

Alma Mater Studiorum Università di Bologna Archivio istituzionale della ricerca

Comprehensive care for haemophilia: A literature review for improving institutional cooperation

This is the final peer-reviewed author's accepted manuscript (postprint) of the following publication:

Published Version: Comprehensive care for haemophilia: A literature review for improving institutional cooperation / Brondi, Sonia; Palareti, Laura; Mazzetti, Greta. - In: INTERNATIONAL JOURNAL OF HEALTHCARE MANAGEMENT. -ISSN 2047-9700. - ELETTRONICO. - 14:2(2021), pp. 402-411. [10.1080/20479700.2019.1652415]

Availability: This version is available at: https://hdl.handle.net/11585/734736 since: 2021-02-27

Published:

DOI: http://doi.org/10.1080/20479700.2019.1652415

Terms of use:

Some rights reserved. The terms and conditions for the reuse of this version of the manuscript are specified in the publishing policy. For all terms of use and more information see the publisher's website.

This item was downloaded from IRIS Università di Bologna (https://cris.unibo.it/). When citing, please refer to the published version.

(Article begins on next page)

This is the final peer-reviewed accepted manuscript of:

Sonia **Brondi**, Laura Palareti & Greta Mazzetti (2021) *Comprehensive care for haemophilia: A literature review for improving institutional cooperation,* International Journal of Healthcare Management, 14:2, 402-411

The final published version is available online at: <u>10.1080/20479700.2019.1652415</u>

Rights / License:

The terms and conditions for the reuse of this version of the manuscript are specified in the publishing policy. For all terms of use and more information see the publisher's website.

This item was downloaded from IRIS Università di Bologna (<u>https://cris.unibo.it/</u>)

When citing, please refer to the published version.

Comprehensive care for haemophilia: A literature review for improving institutional cooperation

Sonia Brondi, Laura Palareti, Greta Mazzetti

Department of Education Studies "Giovanni Maria Bertin", University of Bologna, Italy

Corresponding author

Sonia Brondi, Department of Education Studies "Giovanni Maria Bertin", University of Bologna, Via Filippo Re 6, 40126 Bologna, Italy. Tel.: +39 051 2091620; fax: +39 051 2086181; e-mail: sonia.brondi@unibo.it

Abstract

This paper investigates how the notion of comprehensive care (CC) in haemophilia is proposed in institutional guidelines and specialised understandings. A literature review of the articles published in *Haemophilia* was carried. The main features which authors refer to when mentioning CC were outlined and compared with the principles, assumptions and recommendations set by the World Federation of Haemophilia in relation to this care model. The results show a fragmented scenario. First, very few articles are devoted or explicitly refer to CC. Second, only a few of them completely adhere to the notion of CC as proposed in institutional guidelines. In contrast, most articles stress a single feature, providing a partial view of the model. The focus is mainly on promoting physical health, which is however frequently associated with psychosocial health. Multidisciplinarity has great emphasis, too. Third, some key issues for CC emerge. They are implicitly present in institutional guidelines, but they become the very focus of some articles, which reflect on three challenges to be dealt

with: care for all, transition care and systemic care. Such challenges offer the opportunity to clarify and expand the notion of haemophilia CC, and to propose potential future developments in research designs and interventions.

Keywords

Chronic illness; bleeding disorder; integrated care; psychosocial issue; multidisciplinarity

Introduction

State of the art

To date, knowledge on haemophilia, as well as bleeding disorders at large, among nonspecialist healthcare professionals (HCPs) is quite limited since it is a rare disease. As a consequence, both haemophilia diagnosis and management in emergency situations are complex when conducted outside specific hospital departments.

In recent years, the greater number of therapeutic options and medical devices developed by the pharmaceutical industry and relying on biomedical research and multidisciplinary theoretical approaches have reduced mortality and increased the quality of life of people with haemophilia (PWHs). There are, however, considerable variations between countries due to social, economic, political and cultural differences that affect the amount, availability and type of treatments proposed, as well as the use of a more or less integrated approach to the definition of health and care.

Notion of comprehensive care in institutional guidelines

Today, the care model promoted by the World Federation of Haemophilia (WFH), and set by the guidelines recently revised after their first edition in 2005 [1], is known as 'comprehensive care' (CC) (Figure 1). One of its principles is that PWHs are best managed in

a CC setting. In particular, the model is based on two main assumptions. First, it stresses the significance of jointly promoting physical and psychosocial health, while improving the quality of life of PWHs and their families, and decreasing morbidity and mortality. Second, CC emphasises the necessity of co-ordinately combining different HCPs into multidisciplinary teams that are responsive to the wide-ranging needs of PWHs and their families. CC teams should be able to protect the physical and psychosocial health of PWHs and their families, thus they should include medical directors, nurse coordinators, musculoskeletal experts, laboratory specialists, psychosocial experts, and should be the point person in order to approach different non-specialised HCPs (e.g., dentists, geneticists, gynaecologists).

[Figure 1 near here]

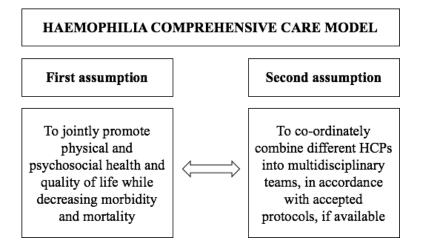


Figure 1. Diagram of the comprehensive care model (adapted from [1]).

These two assumptions translate into concrete actions as well. A CC program should provide continuous, long-term and multidisciplinary support to PWHs and their families [e.g., 2]. This involves the identification of resources and strategies to help to cope with everyday risks and problems, specificities of different life stages, school and/or employment issues, and reproduction options, in accordance with specific national treatment guidelines. In other

words, this implies ensuring prompt management of bleeding events, prevention of (potential) related complications and attention to psychosocial health at the haemophilia comprehensive care centres (HCCCs) [e.g., 3,4]. Moreover, a CC program should guarantee full training and close supervision of home treatments among PWHs, as well as adequate tutoring for their families to meet PWHs' needs in everyday life [e.g., 5,6].

