



The role of the specialist nurse in comprehensive care for bleeding disorders in Europe: An integrative review

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Abstract

Introduction: Managing bleeding disorders (BDs) is complex, requiring a comprehensive approach coordinated by a multidisciplinary team (MDT). Haemophilia nurses (HNs) play a central role in the MDT, frequently coordinating care. As novel treatments bring change to the treatment landscape, ongoing education and development is key. However, understanding of the roles and tasks of HNs is lacking.

Aim: The EAHAD Nurses Committee sought to identify and describe the roles and tasks of the European HN.

Methods: A five-step integrative review was undertaken, including problem identification, literature search, data evaluation, data synthesis and presentation. Relevant literature published from 2000 to 2022 was identified through database, hand and ancestry searching. Data were captured using extraction forms and thematically analysed.

Results: Seven hundred and seventy-seven articles were identified; 43 were included. Five main roles were identified, with varied and overlapping associated tasks: Educator, Coordinator, Supporter, Treater and Researcher. Tasks related to education, coordination and support were most frequently described. Patient education was often 'nurse-led', though education and coordination roles concerned both patients and health care practitioners (HCPs), within and beyond the MDT. The HN coordinates care and facilitates communication. Long-term patient care relationships place HNs in a unique position to provide support. Guidelines for HN core competencies have been developed in some countries, but autonomy and practice vary.

Conclusion: As the treatment landscape changes, all five main HN roles will be impacted. Despite national variations, this review provides a baseline to anticipate educational needs to enable HNs to continue to fulfil their role.

KEYWORDS

blood coagulation disorders inherited, haemophilia, nursing, von Willebrand disease

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1 | INTRODUCTION

Inherited bleeding disorders (BDs) are rare chronic conditions in which the blood does not clot properly. The BD treatment landscape is changing rapidly. Haemophilia has seen the most change to date, with the introduction of extended half-life factor products, novel therapies and gene therapy.¹ Clinical trials for rebalancing therapies are ongoing,² and recombinant treatments are bringing new therapeutic options for von Willebrand disease and rare BDs.^{3,4} Work to update the guidelines for European haemophilia centre certification is currently underway to ensure that the continued delivery of appropriate care includes the monitoring of novel treatment products.⁵

The management of BDs is complex, requiring a comprehensive approach by a multidisciplinary team (MDT).^{6,7} In Europe, people with a BD (PwBD) typically access care either through a European Haemophilia Comprehensive Care Centre (EHCCC) or a European Haemophilia Treatment Centre (EHTC).^{8,9} Guidelines from the European Association of Haemophilia and Allied Disorders (EAHAD) and the World Federation of Hemophilia (WFH) highlight the central role of specialist nursing staff within the MDT.^{6,8} European haemophilia centre certification criteria require at least one specialist nurse among core staff,⁹ and advanced nurse practitioners and clinical nurse specialists may be responsible for nurse-led services.¹⁰ Specialist nurses working in EHCCCs and EHTCs are generally known as haemophilia nurses (HNs) and are referred to as such in this review.

Though recognising the need for ongoing education and development in BD care,⁶ neither the WFH nor the EAHAD curriculum for HN¹¹ specify or clarify what constitutes best practice for the HN. Attempts to better define HN competencies have been undertaken at national level in some countries,^{10,12} but a broader understanding of the roles, tasks and responsibilities is lacking. In challenging the practice of HN, it is important to understand these roles to ensure that educational needs are met. To facilitate this understanding, the EAHAD Nurses Committee sought to identify and describe the role of the European HN through an integrative review.

2 | METHODS

Integrative review is widely used in nursing research to develop more comprehensive understanding of a particular health-care or practice-related issue and can inform policy and clinical practice.¹³ Our review following Whittemore and Knaf's five-stage approach¹⁴.

2.1 | Problem identification

To identify and describe the roles and tasks of the HN in the comprehensive care of PwBD.

