



GUIDELINES FOR HAEMOPHILIA NURSING CARE

Guidelines on factor administration
via IV or CVAD

EAHAD Nurses Committee
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Disclaimer:

The EAHAD Nurses Committee does not provide medical advice or engage in the practice of medicine. The EAHAD Nurses Committee under no circumstances recommends particular treatment for specific individuals.

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How were the guidelines developed?

Introduction

The guidelines for intravenous (IV) and central venous access device (CVAD) infusion were designed to be an up-to-date review of the current evidence in haemophilia care. In the next section, the origin and development methodology of the European guidelines for haemophilia nursing care are described. Literature search and scoring national and local protocols/guidelines on methodological quality (using AGREE II) form the basis for the guidelines.

Despite similarities in core nursing roles across Europe, there has been little attempt to codify the evidence base for nursing interventions. Currently, there are no formal evidence-based guidelines for nursing care for bleeding disorders, such as haemophilia and von Willebrand disease. Patients suffering from these chronic illnesses have to treat themselves intravenously regularly. The haemophilia nurse plays a central role in the treatment of these patients and has a crucial part to play in educating patients about self-managing the bleeding disorder.

The guideline development process

As a first step, a committee was set up with haemophilia nurses from across Europe (n=7), with expert patient representation (n=1) and haemophilia physician/epidemiologist (n=1). All nursing members (n=7) of the steering committee had a clinical background and most of them also had a scientific background or a Master of advanced nurse practice.

Delphi procedure

The guideline was developed based on a review of the literature, analysing local and national protocols, and a Delphi consensus procedure. A literature search was conducted in the known databases. Different search terms were used (haemophilia, self-infusion, prophylaxis, infusion, CVAD, nursing, procedure, intravenous). This search was not restricted by language or year of publication, since it is known that there is limited evidence available. Titles and abstracts were scanned and potential articles were read and summarised. The available evidence about haemophilia and nursing care ranged from limited to non-existent.

Therefore, the committee collected local and national protocols and guidelines. A call for protocols and guidelines was performed through the EAHAD network and the national haemophilia nurses' societies. The guidelines received were critically appraised with the AGREE II tool: Appraisal of Guidelines for Research and Evaluation II¹. The great majority of the collected guidelines did not fulfil the AGREE criteria. The most comprehensive guidelines were compared and the steering committee decided to set up a Delphi procedure^{2,3} to achieve expert consensus. The collected guidelines and protocols were compared in a framework. Similarities and discrepancies were discussed within the steering committee.

During the Delphi meeting the collected guidelines were discussed. Points which were unclear were examined and judged with the adapted scheme of Ewenstein et al⁶. The rating scheme can be found in Table 1. Some issues remained unresolved. When there was no or conflicting evidence available, a rating of D or E was given

Guideline review and update

These guidelines were externally reviewed and approved by the EAHAD Executive Committee. They will be reviewed every five years for any updates or relevant literature which could improve them. The EAHAD Nurses Committee will perform the review.

Table 1: Rating scheme adapted from Ewenstein et al.

A	Strongly recommended for implementation and strongly supported by clinical studies
B	Strongly recommended for implementation and supported by some clinical or epidemiologic studies, widespread clinical use and/or a strong theoretical rationale
C	Recommended and supported by limited clinical evidence and regarded as reasonable clinical practice among haemophilia experts
D	Reasonable and acceptable medical practice, but recommendation is not currently warranted because of limited published evidence and/or conflicting views among haemophilia experts
E	An unresolved issue for which evidence is insufficient, no consensus regarding efficacy exists and more research is needed

Background: Guideline on intravenous infusion (IV)

The first area of clinical practice identified for this process is peripheral infusions of factor concentrates in patients with inherited bleeding disorders. A literature search conducted to identify clinical practice guidelines used in peripheral factor administration (IV) in haemophilia yielded no evidence to support the availability of any current clinical practice guidelines. A literature search in relation to central venous access devices (CVAD) in haemophilia produced no results about clinical practice guidelines for CVAD in haemophilia.

Population

Patients who were prescribed clotting factor concentrate, for example:

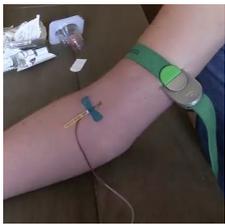
- Patients with haemophilia
- Patients with von Willebrand disease