Rationale of the paper

The aim of this paper is to examine the mutual relationship between the notions of haemophilia CC proposed by institutional guidelines and specialised understandings. More precisely, this paper intends to investigate the implementation of the institutional guidelines in everyday medical practices and, vice versa, the capacity of HCPs' understandings to set the institutional agenda. For this purpose, starting from a systematic review of the articles published in *Haemophilia* journal, it will illustrate the main issues proposed by the academic literature in dealing with this care model. These issues will be framed according to the institutional guidelines set by the WFH, in order to identify the overlapping between scientific sources and WFH guidelines. This literature review will lead to an extensive theoretical reflection on this topical issue and will allow us to identify and emphasise neglected and/or emerging aspects that might be relevant for future research and interventions.

Method

The review of the scientific literature currently available was conducted by applying the following three-steps procedure (i.e., search, qualitative synthesis, discussion), inspired by the Search, AppraisaL, Synthesis and Analysis (SALSA) analytic framework.

The first step regarded the search procedure. The review was focused on *Haemophilia* journal, since this international periodical publication is the official journal of WFH, thus representing an influential venue for providing readers with an overview of the current state of the art on haemophilia and bleeding disorders at large. Moreover, the aim of this review is in line with the Journal's scope: *Haemophilia* is 'dedicated to the exchange of information regarding the comprehensive care of haemophilia'¹. The main goal of the current paper was to explore how the articles published in *Haemophilia* journal apply and describe the notion of CC: that is, whether this notion is coherent with the institutional guidelines previously outlined or, in contrast, it proposes partial or specific views. All the 4,446 articles published since 1995, when *Haemophilia* journal was founded, to the end of 2016 were considered for this review. In particular, these articles were selected according to the presence of the words 'comprehensive' and 'care' within the title or abstract.

Then, these documents were assessed according to the 'relevance' of the notion of CC developed within each article. To be specific, they were categorised into 'high relevance articles' (i.e., the article is explicitly devoted to CC), 'medium relevance articles' (i.e., the article gives definitions, explanations or information about this notion) and 'low relevance articles' (i.e., the article only uses the term CC without adding any definitions, explanations, or information).

Finally, medium and high relevance articles were included in the subsequent qualitative synthesis and discussion aimed at identifying and commenting the main features of CC proposed by the authors.

Results

General information

¹ cf. http://onlinelibrary.wiley.com/journal/10.1111/(ISSN)1365-2516/homepage/ProductInformation.html

The search procedure was completed using the four steps, which are graphically summarised

in Figure 2.

[Figure 2 near here]

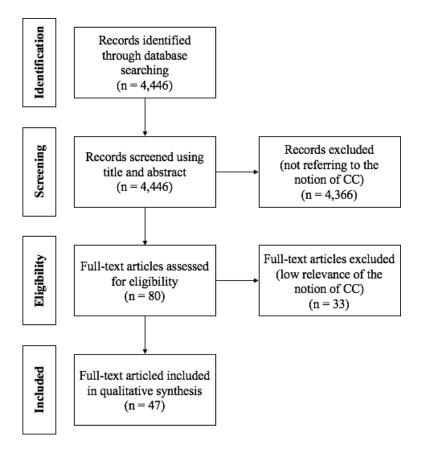


Figure 2. Review PRISMA flow diagram.

CC was mentioned in titles and/or abstracts in 80 articles, which constituted our initial corpus, distributed in volumes and years as illustrated in Figure 3. Figure 3 shows the irregular, but generally increasing, trend of articles that explicitly referred to CC over time.

[Figure 3 near here]

Relevance of comprehensive care

An eligibility screening of the corpus led us to assess the articles according to the relevance of the notion of CC and to exclude 33 low relevance articles, where the notion of CC was just mentioned rather than actually developed throughout the entire paper. Thus, the retained corpus consisted of 47 articles: 36 medium relevance articles and 11 high relevance articles. These three categories of articles were distributed discontinuously over time; however, the frequency of medium and high relevance articles increased within the most recent volumes (Figure 3).

Notion of comprehensive care in specialised understandings

A qualitative investigation of medium and high relevance articles showed that only a few of them (n=7) adopted the notion of CC consistently with the institutional guidelines. In contrast, the majority of articles (n=24) focused on a single feature of this notion, thus providing a partial view of the model. Lastly, some papers (n=16) discussed specific issues related to the construct of CC. Table 1 summarises the main features of CC considered in the analysed articles, providing a comparison between what is proposed in institutional guidelines and in specialised understandings.

[Table 1 near here]

Complete correspondence with the definition of CC in institutional guidelines

The articles adhering to a complete definition of CC can be classified into two groups. A first group consisted of articles providing an overview of CC principles (*n*=4). This is the case of two seminal works by Evatt [7,8], which were specifically devoted to the notion of haemophilia CC. In particular, Evatt et al. [7] illustrated how this care model was adopted in - and adapted to - different geographical contexts around the world; Evatt [8] discussed the

challenges to be faced in order to fully develop and sustain CC globally. In a similar vein, Skinner [9] stressed the importance of ensuring continued global collaboration and advances on the research front to reduce the care gap between developed and developing countries. Lastly, Colvin et al. [10] set out the principles of haemophilia CC by focusing on the European context.

A second group consisted of articles that applied the definition of CC to specific case studies (n=3). Teixeira et al. [11] showed how national and local haemophilia registries could represent valuable tools to ensure CC, as they support HCPs, empower PWHs and improve communication. Escobar et al. [12] described the best practices developed and adopted by HCCCs to adhere and extend the CC model. More recently, Zia et al. [13] highlighted the need to offer CC to women with heavy menstrual bleeding and blood disorders as well as to design specific programmes in this direction.

