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Life for patients with myelofibrosis: the physical, emotional and financial impact, collected using narrative medicine—Results from the Italian 'Back to Life' project

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Abstract

Purpose Myelofibrosis (MF) is a chronic myeloproliferative neoplasm characterised by an aggressive clinical course, with disabling symptoms and reduced survival. Patients experience a severely impaired quality of life and their families face the upheaval of daily routines and high disease-related financial costs. The aim of this study was to investigate the perceptions of Italian patients and their caregivers about living with MF and the burden of illness associated with MF.

Methods A quali-quantitative questionnaire and a prompted written narrative survey were administered to patients affected by primary or post-essential thrombocythemia/post-polycythaemia vera MF and their primary caregiver in 35 Italian haematological centres.

Results In total, 287 questionnaires were returned by patients and 98 by caregivers, with 215 and 62, respectively, including the narrative. At the time of diagnosis, the most commonly expressed emotional states of patients were fear, distress and anger, confirming the difficulty of this phase. A high level of emotional distress was also reported by caregivers. Along the pathway of care, the ability to cope with the disease differed according to the quality of care received. The mean cost to each patient attributable to MF was estimated as €12,466 per year, with an estimated average annual cost of loss of income of €7774 per patient and €4692 per caregiver.

Conclusions Better understanding of the personal life of MF patients and their families could improve the relationships between health workers and patients, resulting in better focused healthcare pathways and more effective financial support to maintain patients in their social roles.

Keywords Quality of life · Myelofibrosis · Narrative medicine · Burden of illness · Indirect costs

Introduction

Primary myelofibrosis (MF) is the most severe form of BCR-ABL1-negative chronic myeloproliferative neoplasm, characterised by the expansion of clonal haematopoietic cells, which release cytokines [1–3]. MF can also arise from other myeloproliferative neoplasms, including polycythaemia vera (post-PV MF) and essential thrombocythaemia (post-ET MF) [4]. Although the genetic trigger of MF is unknown [5, 6], the V617F mutation of the Janus kinase (*JAK*) 2 gene and mutations of the *MPL* and *CALR*

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genes have recently been discovered [7]. Primary MF is a rare disorder, with an estimated annual incidence in Europe of approximately 1 per 100,000 [8]. It primarily affects older individuals (median age at onset, 67 years old), when the individuals may be still active in working [9]. The only curative treatment, but available only to a minority of patients, is allogeneic haematopoietic stem cell transplantation [10, 11]. Other conventional treatments include cytoreductive therapies, erythropoiesis-stimulating agents, and steroids [10]. Recently, the JAK1/2 inhibitor ruxolitinib has demonstrated remarkable success in alleviating the MF symptomatic burden and overall survival [12, 13].

MF indeed not only shortens survival but also severely compromises overall quality of life (QoL) [14, 15]. According to the Myeloproliferative Neoplasm Symptom Assessment Form (MPN-SAF), a validated QoL instrument, the symptoms experienced by patients with MF are heterogeneous and include excessive and disabling fatigue, early satiety, anorexia, pruritus, bone pain, night sweats, cachexia, abdominal pain and discomfort, and cognitive complaints [16].

Despite the emphasis in recent international guidelines on the need to obtain knowledge about QoL to understand the everyday routine of people affected by a chronic disease [17], the existing scientific literature has focused on clinical signs and symptoms and only poorly on patients' and caregivers' experiences [17]. Narrative medicine (NM) is the best tool to investigate QoL, as stated recently by the World Health Organization [17–21]. NM can be defined as 'What is circumscribed between the physician and the patient, from the collection of information on events before the disease focusing on psychological, social and ontological implications' [20]. The NM approach deals with the 'illness' experience, defined according to Kleinman's theory [21, 22] and allows the possibility of understanding patients' and caregivers' needs, which cannot be captured by questionnaires or any other quantitative approach [23, 24]. Although the burden of illness (BOI) is known to be directly related to QoL [25], to date no studies have been performed in Italy to investigate the experience of living with MF that have included not only the medical burden of the disease (such as MF-related symptoms and treatment toxicities), but also financial issues (such as disease-related out-of-pocket costs) [25, 26].

The main aim of this survey was to improve understanding of the experience of MF in Italy to inform and direct the care of MF patients. This was achieved through an investigation of patients' and caregivers' perceptions of living with chronic illness, and their families' level of stress, with a particular focus on the BOI.