2.2 | Literature search

We searched the PubMed, CINAHL, Cochrane, Web of Science and Embase databases for articles written in English and published between January 2000 and October 2022, using the combinations of search terms outlined in Table 1. The cut-off date of 2000 was chosen as older articles do not reflect modern comprehensive care. Journals were also hand-searched to ensure the inclusion of relevant articles not indexed in the databases.

2.3 | Data evaluation

LMW and NU undertook initial screening to remove duplicates and identify sources for secondary screening based on title and abstracts. Sources not relevant to BDs, the roles and tasks of HN, and/or the European or similar cultural contexts were excluded.

All authors were involved in secondary screening where full article texts were reviewed. As gene therapy was only available through clinical trial at this time and not embedded in the nurse role, articles concerning HNs and gene therapy were largely excluded. A data extraction form was used to capture details from included articles, including methodologies and findings.

G.M. and L.M.W. reviewed the secondary screening results and created a definitive list of sources for analysis based on the confirmation of inclusion criteria and assessment of informational value to the research question.

2.4 | Data analysis

Articles were thematically analysed using an approach appropriate to the purpose of the study and the inclusion of sources with descriptive, qualitative and mixed methodologies.¹⁵ Manual coding was undertaken to ensure a thorough analysis of the sources and the development of codes and themes. GM undertook coding in ongoing consultation with the research group. Themes were agreed by consensus during several rounds of discussion between all authors.

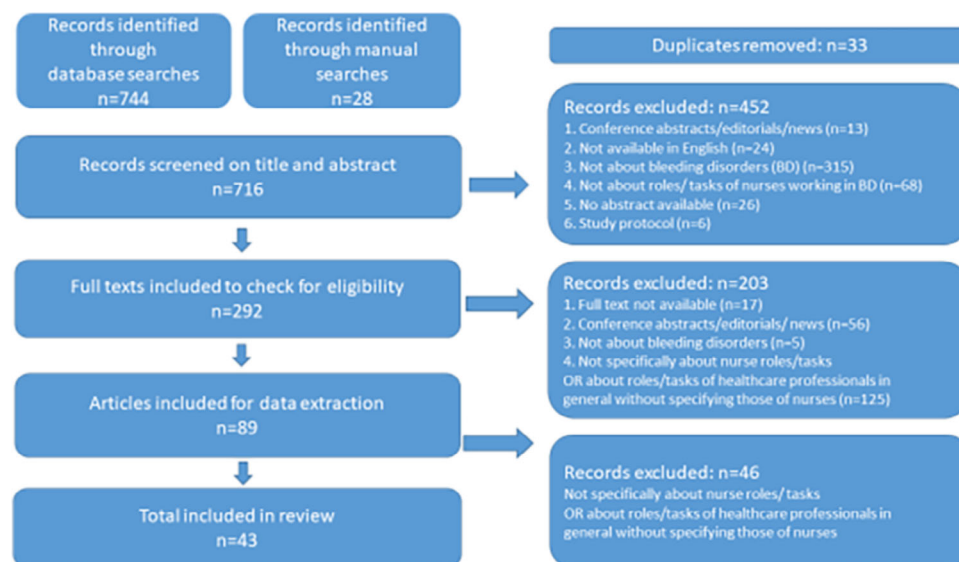
3 | RESULTS

Of 777 articles identified through the two literature searches, 43 were included for review (Figure 1 and Table 2). Five HN roles were identified:

1. Educator
2. Coordinator
3. Supporter
4. Treater
5. Researcher

TABLE 1 Search and MESH terms used in database searches.