Partial correspondence with the definition of CC in institutional guidelines

The articles providing a partial view of CC could be classified into several groups as well. *Focus on the first assumption of CC.* The first group included scientific contributions especially interested in the first assumption underlying the notion of CC, that is, the importance of jointly promoting physical and psychosocial health (*n*=6). In this group, the gender viewpoint came again to the forefront. Kouides [14] and Yang and Ragni [15] discussed how testing and care provision in HCCCs may empower women with bleeding disorders to seek medical care and improve their quality of care. Tedgård [16] and Miller [17] underlined how genetic counselling, carrier testing and prenatal diagnosis of haemophilia, as well as their psychological consequences, represent a main dimension of CC, since they could enable conscious personal decisions concerning marriage and childbearing. Furthermore, Elander and Barry [18] addressed the topic of pain and its management in

haemophilia. Lastly, Wodrich et al. [19, p.593], who stated that 'comprehensive care of boys with haemophilia must include awareness of his psychosocial and physical health status', investigated the hypothesis that boys with extreme levels of attention deficit hyperactivity disorder (ADHD) symptoms are over-represented among boys with haemophilia. Focus on the first assumption of CC, especially on physical health. A further group was composed of articles that underlined the first assumption as well, but with a particular interest in physical aspects rather than in psychosocial ones (n=5). Evatt et al. [20] and Berntorp et al. [21] focused on treatments. They stressed the need to exactly define when and which prophylaxes should be adopted, as well as to periodically re-evaluate and adjust the adopted management techniques and used therapeutic products. Moreover, surgeries were the main topic of the contributions proposed by Rangarajan et al. [22] and Rodriguez-Merchan [23]. These papers discussed that a wide variety of surgeries can be successfully performed in adult PWHs within a CC setting and described the standards for the correct care of postoperative wounds. Lastly, Salem and Eshghi [24] addressed oral and dental health. They stressed the long-term beneficial effects for patients with bleeding disorders of receiving supportive oral and dental care at a young age.

Focus on the first assumption of CC, especially on psychosocial health. On the other hand, only two articles precisely emphasised the promotion of psychosocial health, without taking into account the other aspects proposed in institutional guidelines. This was the case of Wiedebusch et al. [25], who discussed the importance of families' psychosocial care, by reducing psychosocial strains and enhancing the adaptive coping strategies. More recently, Iannone et al. [26] advocated for the incorporation of depression screening and treatment as part of CC for adult PWHs.

Focus on the second assumption of CC. Then, an additional group gathered the articles focused on the second assumption underlying the notion of CC, that is, the need to co-

ordinate different HCPs into multidisciplinary teams (*n*=5). Cahill et al. [27], who mentioned the notion of CC for the first time, and Kuhathong et al. [28] focused on the role of nurses within a multidisciplinary team in HCCCs. These authors discussed how nurses act as both CC providers for patients and as trainers for patients and their caregivers. Heijnen et al. [29] provided an overview of the multiple roles played by the physiatrists as members of multidisciplinary teams in different Countries, including transversal education, specialised treatment and PWHs involvement. Poon and Luke [30] highlighted the importance of a continuative training for multidisciplinary HCPs in order to develop haemophilia CC with an emphasis on service, education and research. Lastly, Mauser-Bunschoten et al. [31] addressed the need for a unique, consistent and exhaustive documentation concerning the patients' status and history: this document should be filled out, consulted and shared among all the members of CC teams.

Focus on the both assumptions of CC, especially on physical health. Further articles stressed both the above-mentioned assumptions, but focusing on the necessity of a multidisciplinary approach for the promotion of physical health, ignoring any psychosocial purpose (*n*=5). De Kleijn et al. [32], Giangrande et al. [33] and Kulkarni [34] addressed the topic of surgeries. They argued that surgeries should only be undertaken in HCCCs, which have the requisite multidisciplinary experience and facilities, and illustrated a number of coordinated procedures carried out by multidisciplinary teams to ensure optimal outcomes before, during and after surgeries. Dargaud and Negrier [35,p.228] described the characteristics that a HCCC should possess: 'collaboration between the widest possible range of specialist staff, 24-h access to medical expertise, laboratory technology for the diagnosis and management of haemophilia, and appropriate and sufficient treatment products'. More recently, Kulkarni et al. [36] suggested that the use of telemedicine may become a useful tool to devise, deliver and monitor multidisciplinary CC to patients.

Focus on the both assumptions of CC, especially on psychosocial health. In contrast, only one article focused on both the assumptions with a specific emphasis on the promotion of psychosocial health, rather than considering issues related to patients' physical health. Coppola et al. [37,p.26] claimed that providing CC, counselling and support to PWHs and their caregivers in HCCCs has always been problematic due to practical and psychological reasons. In this regard, the authors stated that 'the joint analysis of the psychological burden of haemophilia, with the identification of needs arising from clinical problems in every stage of life, should be viewed as the common background for the group of haemophilia specialists and psychologists working together with two goals: developing a network of psychological support services in some Italian haemophilia centres, and promoting specific educational programmes for training professionals in haemophilia comprehensive care'.

Emerging aspects with respect to the definition of CC in institutional guidelines

Finally, the articles proposing specific issues in CC can be gathered into three main perspectives. Starting from the notion of CC as proposed in institutional guidelines, these viewpoints could be labelled as: 'care for all', 'transition care', and 'systemic care'. Although these tags were somehow included in the previous groups of papers, since they implicitly represent transversal assumptions underlying this care model, they explicitly became the central core of this last set of articles.