Methods

Participants' selection criteria

This cross-sectional and observational study included only patients specifically affected by primary MF or post-ET/post-PV MF (independent of the severity of the disease and the type of treatment) and their family informal caregiver. The survey used in this study was based on a mixed methods approach comprising a questionnaire that investigated the BOI and a narrative part designed as described in the following section. The study was conducted under the methodological supervision of ISTUD Foundation between September and November 2012 and between June and October 2013. The surveys were collected anonymously. The target set for the study was to collect surveys from at least 250 patients and 100 caregivers. Survey participation was on a voluntary basis, via an online platform specifically designed for this study, which was accessible only after providing written informed consent, in accordance with Italy's Privacy Law [27]. Participants could also learn of and gain information about the project from 35 haematological centres, situated in big cities across the entire Italian territory, which were chosen for their expertise in the management and treatment of patients with MF. Ethics approval for each data collection period was obtained from the principal centre involved. The entire study was conducted in accordance with the principles of the Declaration of Helsinki.

Burden of illness questionnaire

The BOI questionnaire evaluated six areas of illness impact: (1) socio-demographic information; (2) the beginning of the care pathway, especially the diagnosis and the patient's clinical status at diagnosis; (3) the patient's life along the care pathway, including clinical status after treatments; (4) life with MF from the caregiver's perspective, such as the caregiving assistance provided; and (5) financial implications, including on working productivity (before and after the diagnosis) and out-of-pocket costs. Informal caregivers received the same questionnaire as the patients, but they were additionally given the American Medical Association's Caregiver Self-Assessment Questionnaire [28] to measure the level of stress associated with caring for a patient with a chronic disease; this was translated literally from English to Italian. All other parts of the survey were specifically designed for this study by the authors, in Italian.

Studying the illness through narrative medicine

The narrative part of the survey was structured according to Greenhalgh's methodology [18], which identifies three main phases: falling ill, being ill and getting better/getting worse. The narrative follows a determined plot that begins before diagnosis and continues through the clinical encounter, investigations, development of a management plan, treatment and resolution or progression of the disease. In this study, the plot of each narrative was structured using little prompts, written in Italian (see Online Appendix). The texts were also analysed through an assessment of recurring words and semantic expressions using Nvivo10. This software is based on Grounded Theory [29] and facilitates rigorous textual analysis by generating substantive codes through word clustering, calculating word frequencies and evaluating logical correlations, creating graphs and charts [30].

According to Launer and Robinson's classification of narratives [31], 'progressive' narratives show the individual's ability to activate 'coping', even in a difficult and uncertain situation. Here, 'coping' is defined as the conscious effort to solve personal and interpersonal problems, and to master, minimise or tolerate stress or conflict. Conversely, 'stable' and 'regressive' narratives relate to when the individual did not find a way to manage the situation effectively and deactivated 'coping' [32–35]. Because narratives are not metrics, the validity of the NM analysis phase comes through reading the narratives independently and in a blinded fashion, following the Grounded Theory protocol [36]. This analysis was performed by three authors of this paper (AC, LR, MGM), chosen for their high expertise in qualitative research and the NM approach [37–40] as applied in the field of haematology [41–43].

Statistics

All data collected from the surveys, even incomplete ones, were analysed using standard descriptive statistics as means, ranges and frequencies. No further inferential analysis was performed with any sample size calculation because of the narrative nature of the study. The analysis of specific therapies was not an objective of the present study, so the information regarding treatments was analysed and reported in an aggregated form.

Estimation of caregiving costs

Direct and indirect costs are reported as mean amounts across all participants based on what the participants perceived as having lost because of MF and declared in the questionnaires and/or in narratives. The assistance of the informal caregiver, as perceived by both patients and families, was examined, taking into consideration the tasks carried out and the time spent on such tasks and activities. This time-based information was converted to a financial cost through the attribution of a specific monetary value based on the cost of that number of hours of assistance provided via the Italian National Labour Contract for carers in Italy. This contract establishes the cost per hour as €4.41 (US$5.21) for Level A work (equivalent to that of a generic carer with basic training, cleaner, laundry operator or housework aid), or €7.83 (US$9.26) for Level D work (equivalent to an assistant to patients who are not self-sufficient, or someone holding a professional degree or a specific certificate, such as a general registered nurse or geriatric assistant, who also performs tasks related to feeding and house cleaning). When a carer was involved for over 5 h a day, the monthly cost was considered to be €606.79 (US$717.48) for level A work and €1158.42 (US$1369.74) for level D.

Results

Socio-demographic information

Overall, 287 questionnaires were received from patients with MF and 98 from their caregivers; of these, narrative was included with the responses from 215 patients and 62 caregivers. The basic socio-demographics of participants (both patients and caregivers) are summarised in Table 1 and are representative of the general Italian population [42].

