…' PQoL, a subjective individual perception, was defined by the patients themselves.

Data

The questionnaire was sent by mail to 380 patients recruited from the Reference centre (Lyon, Prof. Plauchu), the competence centre and from AMRO, the HHT patients' association. They were auto questionnaires and remained anonymous. One hundred fifteen (30.2%) completed questionnaires were included.

Results analysis

Statistical analysis was carried out on the results of all 115 patients. Standard frequency analysis was performed for categorical variables such as gender, age and living situation. Significant correlation was assigned only for p values of less than 0.05. The software used was Microsoft Excel 2003® for input data and SAS 9.1® for statistical analysis.

RESULTS

Patient groups

Patients were predominantly female (71 women, 44 men). Mean age was 56 years while average age at HHT diagnosis was 36. Eighty percent were living as a couple. Forty percent of the patients of working age were active. There was no difference in geographic distribution between city and country. Seventy-five percent lived with a household income of between 750 and 3000 euros per month. Twenty-five percent were members of the association.

Eighty-five percent had epistaxis more than once a month and 25% more than once a day. The duration of the nasal haemorrhage was over five minutes in 65% of cases, more than one hour in 10% of cases. Two thirds of patients considered these episodes to be of moderate or great importance.

Relationships within the home were not modified by HHT in 70%, and with friends in 86% of the patients. Forty-five percent considered that their quality of life was good or very good. However, 58% of patients were frightened of speaking in public. Two thirds felt different to others. Twenty percent had never gone away for a weekend or on holiday.

PQoL analysis revealed no significant difference in relation to gender, marital status, household income or housing (rural or urban), but a significant difference was observed with employment. Workers had a better PQoL than unemployed patients (p = 0.027). Epistaxis was correlated to professional duties: workers with less than one episode of epistaxis per month were more active (p = 0.008). When the duration of the bleeding was less than one minute, patients were also Table more active (p = 0.026), whereas if it was more than one hour, patients were less active (p = 0.041).

Table 1. Criteria associated with the risk of poor QoL adjusted for age.

		Univariate*		
	Odds Ratio	CI 95%	p	p-trend
Bleeding frequency			0.04	0.02
Once a month	0.12	[0.02-0.76]		
Once a week	0.36	[0.12-1.05]		
Once a day	1			
Bleeding abundance			< 0.01	< 0.01
Low	15.14	[3.51-65.34]		
Medium	2.84	[0.72-11.19]		
High	1			

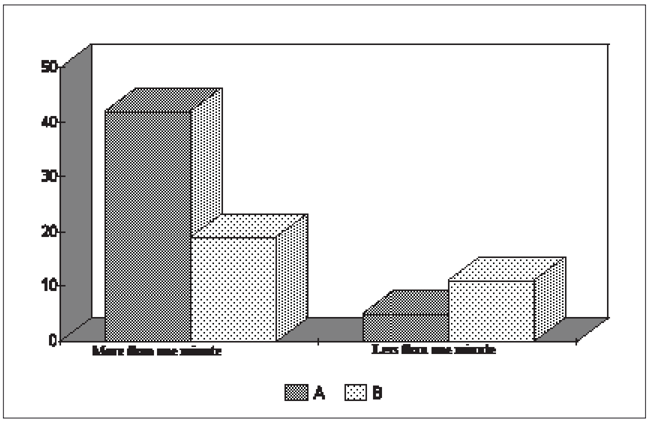
		Multivariate*		
	Odds Ratio	CI 95%	p	p-trend
Bleeding frequency and abundance			< 0.01	0.21
No bleeding	0.80	[0.14-4.69]		
Less than once a week and low abundance	1			
Less than once a week and medium or high abundance	3.99	[0.98-16.21]		
More than once a week	0.59	[0.09-4.07]		
Bleeding abundance and frequency			< 0.01	0.02
No bleeding	1.24	[0.23-6.71]		
Low abundance	1			
Medium or high abundance and less than once a week	6.34	[1.72-23.46]		
Medium or high abundance and more than once a week	2.079	[0.26-16.85]		

*: adjusted for age; CI: Confidence Interval.

Age also had an influence on PQoL. Elderly patients had a poorer PQoL than younger patients (p = 0.016). Nevertheless, it would appear that the expression of poor PQoL was not related solely to age, but rather significantly linked to the frequency and abundance of the episodes of epistaxis. Effectively, when adjusted for age (Table 1), improved PQoL was observed with low frequency and abundance. Furthermore, when adjusted for age, and with frequency and abundance criteria pooled together, the more frequent and abundant the epistaxis, the poorer the PQoL, independent of age (Table1). The analysis also revealed a significant link between PQoL and the duration, abundance and frequency of the episodes of epistaxis.