Care for all. Several papers proposed the 'care for all' view (*n*=6). Srivastava et al. [38] addressed the issue of haemophilia management in developing countries. The authors provided an overview of the main problems emerged: along with the low level of social awareness, these difficulties entail the inadequateness of the facilities and the lack of products for therapy and their costs. Non-affluent countries were the focus of other two following articles as well. Santiago-Borrero et al. [39,p.389] stated that 'the present paradigm

of haemophilia care is the comprehensive care approach, which includes a well-coordinated team of health professionals providing comprehensive continuous services and support to the patients and their relatives'. Services and support were discussed in terms of diagnosis, needs assessment of PWHs and collaboration among HCPs. However, this paradigm requires high charges that usually are not accessible across all countries. Isarangkura [40] highlighted the need for increasing the number and the quality of facilities, as well as ensuring the continuity in product availability therein. More recently, these points were also underlined by Tang et al. [41], who discussed how CC should improve in China. These scholars stressed the importance of establishing many more facilities, which should rely on adequate infrastructures and CC teams. Availability and affordability of the products were considered as well. Developed countries, and specifically the United States of America, were the focus of two other articles. Duncan et al. [42,p.519] addressed cost-management strategies. The authors discussed how 'to curb the cost of specialty pharmaceuticals in conditions when their expenditures represent a significant portion of total annual costs of care'. Similarly, Aledort [43] concluded his historical review with the current challenges for the haemophilia CC, including its maintenance despite decreased funding.

Transition care. A further group fostered the 'transition care' view (*n*=4). Oldernburg et al. [44] explicitly took an age-related perspective and stated that, as a consequence of the growing life expectancy of PWHs, the spectrum of the current CC should be extended to diseases generally considered as a prerogative of elderly patients. In the same issue, specifically devoted to transition care, Dolan et al. [45] took the same stance. The authors stressed that the delivery systems of medical CC for PWHs require a deep modification in order to provide appropriate services for the management of the diseases of older age. More recently, Smith et al. [46,p.e4] further underlined this point, by stating that 'the comprehensive care team needs to adapt to the changing needs of the ageing adult with an

inherited bleeding disorder'. Differently, Breakey et al. [47] focused on adolescence and its challenges, that result in corresponding additional issues for haemophilia CC. Since institutional guidelines do not sufficiently foster a conscientious approach to transition care, the authors suggested to significantly advance research in this specific field. Moreover, two recent articles merged the 'care for all' and 'transition care' views. On the one hand, Page et al. [48,p.535] provided an assessment of CC program services and resources in Canada. The authors concluded that 'many programmes have serious human resource shortcomings in their core disciplines. As a result, some programmes are unable to fully respect key standards of care'. They also added that 'this situation is exacerbated by a new and little studied problem, the ageing of people with bleeding disorders. The co-morbidities of ageing are increasing the complexity of care and treatment, with the potential to significantly increase demands on services'. On the other hand, Buzzi et al. [49,p.e320-321] stated that 'in recent years, public funding for haemophilia support programmes [...] has been reduced across many countries, placing greater responsibility on the community to drive local initiatives'. Therefore, the authors emphasised that 'more community-led programmes are needed to further empower PWH and their families worldwide, with the aim of providing comprehensive support throughout all life stages'.

Systemic care. Finally, some articles suggested the 'systemic care' view (*n*=4). Pritchard and Page [50], as well as Calizzani et al. [51], translated this view in terms of 'multi-level care'. Specifically, Pritchard and Page [50] pointed out that CC should involve a variety of actors (e.g., organisations, institutions, community agencies, governments) working together in the continuum of care; in other words, it should fully reflect the broader healthcare system through a vertical integration process. In parallel, Calizzani et al. [51] recommended that the principles underlying haemophilia CC should be considered for implementation within local policy planning and organisational frameworks. Differently, Hacker et al. [52], as well as

Grogan et al. [53], interpreted the 'systemic care' view in terms of 'bottom-up care'. In particular, Hacker et al. [52] called for a broader PWHs' involvement to helpfully guide decisions concerning CC services. Similarly, Grogan et al. [53] advocated that the role of PWHs in service quality development is crucial. The authors concluded that a model of PWHs collaboration within haemophilia services should be suitable for the evaluation, planning, development and delivery of care.

Discussion

The current literature review allows for drawing some closing remarks about similarities and differences between the notions of haemophilia CC in institutional guidelines and specialised understandings.

Concerning institutional guidelines, haemophilia CC has increasingly become a milestone among the existing care models over time. Consequently, this notion has been defined following precise principles, constantly updated, and corresponding recommendations and practices. These guidelines seem to play an essential role: they drive the actors involved (i.e., PWHs, caregivers, HCPs, but also organisations and institutions) toward the optimal management and care of this rare, chronic and complex disease. The continuous search for excellence in the quality of treatments provided represents a common challenge that characterises health systems at a global level. However, recent studies have defined this challenge as increasingly impelling over time due to demographic and epidemiological transformations underway. Many of these transformations reflect some of the specificities of haemophilia: in particular, literature has been focusing on the importance of promoting a model of integrated care for rare diseases [54] and for chronic diseases, which are constantly increasing [55-57], especially those at risk of co-morbidity [58,59].

As regards specialised understandings, the emerging scenario is more fragmented.

First, it should be noted that very few articles explicitly refer to CC, and that they are rarely devoted to this recognised care model. This lack of reference may be interpreted in a twofold way: the notion of CC may be taken for granted or, on the contrary, it may represent an overmultifaceted care model, which is thus difficult to be exhaustively implemented in everyday medical practices and discussed in the literature. This last interpretation is consistent with recent literature that has dealt with defining integrated care, highlighting the difficulty of proposing a consensual and univocal definition [60-62].

Moreover, in line with the previous point, only a few articles completely adhere to the notion of CC as proposed in the institutional guidelines. In contrast, some gaps between institutional guidelines and specialised understandings can be highlighted.