The beginning of the pathway: diagnosis and clinical status

Quantitative analysis showed that the diagnosis was made an average of 2.5 years after the onset of symptoms, in 21% of cases, the period was more than 3 years. Systemic symptoms (insomnia, bone pain, itching, loss of appetite, stress and muscle ache) occurred in 63% of patients, and constitutional symptoms (sweating, weight loss and fever) were reported by 49%. The participants were asked to evaluate their general QoL before starting treatment; this was reported as excellent/good in 40%, tolerable in 31% and poor/very poor or bad in 29%. The most frequently reported symptom at the time of diagnosis was fatigue (70%), and the narratives confirmed this adversely conditioned the participants' daily activities and forced them to spend several hours in bed during the day.

The narratives confirmed these quantitative results but added further details. In particular, in 40% of cases, the onset of MF occurred at an unexpected moment, in an everyday context, and was perceived as an imbalance in body integrity. In a further 19% of cases, patients realised they had the disease during a course of treatment for a different

Table 1 MF patients and their caregivers' socio-demographic information

	Patients		Caregivers	
	Questionnaires	Narratives	Questionnaires	Narratives
	N. 287 (%)	N. 215	N. 98 (%)	N. 62 (%)
Male	54	54	42	40
Female	46	46	56	60
Age				
< 51	12	14	40	34
51–60	13	13	22	27
61–70	38	39	22	21
> 70	37	34	16	17
Marital status				
Single	7	8	15	11
Married	73	72	80	82
Divorced	7	8	4	5
Widow	13	12	1	2
Educational qualification				
Primary school degree	25	21	10	11
Middle school degree	28	29	29	31
High school degree	34	36	41	44
Bachelor/Master degree	13	14	20	13
MF classification				
PMF	44	46	–	–
Post-ET MF	21	23	–	–
Post-PV MF	19	19	–	–
ND	16	13		

health condition. The remaining 41% of patients made a specific connection with their past perceived symptoms just at the time of the diagnosis communication; among these, 10% reported feeling in good health. In addition, during the period between the onset of symptoms and the MF diagnosis, 16% of patients asked for three or more consultancies before being told they had MF; in these cases, a strong sense of discomfort in the pre-diagnosis period emerged from the narratives, probably because of this long period of uncertainty. The most critical moment reported in the doctor–patient relationship was the communication of the diagnosis, particularly when the haematologist explained that MF is a chronic cancer with no standard therapeutic management. Indeed, the most frequently remembered words by patients in the first description of this disease were 'chronic disease', 'rare pathology to be monitored' and 'curable leukaemia', whereas the most common word remembered by patients with secondary MF was 'evolve', as their PV or ET transformed to MF. Medical language, although described as precise and accurate, was perceived as too technical and subject to misinterpretation. In most cases, the patients reported that the setting and the time dedicated to explaining and commenting on the diagnosis were inadequate and did not promote a good understanding or a satisfactory emotional acceptance of the disease. The narrative analysis showed that the most frequent emotions felt by patients at the time of diagnosis communication were fear (53%) and distress (23%), followed by relief (11%), resignation (7%) and anger (6%). Furthermore, the narratives highlighted that the patients felt they were initially ignorant and uninformed: 'I felt lost because I didn't know of the disease and or how it would evolve'; 'I felt worried because I hadn't heard anyone talking about this disease before'; 'I didn't know anything about the disease but the conversation with Doctor P. got me worried'. Following the initial time of diagnosis, 47% of patients continued to express resignation and despair; these people felt helpless and, in some cases, they decided not to react, simply waiting for death. Conversely, 27% of narratives showed the patients' inner strength in deciding to react and find a way to live with the disease. In addition, 6% of patients tried to be strong for their family and 18% thought pragmatically about the difficulties they would have to face in their ordinary life (see Table 2).

Life for the MF patients along the care pathway

The quantitative part of the survey revealed that patients experienced difficulties in movement caused by

Table 2 MF patients' reactions after the communication of the diagnosis

I thought…	Patients N. 152 (%)
It would be the end	47
To face the disease	27
To my family	6
How my life would be affected by that	18

Table 3 Improvements of symptoms after treatment

	% of patients
Splenomegaly	43
Fatigue	85
Constitutional symptoms	61
Systemic symptoms	58
None	29

Table 4 Restored activities thanks to treatment

	% of patients
None	32
Essential activities	56
Domestic activities	53
Leisure activities	54

splenomegaly (68%) and the narratives showed that this forced the patients to limit their mobility and had a strong impact on their QoL, even in the absence of abdominal pain, with 49% of participants reporting limitation of outdoor activities, 23% of domestic activities and 6% of daily self-care activities.