Search	Query
#9	#4 AND #7 Filters: English Sort by: Most Recent
#8	#4 AND #7 Sort by: Most Recent
#7	#5 OR #6 Sort by: Most Recent
#6	nurses[Title/Abstract] OR nurse[Title/Abstract] OR nursing[Title/Abstract] Sort by: Most Recent
#5	"Nursing"[Mesh] OR "Nursing Staff"[Mesh] OR "Students, Nursing"[Mesh] OR "Nurses"[Mesh] OR "Education, Nursing"[Mesh] OR "Nursing Care"[Mesh] OR "Nursing Research"[Mesh] OR "Nurse's Role"[Mesh] Sort by: Most Recent
#4	Search: #1 OR #2 OR #3 Sort by: Most Recent
#3	(inherited[Title/Abstract] OR hereditary[Title/Abstract]) AND (coagulat*[Title/Abstract]) Sort by: Most Recent
#2	hemoph*[Title/Abstract] OR haemoph*[Title/Abstract] OR bleeding-disorder*[Title/Abstract] OR von Willebrand[Title/Abstract] OR bernard-soulier[Title/Abstract] OR thrombasthenia*[Title/Abstract] OR glanzmann[Title/Abstract] OR factor VIII[Title/Abstract] OR factor IX[Title/Abstract] Sort by: Most Recent
#1	"Hemophilia A"[Mesh] OR "Hemophilia B"[Mesh] OR "Blood Coagulation Disorders, Inherited"[Mesh] OR "Factor VIII"[Mesh] OR "Factor IX"[Mesh] OR "von Willebrand Factor"[Mesh] Sort by: Most Recent

**FIGURE 1** Flow chart showing the combined literature searches and process for inclusion of articles for review.

Tasks related to education, coordination and support were most frequently described.

3.1 | Educator

Almost three quarters of the articles referred to the HN's role as educator. Education of PwBD is defined as 'nurse-led', with the nurse-educator role described as 'crucial'.¹⁶

HN provides education for PwBD on aspects of self-care including bleed recognition, prevention and management, home treatment and mitigating risks,^{10,17-19} as well as explaining the genetics of BDs.²⁰⁻²² Almost 90% of nurses in a global survey identified infusion training as part of their daily practice, with most saying the same of education.²³

This educational role is lifelong for PwBD and begins with diagnosis. Nurses teach the parents/caregivers about their child's condition, treatment and potential complications²⁴; educating on home treatment and management is a core role for paediatric HN.^{17,25-27} Nurses also address bleeding issues specific to women and girls, such as heavy menstrual bleeding,²⁸ providing education around the menstrual cycle, iron deficiency and measuring blood loss, using appropriate language adjusted to the patient's age and culture.²⁹

HN ensure that PwBD are aware of the benefits of regular treatment for their future wellbeing.³⁰ They are a key point of contact for questions, information, instruction and counselling, and support PwBD to develop successful self-management strategies. In some countries, nurse associations develop guidelines and educational resources to support patient education.³¹⁻³³ Beyond the clinic, nurses provide

TABLE 2 Overview of articles included in integrative review of the roles and tasks of the haemophilia nurse in Europe.