Target users

Nurses working in haemophilia care

Benefits, side effects and risks

Health benefits:	<ul style="list-style-type: none"> • Safe injection of clotting factor (hygiene) • Throughout Europe a similar procedure enhances patients' feeling of safety • New nurses can be trained more easily • Establishes a benchmark standard of procedure that may be utilised across Europe
Side effects/ risks ⁴	<ul style="list-style-type: none"> • Pain • Subcutaneous infusion • Infection (thrombophlebitis) • Haematoma • Product-related side effects (check information leaflet of product)
Barriers:	<ul style="list-style-type: none"> • Haematoma • Difficulties with vein palpation • Skin problems (e.g., psoriasis, neurodermatitis) • Patient / parent / caregiver anxiety or unwillingness for treatment
Facilitators:	<ul style="list-style-type: none"> • Warm hands • Being well hydrated • Being relaxed; creating a relaxing environment⁵ • Having time: no hurry in preparation and/or treatment

Step by step instruction for peripheral venous access		
<p>PREPARATION</p>	<ol style="list-style-type: none"> 1. Wash hands and dry properly. Hygiene-aspects: remove jewellery, keep nails short, and ensure a clean workplace 2. Prepare equipment (alcohol swab, dry pad, tourniquet, plaster). Check butterfly connector, stretch tube gently. <i>Optional:</i> prepare tape to fix butterfly. 3. Check medication (name, date of expiration, vial size) 	
<p>RECONSTITUTION</p>	<ol style="list-style-type: none"> 4. Dissolve the powder of coagulation factor concentrate (CFC) using the water for injection provided and according to the medication leaflet 5. Fill a small volume of air in the full syringe with CFC (volume corresponding to the volume of butterfly tube) <p>OR</p> <p>Prepare a syringe with saline water (no air in the syringe with factor concentrate)</p>	
<p>INJECTION</p>	<p><i>Check for warm hands (patient) – if necessary, ask patient to warm up their hands (e.g., warm water, hot pack). Furthermore, it is recommended to switch between puncture places.</i></p> <ol style="list-style-type: none"> 6. Put on the tourniquet 7. Clean the skin of the chosen puncture vein with alcohol swab 8. Wait until the skin is dry and / or at minimum 30 seconds 9. Puncture the vein and let blood fill the butterfly tube 10. Loosen the tourniquet and connect the CFC syringe <i>Keep in mind: only first blood drop in the syringe allowed, not extra aspiration of blood.</i> 11. Bring the CFC syringe in upright position – air must be in the back of the syringe and inject the CFC slowly. End: fill the tube with air so that CFC is fully infused <p>OR</p> <ol style="list-style-type: none"> 12. Inject the CFC slowly. Connect syringe with saline water. Inject the volume of butterfly tube with saline water 	
<p>ROUNDING UP</p>	<ol style="list-style-type: none"> 1. Pull needle out before pressing with dry pad 2. Press with dry pad (minimum 1 minute) and apply plaster (preference of patient) 3. Documentation (substitution diary) 4. Ensure safe disposal 	

Background: Central venous access device (CVAD) infusion

A literature search on central venous access devices (CVAD) in haemophilia showed some results, especially on the incidence of CVAD related infections, yet none of these papers described a clinical practice guideline on how to perform CVAD infusion in haemophilia. Yet, Ewenstein et al. reviewed the current best evidence for some aspects of CVAD practice and the following recommendations were adopted:

1. Strict adherence to effective hand-washing and an aseptic technique is recommended to prevent infections⁶ (A). The use of gloves should not replace the need for hand hygiene. Sometimes gloves give the feeling of 'false security'; always take into consideration what you touch with or without gloves.
2. Povidone–iodine or chlorhexidine can be used as a topical skin antiseptic agent. Evidence exists that use of chlorhexidine may be associated with fewer catheter-related infections, and may be preferred, especially on patients with a history of infection.⁶ (B)
3. When EMLA cream® is applied: minimum 30 minutes, maximum 1 hour because the skin gets soft and is more easily damaged. Local anaesthetics should be thoroughly removed with water and soap prior to injection⁶. (B)
4. Move the skin a bit over the CVAD; do not use always the same insertion place. (D)
5. Flush with NaCl 0.9% between different medicines. After taking a blood sample, always flush with 10ml or 20ml NaCl 0.9%. (A)
6. Always block the CVAD under positive pressure (A), Heparin (100U/ml) not more than 3 ml or saline (E)^{6,7}
7. Never use syringes with a lower volume than 5ml or preferably 10ml⁸. (C)

Population

Patients who have been prescribed clotting factor concentrate, for example:

- Patients with haemophilia
- Patients with von Willebrand disease

Target users

Nurses working in haemophilia care

Benefits, side effects and risks:

Health benefits:	<ul style="list-style-type: none"> • Save injection of clotting factor (hygiene) • Throughout Europe a similar procedure enhances the patient's feeling of safety • New nurses can be trained more easily
Side effects/ risks:	<ul style="list-style-type: none"> • Skin irritation (disinfectant or lido-spray) • Skin-infection • Swelling (locally) • Haematoma <p>CVAD related complications: Infection, thrombosis, or mechanical problems of the CVAD (adopted from Ewenstein, 2004)^{6,9}:</p> <ul style="list-style-type: none"> ○ Infection: <ul style="list-style-type: none"> ▪ Swelling around CVAD site or subcutaneous catheter track ▪ Pain or redness at CVAD site ▪ Pus or drainage from CVAD site ▪ Fever or chills ▪ Systemic infection (possibility of sepsis and septic shock) ○ Thrombosis <ul style="list-style-type: none"> ▪ Local redness, pain or swelling ▪ Oedema distal to CVAD ▪ Difference in colour or temperature of extremities between affected and unaffected sides ▪ Vein engorgement on affected side ▪ Acute onset of pain and swelling in arm or base of neck ○ Mechanical <ul style="list-style-type: none"> ▪ Change in CVAD position or appearance ▪ Pain around CVAD, especially during infusion ▪ Difficulty infusing medication, flushing fluid or withdrawing blood ▪ Swelling around CVAD site or subcutaneous catheter track ▪ Bleeding from CVAD site ▪ Breakdown of skin over CVAD <ul style="list-style-type: none"> • Product-related side effects (check information leaflet of product).
Facilitators:	<ul style="list-style-type: none"> • Having time and being relaxed; creating a relaxing environment⁵ • Remove jewellery, keep nails short, clean workplace / tray with alcohol

Step-by-step instruction for substitution via central venous access device (CVAD)		
PREPARATION	<ol style="list-style-type: none"> 1. Wash hands and dry properly. Hygiene-aspects: remove jewellery, keep nails short, ensure a clean workplace 2. Gather equipment, including needle for CVAD 3. Check medication (name, date of expiration, vial size) 	
RECONSTITUTION	<ol style="list-style-type: none"> 4. Dissolve the powder of coagulation factor concentrate (CFC) using the water for injection provided and according to the medication leaflet. No air in the syringe with factor concentrate 5. Prepare 2 syringes with 10ml NaCl 0.9% 6. Unpack dry sterile gauzes (2 or 3 of size 10x10 and 1 of 5x5) 7. Moisten 2-3 large sterile gauzes with alcohol (10X10cm) 8. Fill the line of the CVAD needle and the needle with NaCl 0.9% without locking the clamp 	
INJECTION	<ol style="list-style-type: none"> 9. Encourage the patient / child to settle in a relaxing and comfortable position 10. Disinfect your hands OR put on sterile gloves 11. Disinfect the area of the CVAD twice and wait the recommended time 12. Palpate the CVAD and insert the needle perpendicularly to the CVAD until you reach the bottom 13. Aspirate slowly until you see blood flowing back OR connect syringe with 10ml NaCl 0.9% and flush it 14. Flush with at least 5ml NaCl 0.9% 15. Connect syringe with factor concentrate and infuse it 16. Connect syringe with 10ml NaCl 0.9% and flush with at least 5ml 17. Connect the syringe with Heparin and flush with 3 ml 18. Pull out the needle under positive pressure 	
ROUNDING UP	<ol style="list-style-type: none"> 19. Put on a sterile compress and apply pressure for at least 1 minute and apply plaster (preference of patient) 20. Documentation (substitution diary) 21. Ensure safe disposal 	

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Appendix 1: Compare and contrast of the highest ranked EU nursing guidelines IV

<i>Topic / City, Country</i>	Norwegian guideline	Swiss guideline (hospital)	Swiss guideline (home care)	Dutch guideline ^{10,11} (Utrecht)	UK guideline (Edinburgh, Scotland)	Recommendation (with level of evidence for it)
<i>Needle</i>	Butterfly	Butterfly	Butterfly	Butterfly	Butterfly	Yes
<i>Check connector of butterfly, stretch tube gently</i>	-	-	Yes	Yes		Yes
<i>Factor-syringe with air / without air</i>	Without	Possible with little bit of air	Without	Not mentioned With air		Discussion (C)
<i>Prefilling the syringe with concentrate</i>	Yes	No, letting blood flow back, then connecting the syringe	No, letting blood flow back, then connecting the syringe	No, letting blood flow back, then connecting the syringe	No, letting blood flow back, then connecting the syringe	(C) No, letting blood flow back, then connecting the syringe.
<i>Tourniquet where?</i>	10-15cm above	-	-	-	-	Irrelevant
<i>Aspiration of blood and mixing in syringe</i>	-	-	No	-	-	Discussion (D)
<i>Fixation of butterfly</i>	No	Possible	Possible	Yes	Possible	Preference of patient
<i>Loosen tourniquet for infusion</i>	Yes	Yes	Yes	Yes		Yes
<i>After injection?</i>	-	Air through tube or nothing	Flushing with NaCl	Make sure no air bubbles are injected; -	Flushing with 5ml NaCl	Air through tube for home treatment Discussion (see factor syringe)
<i>When pressing with gauze pad?</i>	First pulling needle out, then pressing	First pulling needle out, then pressing	First pulling needle out, then pressing	-	First pulling needle out, then pressing	Yes
<i>Press how long after injection?</i>	-	Minimum 2min	Minimum 2min	Sometime bandage	-	Recommendation: at least 1 min pressure (C)
<i>Failure: pulling back concentrate from the tube into syringe after failing</i>	Yes, if there is no blood in it	-	-	-	-	
<i>Having a clean control finger</i>	-	-	-	Yes	-	Recommended (C)
<i>Position of the syringe during injection</i>	-	-	-	Upright, extension for needle downwards	-	Recommended (C)
<i>Others</i>			Remove all jewellery		Place syringe back into sterile empty wrapper	Recommendation: don't touch the tip of the syringe (C)