In the narratives, 52% of patients reported that, before beginning therapy, they had been unable to perform their normal daily activities. The activities suffering the greatest impact were walking and travelling (49%), and domestic activities such as cooking and housekeeping (23%). In 6% of cases, MF impaired essential daily activities such as dressing, washing and heating. Therapies appeared to improve the patients' moods and reduce fear (54%) and depression (36%). According to the narratives, the patients' initial feelings of fear and pain gradually diminished over time with health improvements, a better knowledge of the disease and familiarity with the medical centre. Indeed, all the narratives described therapy as playing a pivotal role in the patients' experience of illness, with most describing it as useful and effective.

Most of the patients reported improvements of symptoms associated with the disease as a consequence of the therapy

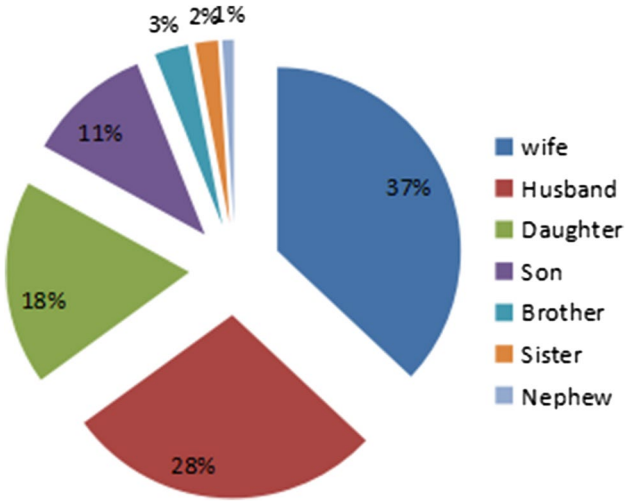

Fig. 1 The relationships between the caregivers and patients

(Table 3). In addition, 48% indicated improvement in their QoL and 52% were able to restart their main daily activities previously interrupted because of MF (Table 4). The narratives also showed that although 60% of patients were able to maintain their working activities, 20% of them experienced a reduction in their productivity and 20% had been forced to interrupt their professional life. In addition, the housekeeping activity is demanded to a maid in 10% of cases, even after treatment.

An investigation of the patients' relationships with others showed that only 35% of patients reported that, at the beginning of the disease, they were able to live in a harmonious relationship with their relatives and speak openly about their condition. Others reported that their relationships had been negatively influenced by emotions of fear, anger and grief. In some cases, problems of isolation were particularly evident. Indeed, the patients often described the people they were closest to as worried but unable to provide a point of reference and support. According to the narratives, the family situation seemed to improve after the haematology centre had taken charge of the patient, and the proportion of patients then living in a harmonious relationship with their family increased to 60%.

Finally, at the end of the narratives, 70% of the patients were facing the future full of hope and courage; however, almost half of the patients expressed hope for a complete recovery, not accepting MF as a chronic disease.

Life with MF: the caregiver's perspective

In all cases, the primary caregiver was a family member: 65% of those participating were spouses and the remaining 35% were other close family members (Fig. 1). No patients' parents were among the caregivers who participated, perhaps

because patients with this disease tend to be elderly [9] and rarely have relatives older than themselves as caregivers in Italy.

According to the questionnaire data, the duration of caregiving was > 4 years in 52% of cases, 2–4 years in 20% and 1–2 years in 14%. In 55% of cases, the caregivers were assisting patients otherwise in good general health and able to preserve their autonomy.

The Caregiver Self-Assessment Questionnaire revealed a high level of stress in 87% of caregivers. This was reflected in 40% of the narratives, in which caregivers reported their own distress because of the patients' fear (23%), anger (19%) and distress (10%): 'I feel misunderstood and abandoned'; 'I see my husband suffering and I feel powerless and unprepared for assisting him'; 'I often cry'; 'I tend to stay isolated to avoid being a burden'.

As with the patients, the narratives indicated that the grimmest time for caregivers was when the diagnosis was first communicated, with the most frequent emotions being fear (28%), discomfort (28%) and bewilderment (18%). Despite these negative feelings, 45% of caregivers reported an immediate desire to find the strength to react and stay close to their loved one, and 23% decided to look for the best treatment. In 70% of cases, caregivers reported that the disease had an overall negative effect on the patients, but 44% nevertheless decided from the beginning to remain close to the patient. All caregivers felt they had been taken into consideration by the doctors.

With regard to the future, fear, anxiety and sadness emerged in 37% of the caregivers' narratives, whereas positive feelings of confidence, serenity and optimism were reported in 27%. In the remaining narratives, the predominant feeling was the hope of a 'miracle' of healing, wishing for divine help or the discovery of a new treatment, or the hope of living peacefully and making best use of every possible moment with the patient.

