

# Appendix

## Search strategies

### ***Hemophilia – general aspects***

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#### **PubMed (NLM) September 2009**

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Von Willebrand disease (TiAb)	AND	Controlled clinical trial (PT)	NOT	Purpura, Thrombocytopenic (Me)
Angiohemophilia (TiAb)		Meta analysis (PT)		
Blood coagulation disorders (Me)		Randomized controlled trial (PT)		
Hemophilia (TiAb)		Systematic (SB)		
Haemophilia (TiAb)				
Bleeding disorder (TiAb)		Random* (Ti)		
Bleeding disorders (TiAb)		OR Control* (Ti)		
		NOT Medline (SB)		

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("controlled clinical trial"[Publication Type] OR "meta analysis"[Publication Type] OR "randomized controlled trial"[Publication Type] OR systematic[sb] OR ((random\*[Title] OR control\*[Title]) NOT Medline[sb])) AND ("von Willebrand Disease"[title/abstract] OR "angiohemophilia"[title/Abstract] OR "Blood Coagulation Disorders"[Mesh terms] OR "hemophilia"[title/abstract] OR "haemophilia"[title/abstract] OR "bleeding disorder"[title/abstract] OR "bleeding disorders"[Title/abstract]) NOT "Purpura, Thrombocytopenic"[MeSH Terms]

## **Hemophilia – general aspects**

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### **Embase.com (Elsevier) September 2009**

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Blood clotting factor deficiency (Exp,MJR)	AND	Clinical trial (Exp)	NOT	Thrombocytopenic purpura (Exp)
Von Willebrand disease (Ti)		Intervention study (De)		
Angiohemophilia (Ti)		Longitudinal study (De)		
Angiohaemophilia (Ti)		Prospective study (De)		
Hemophilia (Ti)		Meta analysis (De)		
Haemophilia (Ti)		Systematic review (De)		
Bleeding disorder (Ti)		Random* (Ti)		
Bleeding disorders (Ti)		Control* (Ti)		

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('blood clotting factor deficiency'/exp/mjOR 'von willebrand disease':ti OR  
'angiohemophilia':ti OR 'angiohaemophilia':ti OR 'hemophilia':ti OR  
'haemophilia':ti OR 'bleeding disorder':ti OR 'bleeding disorders':ti) AND  
('clinical trial'/exp OR 'intervention study':de OR 'longitudinal study':de OR  
'prospective study':de OR 'meta analysis':de OR 'systematic review':de  
OR random\*:ti OR control\*:ti) NOT 'thrombocytopenic purpura'/exp AND  
[embase]/lim

## **Hemophilia – general aspects**

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**CENTRAL (Wiley) September 2009**

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Von willebrand disease (TiAb)        NOT     Purpura, thrombocytopenic (Me)  
Angiohemophilia (TiAb)  
Hemophilia (TiAb)  
Haemophilia (TiAb)  
Bleeding disorder (TiAb)  
Bleeding disorders (TiAb)  
Blood coagulation disorders (Me)

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#1        (von willebrand disease):ti,ab OR (angiohemophilia):ti,ab OR  
(hemophilia):ti,ab  
            OR (haemophilia):ti,ab OR (bleeding disorder):ti,ab OR  
(bleeding disorders):ti,ab  
#2        MeSH descriptor Blood Coagulation Disorders explode all trees  
#3        MeSH descriptor Purpura, Thrombocytopenic explode all trees  
#4        (( #1 OR #2 ) AND NOT #3)

## **Hemophilia – hemorrhage**

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**Embase.com (Elsevier) September 2009**

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Blood clotting disorder/DT (De)	AND	Blood clotting factor deficiency (Exp)	AND	Clinical trial (Exp)	NOT	Thrombocytopenic purpura (Exp)
Blood clotting disorder/TH (De)		Von willebrand disease (TiAb)		Intervention study (De)		
Blood clotting disorder/CO (De)		Angiohemophilia (TiAb)		Longitudinal study (De)		
Blood clotting disorder/DM (De)		Angiohaemophilia (TiAb)		Prospective study (De)		
Blood clotting disorder/PC (De)		Hemophilia (TiAb)		Meta analysis (De)		
Hemorrhage/DT (De)		Haemophilia (TiAb)		Systematic review (De)		
Hemorrhage/TH (De)		Bleeding disorder (TiAb)		Random* (Ti)		
Hemorrhage/CO (De)		Bleeding disorders (TiAb)		Control* (Ti)		
Hemorrhage/DM (De)						
Hemorrhage/PC (De)						
Hemorrhage (TiAb)						
OR Haemorrhage (TiAb)						
OR Bleeding (TiAb)						
AND Life threatening (TiAb)						
OR Fatal (TiAb)						