Appendix 2: Compare and contrast of the highest ranked EU nursing guidelines CVAD

	Norwegian guideline	Swiss guideline (home care)	Swiss guideline (Zurich)	Swiss guideline (Basel)	UK guideline (e.g., Edinburgh, Scotland)	Dutch guideline ^{10,11} (e.g. Utrecht)	Irish guideline
<i>Port-a-Cath (PAC) Needle</i>	PAC needle	PAC needle, Huber Schliff	PAC needle	PAC needle, Huber Schliff	PAC needle, non-coring	PAC needle, Gripper	PAC needle, non-coring
<i>Gloves</i>	Sterile	Sterile	Sterile	Unsterile	Sterile	No	Sterile
<i>Mask</i>	No	No	Yes, if verbal communication during needle insertion	No	No	No	No
<i>Syringes</i>	Not mentioned	Minimum size 10ml	Minimum size 5ml	Minimum size 10ml	Minimum size 10ml	Not mentioned, seems 10ml	Minimum size 10ml
<i>Pre Flushing</i>	3-4ml NaCl (of 10ml)	3ml NaCl (of 10ml)	10ml NaCl	10ml NaCl	No	5ml NaCl	No
<i>Aspiration after needle insertion</i>	Yes NaCl – aspiration – NaCl 6-7ml (rest of 10ml)	Yes NaCl – aspiration – NaCl 7ml (rest of 10ml)	Optional, if yes: Flushing with 2ml Heparin (100U/ml) and 10ml or 20ml NaCl	no	Yes 5ml blood has to be discarded; then directly factor concentrate	Yes Aspiration – 5m NaCl	Yes appropriate volume (1.5-3ml)
<i>Flushing after factor concentrate</i>	10ml NaCl	10ml NaCl	10ml NaCl	10ml NaCl	NaCl, amount not clear, 5ml possible	5ml NaCl	NaCl, amount not clear, 5ml possible
<i>Block (End of treatment)</i>	2ml Heparin 100U/ml	5ml Heparin 100U/ml under positive pressure (Druck)	2-5ml Heparin 100U/ml, under positive pressure after minimum 2ml	10ml NaCl, under positive pressure	5ml Heparin 100U/ml under positive pressure during the last ml	5ml Heparin 100/ml under positive pressure during last ml	5ml Heparin under positive pressure during last ml
<i>Disinfection of the PAC region</i>	Without gloves Klorhexidin (5mg/ml), 2x	Without gloves Disinfection, 2x		Alcohol basis, Octeniderm (Einwirkzeit 30'')	Chlorhexidine 0.5% in 70% alcohol	Alcohol, 2x	Chlorhexidine 0.5% 3x
<i>Position of Patient</i>	Sitting, lap of adult, laying back	comfortable	Turning head to opposite side during needle insertion	-	-	-	-
<i>Work place</i>	Sterile paper from the gloves	Sterile paper from the gloves			Sterile paper from a wound cleansing pack		Sterile paper from a wound cleansing pack or gloves
<i>Miscellaneous</i>	Tube of needle lower than heart level	-	One PAC can be used for up to 2000 insertions	Move the skin over PAC to change the place of insertion into the skin	-	EMLA® 30-60min, no longer because softening skin. Remove needle	Flushing after factor: stop-start action to create pulsated flush, it

						with sterile gauze. No jewellery (e.g., rings	clears infusion set more effectively
<i>Preparing with sterile gloves</i>	Part of it	Everything	Everything	No	Part of it Factor concentrate without gloves. NaCl and Heparin vials and syringe with factor -> cleaning with alcohol wipe and placing on the sterile work place		Part of it Factor concentrate without gloves. NaCl and Heparin vials and syringe with factor -> cleaning with alcohol wipe and placing on the sterile work place
<i>Blood sampling</i>	Not mentioned	20ml NaCl flushing after	2ml Heparin 100U/ml flushing	20ml NaCl flushing after	No flushing after	-	-
<i>Control of needle</i>	Aspiration	Aspiration	Aspiration	Pulling softly after insertion	Aspiration	Aspiration	