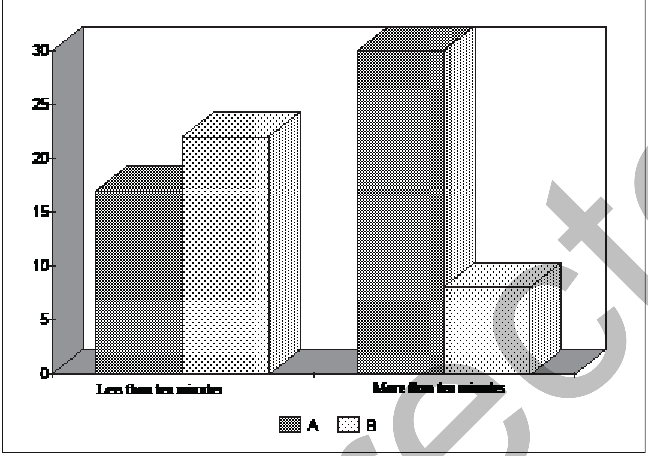
The duration of epistaxis changed the general life of HHT patients. Patients bleeding for less than one minute were satisfied with their life conditions (p = 0.006) (Figure 1), whilst those bleeding for more than ten minutes were dissatisfied (p = 0.043) (Figure 2). Patients bleeding for several hours were more afraid of dying (p = 0.001). Bleeding abundance decreased

Figure 1. Relationship between epistaxis duration (less or more than one minute) and quality of life in hereditary hemorrhagic telangiectasia patients.



Patients bleeding less than one minute are satisfied with their life conditions (p=0.006). A= poor quality of life; B= good quality of life.

Figure 2. Relationship between epistaxis duration (less or more than ten minutes) and quality of life in hereditary hemorrhagic telangiectasia patients.



Patients bleeding more than ten minutes are unsatisfied with their conditions of life (p=0.043). A = poor quality of life; B = good quality of life

quality of life. The frequency of dining at home with friends ($p = 0.012$) and that of eating in restaurants ($p = 0.015$) also decreased if patients felt that the abundance of their bleeding was significant. Finally, sexuality was also modified (lack of satisfaction) in patients with several episodes of epistaxis per day.

DISCUSSION

Did HHT patients' long-term PQoL decrease when measured with general instruments and compared with the general population? There is little literature available on PQoL and HHT. To find a few answers to this question, we searched for information using a Pub-Med search engine and two keywords: 'quality of life' and 'HHT.' Twenty articles have been published. Five used QoL scales. Extensive information can be extracted from these articles. Lennox demonstrated by means of the SF-36, a validated health status survey, that the score for each health dimension was significantly reduced ($p < 0.05$) when compared to the reference population for all dimensions except pain (4). Ninety per cent of patients with HHT considered their recurrent nosebleeds to be the most disturbing symptom, interfering with hobbies and leisure time in 63% (5).

No curative treatment exists for HHT. New studies are evaluating bleeding impairment and PQoL improvement. In a Greek study, Nd/Yag laser treatment seemed to improve Health-Related PQoL in the Physical and Mental Health Dimension in 27 patients two years after treatment (6). Septal dermoplasty reduced the need for transfusion and improved QoL (7). Hitchings, in a prospective study, analysed the QoL of patients after treatment with nasal closure, argon laser alone and argon laser in combination with septodermoplasty. A significant improvement in QoL was shown in the group of patients undergoing nasal closure, for whom bleeding ceased completely (8). Seventy-three percent of patients stated that they would appreciate new treatment methods for their nosebleeds (5). This proves that bleeding plays a major role in QoL impairment. Epistaxis is therefore the most important clinical variable (9).

Contrary to our results, in a study of 50 HHT patients, age was related to lower physical functioning, physical role limitations, bodily pain and emotional role limitations. Illness duration was negatively related to the mental component summary (9). Females had lower scores in several domains (9). Higher haemoglobin levels improved physical functioning (9). The number of episodes of epistaxis was negatively associated with physical role limitations, vitality, social functioning, physical component summary and bodily pain (9). For us, bleeding frequency did not decrease QoL except with regard to work and sexuality. Epistaxis abundance was related to impairment of many QoL criteria with relationship modifications.

In the literature, most authors were more interested in physical QoL than in its psychological and social aspects. No publications deal with the stigma associated with this disease as described by Goffman, a university of Chicago sociologist (10). The stigma, in our case telangiectasia and nasal bleeding, stems from an undesirable attribute. In our study, HHT patients were stigmatised. Altered body image results in dehumanisation and alters relationships with others. In a society that gives value to the body and beauty, the best means of communication requires face-to-face exchange. The PQoL of HHT patients is also decreased, and depression increased. Diagnosis of HHT generates an image of suffering. Independently of patients' individual circumstances, there are periods of 'before' and 'after' the diagnosis and initial treatment. Most patients are no longer able to work. Marital life is often modified with certain modifications in sexual life. Blood is frightening. In such conditions, telangiectasia and the threat of nasal bleeding are limiting factors in social relationships. Stigma transforms relationships, discrediting HHT patients and marginalising them in society.

We would like to point out a few limitations in this study. Firstly, it was not possible to calculate the participation rate. Secondly, we did not study a control group, although we adjusted our results for age. In addition, our questionnaire did not take into consideration any associated pathologies or the genetic pattern of HHT. Finally, we are aware that the PQoL benchmark is subjective, but it cannot be otherwise.

CONCLUSION

Psychosocial quality of life (PQoL) seems to be diminished in HHT patients.

It is not the stigma, but rather the episodes of epistaxis and their characteristics that alter PQoL. The frequency and abundance of the bleeding are negative prognosis factors. Physicians treating this disease need to consider their patients' quality of life and must endeavour to take all possible measures to help these patients remain full members of society with optimal functions, allowing improved social insertion and social rehabilitation.

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CONFLICT OF INTEREST

The authors have no relevant financial interests in this article.

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