Author/s	Year	Article title	Place of origin	Aims	Design
Aradom and Gomez ⁵⁸	2021	The patient gene therapy journey: Findings from qualitative interviews with trial participants at one UK haemophilia centre	UK	To enhance understanding of the perspectives of PwH around gene therapy, and to highlight their concerns about and motivations for having gene therapy	Structured, qualitative, one-to-one interviews (<i>n</i> = 6 PwH)
Beny et al. ⁵⁰	2022	Haemophilia in France: Modelisation of the clinical pathway for patients.	France	To identify the different actors involved in managing PwH and to determine the similarities between them to provide an accurate description of the patient trajectory.	Qualitative exploratory research study, interview-based (<i>n</i> = 31 HCPs)
Bryan ²¹	2013	How nurses built a specialty from scratch	UK	To develop, deliver and evaluate genetic education for haemophilia nurses, based on clinical roles	Expert opinion
Ballmann and Ewers ¹⁶	2022	Nurse-led education of people with bleeding disorders and their caregivers: A scoping review	Germany	To gain deeper insights into nurse-led patient education in haemophilia care and to provide a basis for a more systematic and evidence-based approach to this task	Scoping review
Burke et al. ²²	2012	Developing education tailored to clinical roles: genetics education for haemophilia nurses	UK	To develop, deliver and evaluate genetic education for haemophilia nurses, based on clinical roles	Mixed methods, questionnaire-based (<i>n</i> = 77 nurses)
Chen et al. ⁴⁴	2018	Application of optimized nursing process to perioperative patients with haemophilic pseudo-tumours	China	To optimise care during surgery for haemophilic pseudo-tumours	Retrospective study based on review of patient nursing and medical records (<i>n</i> = 16)
Cygan et al. ⁴⁷	2002	Perceptions of quality health care among parents of children with bleeding disorders	USA	To examine how parents of children with bleeding disorders define quality health care, their expectations of care in clinic and indicators of quality health care important to them	Exploratory study using two open ended questions and the Quality Health Care Questionnaire (<i>n</i> = 54 parents)
de Morais Costa et al. ⁵³	2020	North-eastern Brazilian recommendations for the nurse professional role on the pharmacokinetic-assisted prophylaxis individualization for haemophilia A	Brazil	To present recommendations of a group of expert nurses for the developing a service around the individualisation of therapy for inhibitor-negative PwHA with the aid of pharmacokinetics	Description of the development of recommendations (<i>n</i> = 7 nurses)
Ergün et al. ⁴⁰	2011	Supporting the need for home care by mothers of children with hemophilia	Turkey	To determine the difficulties experienced by mothers of children with haemophilia in maintaining their care at home and establishing appropriate interventions	Descriptive study, questionnaire-based (<i>n</i> = 20 mothers)
Escobar et al. ³⁴	2018	Recommendations on multidisciplinary management of elective surgery in people with haemophilia	USA	To develop practical approaches to implement the principles of multidisciplinary management of elective surgery for PwH	Expert panel opinion presented as a narrative summary (<i>n</i> = 14 HCPs)
Federizo et al. ⁵⁷	2018	The viability and necessity of APRN-led care models in the clinical management of haemophilia and other inherited bleeding and clotting disorders	USA	To review the impact of advanced practice registered nurses not related to haemophilia, and the potential of a haemophilia-specific education	Review

(Continues)

TABLE 2 (Continued)

Author/s	Year	Article title	Place of origin	Aims	Design
Forsyth ⁴⁹	2016	Lessons from life: the role of the advanced nurse specialist in IBDs	UK	To highlight the role of the advanced haemophilia nurse specialist	Case study
Garner et al. ³⁷	2021	The use of rIX-FP in patients with haemophilia B: a nurse's perspective	UK	To inform HCPs on the real-life management of PwHB and to discuss how treatments have advanced in recent years with the introduction of rFIX products	Review
Grieg ⁴²	2013	How specialist nurse-led care can help to lower the costs of prophylaxis	UK	To describe a project in which specialist nurses played a pivotal role in identifying a small cohort of patients eligible for dose reduction	Case study
Guelcher et al. ²⁸	2021	Women and girls with heavy menstrual bleeding and inherited bleeding disorders: A call to action for the haemophilia treatment centre nurse	UK	To discuss the benefit of expanding the role of the HTC nurse to include standardised outreach and education in the context of women and girls with IBDs and HMB, recognising the need for development to prepare nurses for this role	Review
Harrington et al. ¹¹	2016	A European curriculum for nurses working in haemophilia	UK	To develop a curriculum for haemophilia nurses that could be used as a resource in Europe, form the basis of continuous professional development and be used in the preparation of specialised education programmes	Intervention study. Curriculum skeleton reviewed by European haemophilia nurses (n = 200)
Harrison et al. ⁴³	2022	Into a brave new world: Haemophilia A & von Willebrand disease surgery with novel therapies	UK	To review the evidence for managing surgery in the era of novel therapies	Case study (n = 4)
Kadir et al. ⁴⁶	2013	Pregnancy complications and obstetric care in women with inherited bleeding disorders	UK	To summarise a workshop discussing obstetric care for women with bleeding disorders, including the importance of multidisciplinary care, the role of the haemophilia nurse, stages of pregnancy management, risk of bleeding complications, management of labour and mode of delivery and use of haemostatic agents	Review
Khair et al. ²⁴	2008	Assessment of treatment practice patterns for severe hemophilia A: a global nurse perspective	UK	To give insights into international differences in the management of haemophilia A and the evolution of haemophilia care	Intervention study, questionnaire-based (n = 105 nurses)
Khair ⁴⁸	2010	Minimizing joint damage: the role of nurses in promoting adherence to hemophilia treatment	UK	To summarise data from clinical studies evaluating the impact of prophylaxis on joint outcomes and review current findings on factors affecting adherence to prophylactic therapy	Review
Khair et al. ⁵⁹	2012	Why don't haemophilia nurses do research?	UK	To determine the barriers that prevent nurse specialists from engaging in research and to seek ways to promote clinical research by haemophilia nurses in the United Kingdom	Intervention study, survey-based (n = 32 nurses)