Coping strategies in the patients' and caregivers' narratives

The patients' narratives were classified as 'progressive' in 48% of cases. The factors that allowed the positive development of their stories were, primarily, the recovery or maintenance of daily activities and the success of the therapy. Other key elements of coping were the opportunity to share and debate with others and being able to obtain support. In particular, family members provided the support needed, helping the patients to understand that a satisfactory life was still possible. Furthermore, the trusted relationship with physicians allowed patients to understand the course of the pathology, improving their awareness and making them feel reassured and cared for. As a result, patients trusted the treatment, considering it the best possible course of action. Conversely, patients whose narratives were classified as 'stable'

Table 5 The Launer and Robinson's classification of narratives based on coping strategies

	Patients N. 215 (%)	Caregivers N. 62 (%)
Progressive narratives	48	53
Stable narratives	32	30
Not classified narratives	20	17

Table 6 The relationship between the evolution of the patients' narratives and their considerations on physicians

Considerations on physicians	Progressive narratives N. 93 (%)	Stable narratives N. 61 (%)
Great technician	48	71
Great empathy	49	23
Not adequate	3	6

described their physician as professionally and technically prepared, but less empathic and helpful; only in 22% of cases they were satisfied with them (Tables 5, 6).

Of the caregivers' narratives, 51% were classified as 'progressive' because of the caregiver's strong and motivated dedication to the patient acting as the positive stimulus to make the narrative 'evolving'. Two categories of feelings were detected: the first one was a 'moralistic' sense of duty towards the family and a willingness to continue, struggle and fight together, regardless of the outcome of treatment. The second category was related to the affection sphere, where 'love' was one of the most frequently used words. Love enabled the caregivers to endure the constant mood swings and the difficult moments in the relationships.

The type of evolution of the narratives was found to correlate with cost issues. The patients who wrote 'progressive' narratives were those also reporting lower indirect costs, up to 18% (€1365) lower than the average annual loss across all the patients.

Financial impact of MF for patients and caregivers

Although not all participants answered to the part of the survey focused on financial issues, probably because of the Italian Healthcare System that mostly pays for many health services, the extended concept of BOI was substantial. It included also a broad array of intangible non-medical costs (e.g. domestic care assistance) and indirect costs (e.g. loss of productivity) for both patients and caregivers [25, 26].

Direct costs to access the Healthcare System resources (transport costs, examinations and tests) were reported by 12% of patients (as an average of €137/month). Overall, 91% of patients attended a haematology centre close to

their homes, in the region where they lived. The average travel time to the hospital was 55 min (range 3–720 min). The majority (87%) of participants usually travelled to the hospital by car, whereas 7% used train and 3% the bus. The average transport cost corresponded to €42 per visit.

At the time of the interview, 86 of 144 patients (60%) had not stopped working nor reduced their work activity since the MF diagnosis, whereas 63 patients (22%) reported an annual earning loss of €7774. Overall, 63% of patients needed the daily assistance of an informal caregiver. Requested activities were household duties (49%) and tasks outside the home (51%). In addition, the caregivers provided social support, which mostly consisted of not leaving the patient alone. Overall, 41% of caregivers spent more than 6 h/day on this activity. The estimated monthly average cost for the time spent on informal caregiving was €655. In addition, caregivers reported a reduction in their annual earnings after the MF diagnosis, with an average annual loss of earnings of €4692 (Table 7).

Discussion

The integration of the NM approach with a quantitative questionnaire revealed, for the first time, a more complete view of MF patients' and their caregivers' experiences. Indeed, their direct testimonies represent the most accurate way to follow their journey into the 'falling ill' phase, describing the inner and more intimate world of the meaning of the disease to these individuals. This is particularly relevant in the setting of chronic neoplasms such as MF that are projected to have a relatively long survival but may be burdened by disabling symptoms and reduced QoL [14, 15].

This study mainly assesses in detail the strong negative perception of MF patients of their condition. According to narratives, the main feelings that affected patients at the beginning were discomfort, fear and distress, which might persist over time. A strong perception of social isolation and loss of autonomy in terms of not being able to do hobbies or any intense physical activity (e.g. housekeeping, heavy lifting…), as well as essential activities (e.g. walking, dressing…) emerged from the patients' narratives.

A further message of this analysis related to the crucial role played by caregivers in the care pathway of MF patients. Their heavy burden of daily tasks resulted in a high level of stress; this could be reduced through greater consideration of their needs and points of view, both in the organisation of support services and in the relationship of care.