Most articles focus on a single feature of this notion, thus providing a partial view of CC. The emphasis is mainly on promoting physical health, which is however commonly associated with psychosocial health. This reflects the more recent and increasing interest of scholars in the psychosocial issues faced by PWHs and their families, shown by many empirical studies aimed to highlight subjective experiences in everyday life [e.g., 63], or to assess the impact of psychological and socio-educational interventions implemented in collaboration with HCCCs [e.g., 64-67].

Nevertheless, if the psychosocial dimension of haemophilia has been largely treated in the literature with regard to PWHs and their families, the psychosocial experience of professionals working in this field has been marginally explored. By exploring the literature tackling this issue, a limited production was identified, and it was mainly focused on the risks of burnout due to the treatment of a chronic illness potentially subject to complications and co-morbidities [e.g., 68-70]. Yet, psychological literature has indicated that an increased awareness of the psychosocial aspects of work can improve the organisational well-being, encourage increasing attention to multidisciplinary teams and teamwork, and reduce the

incidence of psycho-emotional difficulties in HCPs, thereby improving the quality of care [e.g., 71]. For instance, the current review indicates that those papers focusing on multidisciplinary teams highlights the potential impact of several psychosocial issues related to the CC model, such as the need to be coordinated, to assume new roles, and to share information. These issues imply corresponding skills, which are transversal across various professional roles, but are also expressed and contextualised depending on the specific duties and responsibilities pertaining to each role. Such skills – defined as soft skills in the field of work and organisational psychology [72,73] – are thus pivotal in order to achieve a CC approach, and may be integrated in forthcoming research designs, tools development, training paths and may orient how to organise work in HCCCs.

In addition, the notion of CC that emerged from the literature review puts at the forefront some specific issues. These issues are implicitly included in institutional guidelines as well, but they become the core topic of several articles.

Therefore, starting from the institutional definition of CC, these articles explicitly reflect on three current challenges that should be carefully handled: 'care for all' (i.e., the accessibility to care, both in terms of suitability/reachability of facilities and affordability/availability of treatments), 'transition care' (i.e., attention to specific age-related issues), and 'systemic care' (i.e., inclusive care, both in terms of involving all the institutional actors included in the healthcare system and promoting PWHs' participation in decision-making processes). This result is coherent with a previous study reporting the results of a lexico-metric analysis of interviews with HCPs working with haemophilia across different countries, with the main purpose of mapping their shared cognitive, affective and behavioural experiences [74,75]. This research pointed out the recurrence of relevant themes that overlap to a certain extent with the challenges identified in the present literature review. Specifically, along with communicating diagnosis and counselling, and considering PWHs' relational networks, the

following themes arise: dealing with policies, resources and stakeholders in the local care system (i.e., 'care for all' and 'systemic care'); taking care of impaired adult or elderly PWHs (i.e., 'transition care'); coping with child or adolescent PWHs (i.e., 'transition care'). Such emerging issues should be developed further as they offer the opportunity to clarify and expand the notion of haemophilia CC, by increasing its coherence and accuracy. Achieving a definition of CC that is both inclusive and accurate has important implications for everyday medical practices. On the one hand, it allows to formulate practical recommendations and proper indicators for evaluating the effectiveness of healthcare programmes. On the other hand, it allows to better comprehend the options available and to stimulate developments in research designs and interventions.

In this regard, starting from these closing remarks, a research program funded by the HERO Research Grant was started². This program aims to integrate the afore-mentioned observations, introducing a broader psychosocial perspective in the notion of CC. The project is based on the idea that each of the five themes identified by Palareti et al. [74] and Poti et al. [75] represents a subject where the use of different perspectives (HCPs, PWHs, families and patients' associations) can lead to the identification of relevant cross-professional and non-technical skills that support haemophilia CC in both its assumptions [cf. also 76-78]. Asking all the actors and stakeholders involved to express their viewpoints and needs is a crucial point for improving the coordination of the care provided: indeed, in line with previous research, this process help to 'understand sense-making processes and how roles are redefined and negotiated' [61,p.38].

² Palareti L. & Guglielmi D. (2016). 'Health Professionals and the psychosocial aspects of their work: a proposal for the expansion of core-curriculum based on the experiences of professionals and clients', Hero research grant 2016.

References

[1] Srivastava A, Brewer AK, Mauser-Bunschoten EP, et al. Guidelines for the management of hemophilia. Haemophilia. 2013;19:e1–e47.

[2] De Moerloose P, Fischer K, Lambert T, et al. Recommendations for assessment, monitoring and follow-up of patients with haemophilia. Haemophilia. 2012;18:319–325.

[3] Batorova A, Martinowitz U. Intermittent injections vs. continuous infusion of factor VIII in haemophilia patients undergoing major surgery. Br J Haematol. 2000;110:715–720.

[4] Hermans C, Altisent C, Batorova A, et al. European Haemophilia Therapy Standardisation Board. Replacement therapy for invasive procedures in patients with haemophilia: literature review, European survey and recommendations. Haemophilia. 2009;15:639–658.

[5] Soucie JM, Symons J, Evatt B, et al. Home-based factor infusion therapy and hospitalization for bleeding complications among males with haemophilia. Haemophilia. 2001;7:198–206.

[6] Teitel JM, Barnard D, Israels S, et al. Home management of haemophilia. Haemophilia.2004;10:118–133.

[7] Evatt BL, Black C, Batorova A, et al. Comprehensive care for haemophilia around the world. Haemophilia. 2004;10(Suppl.4):9–13.

[8] Evatt BL. The natural evolution of haemophilia care: developing and sustaining comprehensive care globally. Haemophilia. 2006;12(Suppl.3):13–21.

[9] Skinner MW. WFH-the cornerstone of global development: 45 years of progress.Haemophilia. 2008;14(Suppl.3):1–9.