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(('blood clotting disorder'/dm\_dt,dm\_th,dm\_co,dm\_dm,dm\_pc  
'hemorrhage'/dm\_dt,dm\_th,dm\_co,dm\_dm,dm\_pc OR ('hemorrhage':ti,ab  
OR 'haemorrhage':ti,ab OR 'bleeding':ti,ab) AND ('life threatening':ti,ab  
OR 'fatal':ti,ab)) AND ('blood clotting factor deficiency'/exp OR 'von  
willebrand disease':ti,ab OR 'angiohemophilia':ti,ab OR  
'angiohaemophilia':ti,ab OR 'hemophilia':ti,ab OR 'haemophilia':ti,ab OR  
'bleeding disorder':ti,ab OR 'bleeding disorders':ti,ab) AND ('clinical  
trial'/exp OR 'intervention study':de OR 'longitudinal study':de OR  
'prospective study':de OR 'meta analysis':de OR 'systematic review':de  
OR random\*:ti OR control\*:ti) NOT 'thrombocytopenic purpura'/exp AND  
[embase]/lim)

## **Hemophilia – arthropathy**

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**Embase.com (Elsevier) September 2009**

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Hemophilic arthropathy (TiAb)	AND	Blood clotting factor deficiency (Exp)	AND	Clinical trial (Exp)	NOT	Thrombocytopenic purpura (Exp)
OR Haemophilic arthropathy (TiAb)		Von willebrand disease (TiAb)		Intervention study (De)		
AND Prevention (TiAb)		Angiohemophilia (TiAb)		Longitudinal study (De)		
OR Prevention (EXP)		Angiohaemophilia (TiAb)		Prospective study (De)		
OR Prophylaxis (TiAb)		Hemophilia (TiAb)		Meta analysis (De)		
OR Prophylactic (TiAb)		Haemophilia (TiAb)		Systematic review (De)		
Arthroplasty (Exp)		Bleeding disorder (TiAb)		Random* (Ti)		
Arthrodesis (Exp)		Bleeding disorders (TiAb)		Control* (Ti)		
Arthroplast* (Ti)						
Surgery (Ti)						
Surgical (Ti)						
Arthrodesis (Ti)						

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((('hemophilic arthropathy':ti,ab OR 'haemophilic arthropathy':ti,ab) AND ('prevention':ti,ab OR 'prevention'/exp OR 'prophylaxis':ti,ab OR 'prophylactic':ti,ab)) OR 'arthroplasty'/exp OR 'arthrodesis'/exp OR 'arthroplast\*:ti OR 'surgery':ti OR 'surgical':ti OR 'arthrodesis':ti) AND ('blood clotting factor deficiency'/exp OR 'von willebrand disease':ti,ab OR 'angiohemophilia':ti,ab OR 'angiohaemophilia':ti,ab OR 'hemophilia':ti,ab OR 'haemophilia':ti,ab OR 'bleeding disorder':ti,ab OR 'bleeding disorders':ti,ab) AND ('clinical trial'/exp OR 'intervention study':de OR 'longitudinal study':de OR 'prospective study':de OR 'meta analysis':de OR 'systematic review':de OR random\*:ti OR control\*:ti) NOT 'thrombocytopenic purpura'/exp AND [embase]/lim)

## ***Hemophilia – arthropathy***

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**CENTRAL (Wiley) September 2009**

Hemophilic arthropathy (TiAb)  
Haemophilic arthropathy (TiAb)

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#1 (hemophilic arthropathy):ti,ab OR (haemophilic  
arthropathy):ti,ab

## **Hemophilia – inhibitors**

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### **Embase.com (Elsevier) September 2009**

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Inhibitor (TiAb)	AND	Blood clotting factor deficiency (Exp)	AND	Clinical trial (Exp)	NOT	Thrombocytopenic purpura (Exp)
Inhibitors (TiAb)		Von willebrand disease (TiAb)		Intervention study (De)		
		Angiohemophilia (TiAb)		Longitudinal study (De)		
		Angiohaemophilia (TiAb)		Prospective study (De)		
		Hemophilia (TiAb)		Meta analysis (De)		
		Haemophilia (TiAb)		Systematic review (De)		
		Bleeding disorder (TiAb)		Random* (Ti)		
		Bleeding disorders (TiAb)		Control* (Ti)		

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((('inhibitor':ti,ab OR 'inhibitors':ti,ab) AND ('blood clotting factor deficiency':exp OR 'von willebrand disease':ti,ab OR 'angiohemophilia':ti,ab OR 'angiohaemophilia':ti,ab OR 'hemophilia':ti,ab OR 'haemophilia':ti,ab OR 'bleeding disorder':ti,ab OR 'bleeding disorders':ti,ab) AND ('clinical trial':exp OR 'intervention study':de OR 'longitudinal study':de OR 'prospective study':de OR 'meta analysis':de OR 'systematic review':de OR random\*:ti OR control\*:ti) NOT 'thrombocytopenic purpura':exp) AND [embase]/lim)