Although the feedback from some participants confirmed that their direct health costs were mostly covered by the Italian National Health Service, most of cases reported MF as a burden with high intangible and indirect costs because of the progressive impairment of QoL. More than 50% of patients were still employed at the time of diagnosis of MF. Consequently, the limitations to their daily activities led to a loss of income for those who were still at a working age. The perceived BOI of MF was calculated to be as high as €12,400 per year due to the loss of family income; if the informal caregiver had been replaced by a professional career, the family income loss would have risen to €20,326. These data agree with those from a Spanish study on MF [44], which quantified the total burden of indirect and non-medical costs as €15,142 a year, as well as with a study on cancer in general [45, 46], which confirmed a substantial financial impact of the disease on patients' families.

The AMA Questionnaire used to investigate caregivers level of stress was literally translated into Italian from American English, without formal back-translation into English or pre-testing for equivalence. Further, it has only been partially validated for depression [28], and the dichotomous response options, while simple, can be misleading and harder to use analytically. These may be possible sources of bias. To overcome some of these limitations and to complement the quantitative findings, qualitative work was undertaken. Further,

Table 7 Burden of illness (BOI)

Annual earning loss due to non-medical costs		
Loss of productivity of patients	7774	10.537 US$
Loss of productivity of caregivers	4692	6359.50
		10.653 US$
Annual earning loss due to informal caregiving costs		
In terms of housekeeping activities and assistance to patients	7860	
	Total 20.326 €	27.550 US$
		2.228 US$
Annual earning loss due to direct costs	1644	
Of whom transport costs	− 555.66	(753 US$)
Total	21,970	23.418 US$

The value of the monetary exchange rate for October 2013 was used

although Italian and American have different cultures, the AMA questionnaire is composed of simple questions about everyday activities of caregivers (for example, sleeping, recurrent thoughts…) that are not expected to be affected by different cultures and contexts. The BOI questionnaire, although project specific, was designed by the authors to meet the study objectives.

Differing perceptions between patients and physicians of the BOI and poor patient–physician communication may delay or prevent the achievement of treatment goals [47]; however, the present preliminary study did not involve the professionals' point of view. Nevertheless, healthcare professionals and managers can benefit from the new metric offered by the integration of the NM approach with BOI tools in creating solutions for the problems and needs experienced by patients and their families found in this study. In particular, findings of this study suggest the following conclusions of relevance to health professionals. First, the relationships of care have an impact on patients' and caregivers' illness experience. The narratives provided evidence of the key role of the relationship between the haematologist and both patient and caregiver in enabling them to cope with MF. A successful relationship was based not only on technical expertise, but also on emotional empathy and communication skills in accordance with the patients' wills, needs and possibilities. Second, medical teams should not underestimate the stress and loneliness felt by caregivers, who form part of the care, as reported in previous studies [48]. Finally, when the patient is able to cope, there is a positive impact not only on QoL but also on financial costs, because the positive attitude allows improved therapy adherence and greater effort to be applied to working and daily activities. In conclusion, the findings of this study showed that an understanding of MF patients' and their families' resources was essential for improving care relationships, better organisation of the care pathway and guiding economic choices to support the patients, allowing them to maintain their productivity and social role.

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Compliance with ethical standards

Conflict of interest A. Iurlo has acted a consultant for and received honoraria from Novartis; M. Pellegrino has received honoraria from Novartis, as producer of deferasirox and ruxolitinib that are agents used for the treatment of myelofibrosis; M.Breccia has received honoraria from Novartis, BMS, Pfizer and Incyte; M. Bonifacio has received research fundings from Novartis; all other authors declared no conflict of interests.

Informed consent Informed consent was obtained from all individual participants included in the study.