[10] Colvin BT, Astermark J, Fischer K, et al. Inter Disciplinary Working Group. European principles of haemophilia care. Haemophilia. 2008;14:361–374.

[11] Teixeira L, Ferreira C, Santos BS, et al. Web-enabled registry of inherited bleeding disorders in Portugal: conditions and perception of the patients. Haemophilia. 2012;18:56–62.
[12] Escobar M, Maahs J, Hellman E, et al. Multidisciplinary management of patients with haemophilia with inhibitors undergoing surgery in the United States: perspectives and best practices derived from experienced treatment centres. Haemophilia. 2012;18:971–981.
[13] Zia A, Lau M, Journeycake J, et al. Developing a multidisciplinary Young Women's

Blood Disorders Program: a single-centre approach with guidance for other centres.

Haemophilia. 2016;22:199-207.

[14] Kouides PA. Females with von Willebrand disease: 72 years as the silent majority.Haemophilia. 1998;4:665–676.

[15] Yang MY, Ragni MV. Clinical manifestations and management of labor and delivery in women with factor IX deficiency. Haemophilia. 2004;10:483–490.

[16] Tedgård U. Carrier testing and prenatal diagnosis of haemophilia: utilisation and psychological consequences. Haemophilia. 1998;4:365–369.

[17] Miller R. Counselling about diagnosis and inheritance of genetic bleeding disorders: haemophilia A and B. Haemophilia. 1999;5:77–83.

[18] Elander J, Barry T. Analgesic use and pain coping among patients with haemophilia.Haemophilia. 2003;9:202–213.

[19] Wodrich DL, Recht M, Gradowski M, et al. Is attention deficit hyperactivity disorder over-represented among HIV-seronegative boys with haemophilia? Preliminary results from our centre. Haemophilia. 2003;9:593–597.

[20] Evatt BL, Farrugia A, Shapiro AD, et al. Haemophilia 2002: emerging risks of treatment. Haemophilia. 2002;8:221–229.

[21] Berntorp E, De Moerloose P, Ljung CR. The role of prophylaxis in bleeding disorders.Haemophilia. 2010;16(Suppl.5):189–193.

[22] Rangarajan S, Yee TT, Wilde J. Experience of four UK comprehensive care centres using FEIBA[®] for surgeries in patients with inhibitors. Haemophilia. 2011; 17, 28–34.
[23] Rodriguez-Merchan EC. Surgical wound healing in bleeding disorders. Haemophilia. 2012;18:487–490.

[24] Salem K, Eshghi P. Dental health and oral health-related quality of life in children with congenital bleeding disorders. Haemophilia. 2013;19:65–70.

[25] Wiedebusch S, Pollmann H, Siegmund B, et al. Quality of life, psychosocial strains and coping in parents of children with haemophilia. Haemophilia. 2008;14:1014–1022.
[26] Iannone M, Pennick L, Tom A, et al. Prevalence of depression in adults with

haemophilia. Haemophilia. 2012;18:868-874.

[27] Cahill MR, Woosey CP, Hayden SM, et al. Implementation of a nurse practitioner policy for the requisition and administration of drugs in a haemophilia comprehensive care centre. Haemophilia. 1995;1:172–174.

[28] Kuhathong K, Chuansumrit A, Keorochana S, et al. Nursing roles in orthopaedic joint correction in haemophiliac patients. Haemophilia. 2000;6:672–676.

[29] Heijnen L, Dirat G, Chen L, et al. The role of the physiatrist in the haemophilia comprehensive care team in different parts of the world. Haemophilia.

2008;14(Suppl.3):153-161.

[30] Poon MC, Luke KH. Haemophilia care in China: achievements of a decade of World
Federation of Hemophilia treatment centre twinning activities. Haemophilia. 2008;14:879–
888.

[31] Mauser-Bunschoten EP, Hamers MJ, De Roode D, et al. Improvement of patient education and information: development of a patient's information dossier. Haemophilia. 2001;7:397–400. [32] De Kleijn P, Blamey G, Zourikian N, et al. Physiotherapy following elective orthopaedic procedures. Haemophilia. 2006;12(Suppl.3),108–112.

[33] Giangrande PL, Wilde JT, Madan B, et al. Consensus protocol for the use of recombinant activated factor VII [eptacog alfa (activated); NovoSeven[®]] in elective orthopaedic surgery in haemophilic patients with inhibitors. Haemophilia. 2009;15:501–508.
[34] Kulkarni R. Comprehensive care of the patient with haemophilia and inhibitors

undergoing surgery: practical aspects. Haemophilia. 2013;19:2–10.

[35] Dargaud Y, Negrier C. Thrombin generation testing in haemophilia comprehensive care centres. Haemophilia. 2010;16:223–230.

[36] Kulkarni R, Mauser-Bunschoten EP, Stedman C, et al. Medical co-morbidities and practice. Haemophilia. 2014;20(Suppl.4):130–136.

[37] Coppola A, Cerbone AM, Mancuso G, et al. Confronting the psychological burden of haemophilia. Haemophilia. 2011;17:21–27.

[38] Srivastava A, Chuansumrit A, Chandy M, et al. Management of haemophilia in the developing world. Haemophilia. 1998;4:474–480.

[39] Santiago-Borrero PJ, Ortiz I, Rivera-Caragol E, et al. Financial aspects of haemophilia care in Puerto Rico and other Latin American countries. Haemophilia. 1999;5:386–391.

[40] Isarangkura P. Haemophilia care in the developing world: benchmarking for excellence.Haemophilia. 2002;8:205–210.

[41] Tang L, Wu R, Sun J, et al. Short-term low-dose secondary prophylaxis for severe/moderate haemophilia A children is beneficial to reduce bleed and improve daily activity, but there are obstacle in its execution: a multi-centre pilot study in China. Haemophilia. 2013;19:27–34.