(Continues)

TABLE 2 (Continued)

Author/s	Year	Article title	Place of origin	Aims	Design
Khair et al. ²³	2016	Haemophilia nursing practice: A global survey of roles and responsibilities	UK	To explore the existing clinical experience, attitudes and barriers towards developing evidence-based care for haemophilia nursing globally	Intervention study, web-based survey (n = 297 nurses/22 countries)
Khair et al. ¹²	2013	A core competency framework for haemophilia nurses in the UK	UK	To describe the process of defining competence in haemophilia nursing, both those that are transferable from other areas of nursing, and those specific to haemophilia	Descriptive article focused on competency framework
Kuhathong et al. ⁴⁵	2000	Nursing roles in orthopaedic joint correction in haemophiliac patients	Thailand	To present the various roles of nurses in orthopaedic joint corrections of PwH	Descriptive article based on review of patient records (n = 14 PwH/16 surgeries)
Lazure et al. ³⁵	2018	Education needs of nurses in thrombosis and hemostasis: An international, mixed-methods study	Canada	To identify areas where nurses experience challenges when educating, treating and communicating with patients with a coagulation disorder, and to identify the causes of these challenges	Mixed-methods study, using qualitative (semi-structured interviews) and quantitative (online survey) data
Lindvall et al. ³⁰	2006	Compliance with treatment and understanding of own disease in patients with severe and moderate haemophilia	Sweden	To evaluate patients' knowledge of haemophilia and adherence to prophylactic treatment	Intervention study questionnaire (n = 108 nurses)
Lock et al. ⁴¹	2016	Optimization of home treatment in haemophilia: effects of transmural support by a haemophilia nurse on adherence and quality of life	Netherlands	To measure the effect of structured home visits by a haemophilia nurse on aspects of prophylactic home treatment in families with children with haemophilia	Intervention study based on structured home visits and questionnaires (n = 46 PwH)
Mausser-Bunschoten et al. ²⁹	2021	Managing women-specific bleeding in inherited bleeding disorders: A multidisciplinary approach	Netherlands	To support appropriate multidisciplinary care for women with bleeding disorders in the HTC	Two case studies
Mulders et al. ³⁹	2020	Do nurses have the switch factor?	Netherlands	To explore the role of specialist nurses in switching between clotting factor products and their use of a protocol	Intervention study questionnaires (n = 45 nurses)
Myrin Westesson et al. ²⁶	2018	Reaching independence through forced learning: Learning processes and illness management in parents of children affected by hemophilia	Sweden	To explore parents' learning processes and disease management in daily life during the first year after the start of their child's treatment	Grounded theory study using a longitudinal qualitative design (n = 8 parents/4 children)
Oyesiku ²⁷	2011	Home treatment of haemophilia patients with inhibitors	UK	To discuss recent guidelines and educational materials for haemophilia home care identified during a literature survey	Review
Pollard et al. ¹⁰	2020	The UK haemophilia specialist nurse: Competencies fit for practice in the 21st century	UK	To summarise the evidence supporting the nurse's advanced role in haemophilia care and develop new competencies to deliver comprehensive care within the MDT	Systematic review and results from a workshop

(Continues)

TABLE 2 (Continued)