References

1. Smith, R. E., Chelmowski, M. K., & Szabo, E. J. (1990). Myelofibrosis: A review of clinical and pathologic features and treatment. *Critical Reviews in Oncology/Hematology, 10*, 305.
2. Campbell, P. J., & Green, A. R. (2006). The myeloproliferative disorders. *The New England Journal of Medicine, 355*, 2452.
3. Lataillade, J. J., Pierre-Louis, O., Hasselbalch, H. C., et al. (2008). Does primary myelofibrosis involve a defective stem cell niche? From concept to evidence. *Blood, 112*, 3026.
4. Mesa, R. A., Verstovsek, S., & Cervantes, F. (2007). Primary myelofibrosis (PMF), post polycythemia vera myelofibrosis (post-PV MF), post essential thrombocythemia myelofibrosis (post-ET MF), blast phase PMF (PMF-BP): Consensus on terminology by the international working group for myelofibrosis research and treatment (IWG-MRT). *Leukemia Research, 31*(6), 737–740.
5. Ciurea, S. O., Merchant, D., Mahmud, N., et al. (2007). Pivotal contributions of megakaryocytes to the biology of idiopathic myelofibrosis. *Blood, 110*, 986.
6. Tefferi, A. (2005). Pathogenesis of myelofibrosis with myeloid metaplasia. *Journal of Clinical Oncology, 23*, 8520.
7. Cervantes, F. (2014). How I treat myelofibrosis. *Blood, 124*(17), 2635–2642.
8. Moulard, O., Mehta, J., Fryzek, J., et al. (2014) Epidemiology of myelofibrosis, essential thrombocythemia, and polycythemia vera in the European Union. *European Journal of Hematology, 92*(4), 289–297.
9. Komrokji, R. S., Verstovsek, S., Padron, E., et al. (2012) Advances in the management of myelofibrosis. *Cancer Control. 19*(4 Suppl), 4–15.
10. Mesa, R. A. (2010). Assessing new therapies and their overall impact in myelofibrosis. *Hematology Am Soc Hematol Educ Program, 2010*, 115–121.
11. Gupta, V., Malone, A. K., Hari, P. N., et al. (2014). Reduced-intensity hematopoietic cell transplantation for patients with primary myelofibrosis: a cohort analysis from the center for international blood and marrow transplant research. *Biology of Blood Marrow Transplant, 20*(1), 89–97.
12. Scherber, R. M., Geyer, H. L., & Mesa, R. A. (2014). Quality of life in MPN comes of age as a therapeutic target. *Current Hematologic Malignancy Reports, 9*(4), 324–330.
13. Verstovsek, S., Kantarjian, H., Mesa, R. A., et al. (2010). Safety and efficacy of INCB018424, a JAK1 and JAK2 inhibitor, in myelofibrosis. *The New England Journal of Medicine, 363*(12), 1117–1127.
14. Steensma, D. P., Heptinstall, K. V., Johnson, V. M., et al. (2008) Common troublesome symptoms and their impact on quality of life in patients with myelodysplastic syndromes (MDS): Results of a large internet-based survey. *Leukemia Research, 32*(5), 691–698.