[42] Duncan N, Roberson C, Lail A, et al. A haemophilia disease management programme targeting cost and utilization of specialty pharmaceuticals. Haemophilia. 2014;20:519–526.

[43] Aledort LM. The evolution of comprehensive haemophilia care in the United States: perspectives from the frontline. Haemophilia. 2016;22:676–683.

[44] Oldenburg J, Dolan G, Lemm G. Haemophilia care then, now and in the future. Haemophilia. 2009;15(Suppl.1):2–7.

[45] Dolan G, Hermans C, Klamroth R, et al. Challenges and controversies in haemophilia care in adulthood. Haemophilia. 2009;15(Suppl.1):20–27.

[46] Smith N, Bartholomew C, Jackson S. Issues in the ageing individual with haemophilia and other inherited bleeding disorders: understanding and responding to the patients' perspective. Haemophilia. 2014;20:e1–e6.

[47] Breakey VR, Blanchette VS, Bolton-Maggs PH. Towards comprehensive care in transition for young people with haemophilia. Haemophilia. 2010;16:848–857.

[48] Page D, Crymble S, Lawday K, et al. Penny wise, pound foolish: an assessment of Canadian Hemophilia/inherited bleeding disorder comprehensive care program services and resources. Haemophilia. 2016;22:531–536.

[49] Buzzi A, Kelley L, Gregory M, et al. Improving comprehensive care in the haemophilia community: building on the HERO Study. Haemophilia. 2016;22:e320–e322.

[50] Pritchard AM, Page D. Integrated specialty service readiness in health reform:

connections in haemophilia comprehensive care. Haemophilia. 2008;14:436–443.

[51] Calizzani G, Vaglio S, Arcieri R, et al. Models for institutional and professional accreditation of haemophilia centres in Italy. Haemophilia. 2013;19:e248–e255.

[52] Hacker MR, Primeaux J, Manco-Johnson MJ. A patient satisfaction survey for haemophilia treatment centres. Haemophilia. 2006;12:163–168.

[53] Grogan A, Coughlan M, O'Mahony B, et al. The development of a patient partnership programme and its impact on quality improvements in a comprehensive haemophilia care service. Haemophilia. 2012;18:875–880.

[54] Hannemann-Weber H. Influence of team composition and knowledge sharing on the ability to innovate in patient-centered healthcare teams for rare diseases. J Manag Mark Healthc. 2011;4:265-272.

[55] Nuño-Solinis R, Vázquez Pérez P, Polanco NT, et al. Integrated care: The Basque perspective. Int J Healthc Manag. 2013;6:211-216.

[56] Senitan M, Alhaiti AH, Gillespie J. Improving integrated care for chronic non-communicable diseases: A focus on quality referral factors. Int J Healthc Manag. In press.
[57] Vilaça Mendes E, de Cássia Bertão Cataneli R, Arroio Nicoletti RE, et al. Integrated care in the unified health system of Brazil: The laboratory for innovation in chronic conditions in Santo Antônio do Monte. Int J Healthc Manag. In press.

[58] Cabrera-León A, Jadad AR, Nuño-Solinís R, et al. Improving care for people living with chronic diseases: Innovative examples from Spain. Int J Healthc Manag. 2012;5:208-215.

[59] Urtaran Laresgoiti M, Nuño Solinis R. Integrated care innovations for patients with multimorbidity: A scoping review. Int J Healthc Manag. In press.

[60] Antunes V, Moreira JP. Approaches to developing integrated care in Europe: A systematic literature review. J Manag Mark Healthc. 2011;4:129-135.

[61] Lerum SV, Frich JC. Normative assumptions in integrated care: A conceptual discussion. Int J Healthc Manag. 2012;5:32-39.

[62] Bojakowski S, Filer L. Medicines management: An integrated approach? J Manag Mark Healthc. 2009;1:102-111.

[63] Cassis FY, Querol F, Forsyth A, et al. Psychosocial aspects of haemophilia: a systematic review of methodologies and findings. Haemophilia. 2012;18:e101–e114.

[64] Thomas D, Gaslin TC. "Camping up" self-esteem in children with hemophilia. Issues Compr Pediatr Nurs. 2000;24:253–263.

[65] Cassis F, Emiliani F, Pasi J, et al. Psychological interventions for people with hemophilia. The Cochrane Library. 2012.

[66] Breakey VR, Ignas DM, Warias AV, et al. A pilot randomized control trial to evaluate the feasibility of an Internet-based self-management and transitional care program for youth with haemophilia. Haemophilia. 2014;20:784–793.

[67] Beheshtipour N, Ghanavati S, Edraki M, et al. The Effects of Computer-BasedEducational Games on Self-Efficacy of 8-12 Children with Hemophilia. Iran J Blood Cancer.2015;7:149–154.

[68] Brown LK, Stermock AC, Ford HH, et al. Emotional reactions of haemophilia health care providers. Haemophilia. 1999;5:127–131.

[69] Brown LK, Schultz JR, Forsberg AD, et al. Predictors of retention among

HIV/hemophilia health care professionals. Gen Hosp Psychiat. 2002;24:48-54.

[70] Arranz P, Rodriguez-Merchan EC, Hernández-Navarro F. Burnout syndrome in haemophilia staff. In: Rodriguez-Merchan EC, Goddard NJ, Lee C, editors. Musculoskeletal Aspects of Haemophilia. Wiley Online Library; 2008. p. 212–217.

[71] Vegni E, Leone D, Biasoli C, et al. Difficult encounters with a hemophilic patient: The inner perspective of physicians. J Health Psychol. 2014;19:1499–1507.

[72] Laker DR, Powell JL. The differences between hard and soft skills and their relative impact on training transfer. Hum Resour Dev Q. 2011;22:111–122.