Author/s	Year	Article title	Place of origin	Aims	Design
Santaella et al. ¹⁹	2017	Home infusion teaching practices at federally funded hemophilia treatment centres in the United States of America	USA	To identify and compare tools and resources used, and areas of unmet need, and to discuss implications for nurses in practice	Descriptive article
Santoro et al. ³¹	2009	Registered nurses for the home treatment of children with haemophilia who have developed inhibitors: a case report	Italy	To highlight the role of a registered nurse in achieving successful outcomes for a young child with haemophilia and inhibitors, and reducing the number of visits to the HTC	Case study
Schrijvers et al. ³²	2012	Learning intravenous infusion in haemophilia: experience from the Netherlands	Netherlands	To explore the burden and time-investment needed to learn intravenous infusion	Retrospective study (n = 168 PwH)
Schrijvers et al. ¹⁷	2014	The role of the European haemophilia nurse	Europe	To assess and quantify haemophilia nursing care in Europe	Descriptive study, online survey (n = 94 nurses)
Schrijvers et al. ³³	2016	Achieving self-management of prophylactic treatment in adolescents: The case of haemophilia	Netherlands	To gain insight into how adolescents achieve self-management of prophylactic treatment	Intervention study, questionnaire-based (n = 100 PwH)
St-Louis et al. ⁵⁶	2021	Multidisciplinary team care of patients with hemophilic arthropathy: A qualitative assessment of contemporary practice in the UK and Canada; Canada/UK: MDT practices for hemophilia.	Canada	To investigate MDT practices across early and late arthropathy, including specialty-specific and team-based practices; to highlight approaches to comprehensive haemophilia care; and to identify system-wide and specialty-specific barriers to successful MDT care	Structured, qualitative survey
Sulochana et al. ³⁶	2018	Head injury in a haemophilia patient—Nursing considerations: a case report	India	To address the nurse-coordinated care of a haemophilic patient with ICH in a developing country, and provide insight into the benefits of a haemophilia nurse undertaking a coordinating role	Case study
Walsh et al. [61]	2021	Identified unmet needs and proposed solutions in mild-to-moderate haemophilia: A summary of opinions from a roundtable of haemophilia experts	USA	To identify unmet needs of patients with mild-to-moderate haemophilia.	Literature review and round table summary
Witkop et al. ⁵²	2017	US haemophilia centre nurses and advanced practice providers: Demographics, roles/responsibilities, training, educational barriers and employment benefits	USA	To describe the demographics, roles/responsibilities, practice patterns, educational opportunities/barriers and employment benefits of nurses and advanced practice providers, including advanced practice registered nurses and physician assistants employed by HTCs across the United States	Qualitative intervention study, online survey (n = 438 nurses)
Witkop et al. ⁵⁵	2022	Understanding the pain management landscape within the US bleeding disorder community: a multi-centre survey.	USA	To describe pain management practice patterns of HTC providers; to identify gaps and areas of alignment with national pain guidelines; to address educational opportunities for pain management	Descriptive study; online survey (n = 236 HCPs)

(Continues)

TABLE 2 (Continued)

Author/s	Year	Article title	Place of origin	Aims	Design
Zia et al. ⁵⁴	2016	Developing a multidisciplinary Young Women's Blood Disorders Program: a single-centre approach with guidance for other centres	USA	To describe the process of developing this programme involving paediatric haematology, adolescent medicine and paediatric/adolescent gynaecology, and the expertise of a laboratory coagulationist, a nutritionist and nursing professionals	Descriptive article, expert opinion

educational outreach to schools, day-care centres and residential homes.^{12,17}

HN has a mandate to raise awareness and provide support to non-specialist health care professionals (HCPs) involved in the care of PwBD.^{18,34} Within the MDT, experienced nurses also have a role in peer-to-peer training of less experienced colleagues.³⁵