15. Johansson, P., Mesa, R., Scherber, R., et al. (2012). Association between quality of life and clinical parameters in patients with myeloproliferative neoplasms. *Leukemia & Lymphoma, 53*(3), 441–444.
16. Scherber, R., Dueck, A. C., Johansson, P., Barbui, T., Barosi, G., Vannucchi, A. M., et al. (2011) The Myeloproliferative Neoplasm Symptom Assessment Form (MPN-SAF): International prospective validation and reliability trial in 402 patients. *Blood, 118*(2), 401–408. https://doi.org/10.1182/blood-2011-01-328955. (**Epub 2011 May 2**).
17. Greenhalgh, T. (2016) Cultural contexts of health: the use of narrative research in the health sector. Copenhagen: WHO Regional Office for Europe; (Health Evidence Network (HEN) synthesis report 49). http://www.euro.who.int/__data/assets/pdf_file/0004/317623/HEN-synthesisreport-49.pdf. Accessed on 16 Jan 2017
18. Hurwitz, B., Greenhalgh, T., & Skultans, V. *Narrative research in health and illness*. London: British Medical Association Ed (2004).
19. Greenhalgh, T., & Hurwitz, B. (1998). Why study narrative? In T. Greenhalgh & B. Hurwitz (Eds.), *Narrative based medicine: dialogue and discourse in clinical practice* (pp. 3–16). London: British Medical Journal Books.
20. Charon, R. (2012). At the membranes of care: narratives in narrative medicine. *Academic Medicine, 87*(3), 342–347.
21. DasGupta, S., & Charon, R. (2004). Personal illness narratives: Using reflective writing to teach empathy. *Academic Medicine, 79*, 4.
22. Kleinman, A. (1988). *The illness narratives: Suffering, healing, and the human condition*. New York: Basic books.
23. Marini, M. G., & Arreghini, L. (2012). *Narrative medicine for a sustainable healthcare system*. Milano: Lupetti.
24. Marini, M. G. (2016). *Narrative medicine: Bridging the gap between evidence-based care and medical humanities*. London: Springer International Publishing.
25. Tarricone, R. (2006). Cost-of-illness analysis. *What Room in Health Economics? Health Policy, 77*, 51–63.
26. Pieri, L., Loiacono, I., & Vannucchi, A. M. (2013). The burden of symptoms in myelofibrosis: From patient-reported outcomes to health economics. *Leukemia Research, 37*(8), 855–856.
27. Personal data code protection. (2003). Law 30/06/2003 n.196. Published on the Italian Official Journal n.174., 29th July 2003, Supplementary n.123.
28. Epstein-Lubow, G., Gaudiano, B. A., Hinckley, M., et al. (2010). Evidence for the validity of the American Medical Association's Caregiver Self-Assessment Questionnaire as a screening measure for depression. *Journal of American Geriatric Society, 58*(2), 387–388.
29. Corbin, J., & Strauss, A. (2015) *Basics of qualitative research. Techniques and procedures for developing Grounded Theory*. Los Angeles: SAGE Publications.
30. Bazeley, P., & Jackson, K. (2013) *Qualitative Data Analysis with NVivo* (pp. 68–73). London: Sage.
31. Launer, J. (2006). New stories for old: Narrative-based primary care in Great Britain. *Families, Systems and Health, 24*(3), 336–344.
32. Weiten, W., & Lloyd, M. A. (2008) *Psychology applied to modern life*. (9th edn.). Boston, MA: Wadsworth Cengage Learning
33. Carver, C. S., et al. (1989). Assessing coping strategies: A theoretically based approach. *Journal of Personality and Social Psychology, 56*(N.2), 267–283.
34. Heather, N., Rasmussen, C., Wrosch, M. F., Scheier, & Carver, C. S. (2006). Self-regulation processes and health: The importance of optimism and goal adjustment. *Journal of Personality, 74*(6), 1721–1748.
35. Carver, C. S., & Connor-Smith, J. (2010). Personality and coping. *Annual Review of Psychology, 61*, 679–704.
36. Kennedy, T. J., & Lingard, L. A. (2006). Making sense of grounded theory in medical education. *Medical Education, 40*, 101–108.
37. Marini, M. G., Chesi, P., Bruscagnin, M., Ceccatelli, M., & Ruzzon, E. (2017). Digits and narratives of the experience of Italian families facing premature births. *Journal of Maternal Fetal & Neonatal Medicine*. https://doi.org/10.1080/14767058.2017.1339272.
38. Marini, M. G., Chesi, P., Mazzanti, L., Guazzarotti, L., Toni, T. D., Salerno, M. C., et al. (2016) Stories of experiences of care for growth hormone deficiency: The CRESCERE project. *Future Science OA, 2*(1), FSO82. https://doi.org/10.4155/fso.15.82. (**eCollection 2016 Mar**).
39. Cappuccio, A., Limonta, T., Parodi, A., Cristaudo, A., Bugliaro, F., Cannavò, S. P., et al. (2017) Living with chronic spontaneous Urticaria in Italy: A narrative medicine project to improve the pathway of patient care. *Acta Dermato-Venereologica, 97*(1):81–85. https://doi.org/10.2340/00015555-2478.
40. Marini, M. G., Reale, L., Cappuccio, A., Spizzichino, M., Zini, P., Amato, F., et al. (2014). Narrative medicine to highlight values of Italian pain therapists in a changing healthcare system. *Pain Management, 4*(5), 351–362. https://doi.org/10.2217/pmt.14.35.
41. Research, & Projects. http://www.narrativemedicine.eu/projects. Accessed on 28 Nov 2017.
42. Istat. (2012). Annuario statistico italiano.
43. Gimenez, E., Besses, C., Boque, C., et al. (2014). Indirect and non-medical economic burden, quality-of-life, and disabilities of the myelofibrosis disease in Spain. *Journal of Medical Economics, 17*(6), 435–441.
44. Mesa, R., Miller, C. B., Thyne, M., Mangan, J., Goldberger, S., et al. (2014). Gaps in perception between patients and physicians regarding symptomatology and treatment attitudes for myeloproliferative neoplasms: MPN LANDMARK SURVEY. *Blood*, *24*, 4827.
45. Robin Yabroff, K., Lawrence, W. F., Clauser, S., Davis, W. W., & Brown, M. L. (2004). Burden of illness in cancer survivors: Findings from a population-based national sample. *Journal of the National Cancer Institute*, *96*(17), 1322–1330.
46. Ayres, L. (2000) Narratives of family caregiving: Four story types. *Research in Nursing and Health, 23*, 359–371.
47. Levit Laura, A., et al. (Eds.). (2013). Patient-centered communication and shared decision making. In *Delivering high-quality cancer care: Charting a new course for a system in crisis*. Washington, DC: National Academies Press.
48. Sklenarova, H., et al. (2015). When do we need to care about the caregiver? Supportive care needs, anxiety, and depression among informal caregivers of patients with cancer and cancer survivors. *Cancer*, *121*(9), 1513–1519.

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