[73] Ribeiro L, Severo M, Ferreira MA. Performance of a core of transversal skills: selfperceptions of undergraduate medical students. BMC Med Educ. 2016;16:18.

[74] Palareti L, Potì S, Iorio A, et al. When healthcare professionals express themselves:

results overview of the qualitative phase of the HERO initiative. Haemophilia. 2016;22:13.

[75] Potì S, Palareti L, Cassis F, et al. Healthcare professionals dealing with haemophilia:

Insights from the international qualitative study of the HERO Initiative. Forthcoming.

[76] Potì S, Palareti L, Emiliani F, et al. The subjective experience of living with haemophilia in the transition from early adolescence to young adulthood: The effect of age and the therapeutic regimen. Int J Adolesc Youth. 2018;23:133-144.

[77] Palareti L, Potí S, Cassis F, et al. Shared topics on the experience of people with haemophilia living in the UK and the USA and the influence of individual and contextual variables: Results from the hero qualitative study. Int J Qual Stud Health Wellbeing. 2015;10:28915.

[78] Emiliani F, Bertocchi S, Potì S, et al. Process of normalization in families with children affected by haemophilia. Qual Health Res. 2011;21:1667-1678.

Acknowledgements

This review is supported by the Novo Nordisk HERO Research Grant 2016 within the research project "Health Professionals and the psychosocial aspects of their work: A proposal for the expansion of core-curriculum based on the experiences of professionals and clients".

Disclosure of interest

The authors report no conflicts of interest.

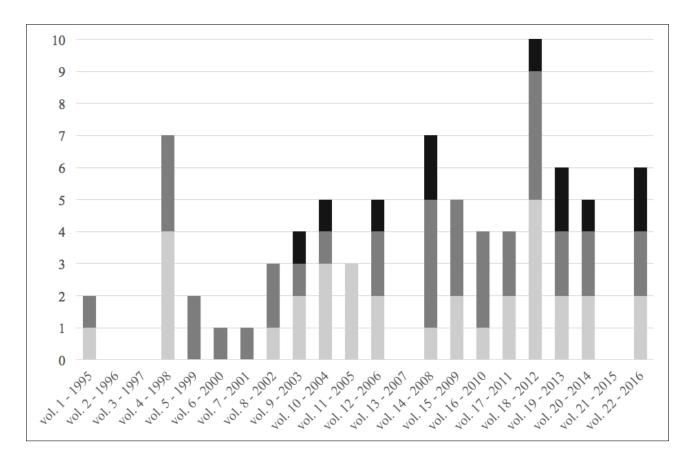
Biographical note

Sonia Brondi, PhD, is a Social Psychologist and Post-Doc Researcher at the University of Bologna (Italy). She currently works at the HERO Research Grant 2016 on the psychosocial aspects related to the work of healthcare professionals in haemophilia field. ORCID 0000-0002-9651-2216

Laura Palareti, PhD, is Assistant Professor in Social Psychology at the University of Bologna (Italy). She is the Principal Investigator of the Novo Nordisk HERO Research Grant 2016 "Health professionals and the psychosocial aspects of their work: a proposal for the expansion of core-curriculum based on the experiences of professionals and clients". Her interests are about the psychosocial and relational aspects involving families and healthcare professionals in the management of congenital bleeding diseases. She has published papers in this domain. ORCID 0000-0001-5964-1409

Greta Mazzetti, PhD, is Adjunct Professor and Research Fellow at the University of Bologna (Italy), where she teaches work and organizational psychology. She has published papers on psychological capital, organizational wellbeing and workaholism, with particular attention to the organizational and individual antecedents of this obsession with work. ORCID 0000-0002-2820-3315

Figure 2. Trend of articles that mention the notion of 'comprehensive care' by volume/year and relevance.



Note: The X axis indicates volumes and years; the Y axis indicates the number of articles in which the notion of comprehensive care has low (light grey), medium (dark grey) and high (black) relevance.

Table 1. Summary of the main features characterising the notion of comprehensive care in medium (lower cases) and high (upper cases) relevance articles.

	First assumption –	First assumption –	Second assumption –	Specific
	Physical	Psychosocial	Multidisciplinary	_
	health	health	team	issues
EVATT ET AL. [7]	*	*	*	
EVATT [8]	*	*	*	
Skinner [9]	*	*	*	
Colvin et al. [10]	*	*	*	
Teixeira et al. [11]	*	*	*	

Escobar et al. [12]	*	*	*	
Zia et al. [13]	*	*	*	
Kouides [14]	*	*		
Yang and Ragni [15]	*	*		
Tedgård [16]	*	*		
Miller [17]	*	*		
ELANDER AND BARRY [18]	*	*		
Wodrich et al. [19]	*	*		
Evatt et al. [20]	*			
Berntorp et al. [21]	*			
Rangarajan et al. [22]	*			
Rodriguez-Merchan [23]	*			
Salem and Eshghi [24]	*			
Wiedebusch et al. [25]		*		
Iannone et al. [26]		*		
Cahill et al. [27]			*	
Kuhathong et al. [28]			*	
HEIJNEN ET AL. [29]			*	
Poon and Luke [30]			*	
Mauser-Bunschoten et al. [31]			*	
	*		*	
De Kleijn et al. [32]			*	
Giangrande et al. [33]	*			
KULKARNI [34]	*		*	
Dargaud and Negrier [35]	*		*	
Kulkarni et al. [36]	*		*	
Coppola et al. [37]		*	*	
Srivastava et al. [38]				*
Santiago-Borrero et al. [39]				*
Isarangkura [40]				*
	I			1

			*
			*
			*
			*
			*
			*
			*
			*
			*
			*
			*
			*
			*
23	16	18	16
		23 16	

Note. Articles are reported following the presentation order in the text.