3.2 | Coordinator

HN coordinates care within the MDT and with other HCPs involved in the care of PwBD, facilitating communication between patients and care teams.^{11,17} This includes developing treatment plans, management and follow-up and concerns long-term treatment and acute episodes.³⁶ Specific tasks include liaison with laboratory staff to ensure timely processing of samples,³⁷ coordination of switches between treatment products^{38,39} and organising home care.^{40,41} This role can also extend to ensuring healthcare activities are covered by regulations and insurers.⁴² Nurse communication with all relevant HCPs is essential to ensuring that PwBD receive time-critical medicines.⁴³ Nurses play a central role in coordinating clinical care and logistics for surgical procedures, ensuring that treatment plans are understood and followed.^{34,44} They contribute to pre-, peri- and post-surgery planning, scheduling procedures, patient counselling, initiating surgical treatment plans and haemostasis support.⁴⁵ HN coordinates and supports gynaecological and obstetric care for women with BDs.^{28,46} Nurses are central to coordinating extended MDT support and planning for pregnancy, labour and delivery.⁴⁶

3.3 | Supporter

Life stages, lifestyle, cultural beliefs and support systems may raise challenges for PwBD resulting in a need for adjustment to treatment, skills, attitude and behaviour to enable good clinical outcomes.⁴⁷ The long-term, professional caring relationships HN often have with PwBD places them in a unique position to identify behavioural changes and concerns, and to work with patients to address them.¹⁰ Among the MDT, HN are often best able to advocate for and support PwBD and their families in navigating healthcare systems, helping to create a

supportive environment,³³ providing practical and psychological support individualised to patient needs.^{45,48} Nurse support is provided within the treatment centre, and via telephone, email correspondence, video calls and home visits, and extends supporting community care providers.^{17,12}

Supporting knowledge and skills is associated with education, but the nurse role in counselling and psychological support addresses patient attitude and behaviour including acceptance of their condition and treatment adherence.⁴⁹ Nurses help promote positive patient attitudes through shared decision-making, tailored and evidence-based education and advice and timely, effective discussion.⁵⁰ Good therapeutic relationships enable HN to recognise when PwBD are not coping, offer support and make referrals as needed.⁵¹

3.4 | Treater

Guidelines have been produced in Europe and the United Kingdom (UK) defining 'core competencies' and responsibilities of HN,¹⁰⁻¹² with the aim of standardising practice and ensuring nurse's central role in haemophilia care. A common core activity is the preparation and administration of medication for patients/parent/caregiver who do not self-inject.^{17,23,50} Reviewing lab results and coordinating bleed treatment are also part of regular care duties.^{10,11,49,52} Nurse practitioners and those in specialist nurse roles may assess bleeds, conduct consultation, prescribe and make referrals.^{10,11,49,52} Their role can be an essential part of providing individualised prophylactic regimens based on pharmacokinetics⁵³ and facilitating patient outcomes.²³ For women and girls presenting with menorrhagia or other gynaecological issues, HN may have the knowledge to take a gynaecological history, assess the main symptoms and treat.⁵⁴ HN role as a treater includes assessing and treating pain caused by acute bleeding episodes and chronic musculoskeletal pain.^{52,55,56} The nurse facilitates consultations and examinations with other clinical specialties, supports patient decision-making^{10,17} and is often best placed to address patient motivation and expectations around surgery, including fear, reluctance and commitment to rehabilitation.³⁴ Communication with ward staff and pharmacists is also undertaken by the nurse, who identifies and discusses particular requirements with relevant MDT members.⁵⁷

3.5 | Researcher

'Research' for HN concerns their own knowledge and that of the wider community of BD treaters, and in both cases contributes to improving health outcomes for PwBD. It is essential for HN to keep their knowledge of new research and changes in the treatment landscape up to date, and they possess the skills to do this.^{11,12,58} Nurses also participate in clinical research teams as BD experts and supports of PwBD who participate in clinical trials.¹⁷ Their roles and tasks vary here, ranging from ensuring accurate data collection to being involved in study planning and improving study protocols.^{11,38,12} While nursing aspects in clinical study protocols remain uncommon, HN have a valuable role to play in ensuring that research considers the patient rather than being solely disease-focused.⁵⁹ In some European countries, nurse-led haemophilia care is increasing.^{10,16,18,41} Experienced HNs are well placed to contribute to improvements in patient care through executing research within this field,⁵⁹ for example through service evaluation around patient journeys/pathways, patient satisfaction and implementing national guidelines and targets.

4 | DISCUSSION

Our review shows that HNs undertake a variety of linked and often overlapping roles involving education, coordination of care, nursing care support, treating and research, each of which relates to various 'nursing process components'⁵⁹ or core tasks. As well as being the primary point of contact for PwBD, they understand the viewpoint and experience of patients and their families, which supports and enables the delivery of personalised care.¹⁰ Alongside ensuring that their skills are up to date and relevant to the roles they need to fulfil, the HN role requires that they are able to educate both patients and other HCPs on BD care and management, including understanding how BDs impact other treatment needs.²⁵

Coordinating multidisciplinary care, including referral to other specialties and the organisation of care beyond the hospital setting, are tasks undertaken by the HN.^{11,17} Treatment advances in recent years mean that more PwBD are living into old age; therefore, alongside the multidisciplinary organisation of BD care, a greater number of non-specialist HCPs are likely to be involved the care of PwBDs.¹⁸ This places additional focus on the role of the nurse in facilitating a comprehensive approach to care and ensuring best outcomes in the longer term.⁶⁻⁸

As the treatment landscape continues to change, education of HN will be key to enabling them to continue to fulfil their own educational role. A global survey in 2016 found that almost 80% of HN considered patient education as a day-to-day responsibility²⁴; however, there is also evidence that many nurses lack the confidence to teach patients to self-infuse,¹⁹ and there seems to be a lack of systematic discussion and theory-guided, evidence-based practice about this role.

In Europe, as elsewhere in the world, levels of nurse education and scope of practice can vary significantly. The range of activities HN are able to undertake autonomously (e.g. modifying a treatment plan, pre-

scribing) is subject to health laws.¹⁶ Nevertheless, the organisation of haemophilia care in Europe requires a specialist nurse.⁹ An exercise undertaken by the EAHAD Nurses Committee in 2016 outlined the skills and attitudes needed to practice as a HN,¹¹ and the review presented here builds on this by confirming the range of roles and tasks that HNs may perform. However, there is still no formal standard of haemophilia nursing care or education programme to support this curriculum.

Due to time constraints, a pragmatic decision was taken to exclude conference abstracts and grey literature from our analysis, which could introduce bias.

Defining the role of the HN is complex due to variances in how haemophilia care is organised across Europe. Nurse-led BD care is increasing in some countries but does not exist in others, and the titles and roles of specialist HNs vary, as in other disease areas.

5 | CONCLUSION

This integrative review shows that HN performs multiple, complex tasks in a variety of settings, based around five key, inter-related roles – educator, coordinator, supporter, treater, researcher – among which education and coordination of care are central. Current and ongoing changes in the treatment landscape, including the introduction of novel and gene therapies, impact all five roles, and it is essential that nurses engage in continuous professional development. Although there is a need to acknowledge national variations in practice, this can and should be used to inform and prepare for the education, training and competency development that will ensure HNs remain equipped to fulfil their role and responsibilities.

AUTHOR CONTRIBUTIONS

Greta Mulders designed the study. All authors analysed the data, wrote the first draft of the paper and critically reviewed revisions of the manuscript before agreeing on its final version.

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CONFLICT OF INTEREST STATEMENT

G.M. has received a research grant support from Sobi, consultancy fees from Novo Nordisk and speaker fees from CSL Behring. N.U. has received research grant from Sobi, MFB has received consultant fees from Sobi, Novo Nordisk, Roche, Takeda, CSL-Behring and research grants from Pfizer, Sobi, Takeda, CSL-Behring, Roche, Octapharma. L.M.W. has received research grants from Takeda and is a member in a study steering committee Roche. M.K., R.K. and G.N. declare no conflicts of interest.

DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available from the corresponding author upon reasonable request.

ETHICS STATEMENT

This review is based on previously conducted studies and does not report any new data; therefore, ethical approval was not required.

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