## ***Hemophilia – complications***

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PubMed (NLM) September 2009

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Blood coagulation disorders, inherited/CO (Me) AND Hemophilia (Ti) AND Clinical trial (PT)  
Blood coagulation disorders, inherited/DT (Me) AND Haemophilia (Ti) Review (PT)  
Blood coagulation disorders, inherited/TH (Me) AND Retrospective studies (Me)  
Blood coagulation disorders (MJR) AND Humans (Me)  
AND English (La)

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("Blood Coagulation Disorders, Inherited/complications"[Mesh] OR  
"Blood Coagulation Disorders, Inherited/drug therapy"[Mesh] OR "Blood  
Coagulation Disorders, Inherited/therapy"[Mesh] OR "blood coagulation  
disorders"[MeSH Major Topic]) AND ("clinical trial"[Publication Type] OR  
"review"[publication type] OR "Retrospective studies"[MeSH Terms])  
AND "humans"[MeSH Terms] AND "english"[Language] AND  
("hemophilia"[title] OR "haemophilia"[title])

## **Hemophilia – complications**

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### **Embase.com (Elsevier) September 2009**

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Blood clotting disorder/DT (Exp,MJR) AND Hemophilia (Ti) AND Clinical trial (Exp)  
Blood clotting disorder/TH (Exp,MJR) AND Haemophilia (Ti) AND Intervention study (De)  
Blood clotting disorder/DR (Exp,MJR) AND Longitudinal study (De)  
Blood clotting disorder/CO (Exp,MJR) AND Prospective study (De)  
Blood clotting disorder/SI (Exp,MJR) AND Meta analysis (De)  
NOT Thrombocytopenic purpura (Exp)  
Systematic review (De)  
Retrospective study (De)

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(('blood clotting disorder'/exp/mj/dm\_dt,dm\_th,dm\_dr,dm\_co,dm\_si AND  
('hemophilia':ti OR 'haemophilia':ti)) AND ('clinical trial'/exp OR  
'intervention study':de OR 'longitudinal study':de OR 'prospective  
study':de OR 'meta analysis':de OR 'systematic review':de OR  
'retrospective study':de) NOT 'thrombocytopenic purpura'/exp AND  
[embase]/lim)

## ***Hemophilia – complications***

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**CENTRAL (Wiley) September 2009**

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Blood coagulation disorders, inherited/CO (Me) AND Hemophilia (Ti)  
Blood coagulation disorders, inherited/DT (Me) Haemophilia (Ti)  
Blood coagulation disorders, inherited/TH (Me)

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#1 MeSH descriptor Blood Coagulation Disorders, Inherited  
explode all trees with  
    qualifiers: CO,DT,TH  
#2 (hemophilia):ti OR (haemophilia):ti  
#3 (#1 AND #2)

## **Hemophilia – von Willebrand disease**

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PubMed (NLM) September 2009

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Von Willebrand disease/DT (Me) AND Clinical trial (PT)  
Von Willebrand disease/TH (Me) Controlled clinical trial (PT)  
Meta analysis (PT)  
Multicenter study (PT)  
Randomized controlled trial (PT)  
Review (PT)  
Systematic (SB)

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(("von Willebrand Disease/drug therapy"[Mesh] OR "von Willebrand  
Disease/therapy"[Mesh]) AND ("clinical trial"[Publication Type] OR  
"controlled clinical trial"[Publication Type] OR "meta analysis"[Publication  
Type] OR "multicenter study"[Publication Type] OR "randomized  
controlled trial"[Publication Type] OR "review"[Publication Type] OR  
systematic[sb]) AND "english"[Language])

## **Hemophilia – von Willebrand disease**

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### **Embase.com (Elsevier) September 2009**

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Von willebrand disease/DT (Exp)	AND	Clinical trial (Exp)	NOT	Thrombocytopenic purpura (Exp)
Von willebrand disease/TH (Exp)		Intervention study (De)		
		Longitudinal study (De)		
		Prospective study (De)		
		Meta analysis (De)		
		Systematic review (De)		
		Random* (Ti)		
		Control* (Ti)		

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('von willebrand disease'/exp/dm\_dt, dm\_th AND ('clinical trial'/exp OR  
'intervention study':de OR 'longitudinal study':de OR 'prospective  
study':de OR 'meta analysis':de OR 'systematic review':de OR random\*:ti  
OR control\*:ti) NOT 'thrombocytopenic purpura'/exp AND [embase]/lim)

## ***Hemophilia – von Willebrand disease***

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**CENTRAL (Wiley) September 2009**

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Von willebrand disease/DT (Me)  
Von willebrand disease/TH (Me)

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#1 MeSH descriptor von Willebrand Disease explode all trees with  
qualifiers:  
DT,TH

## ***Hemophilia – immune tolerance***

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PubMed (NLM) September 2009

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Immune tolerance induction (TiAb) AND Hemophilia (Ti)  
Tolerance induction (Ti) AND English (LA)  
Immune tolerance (Ti) AND Haemophilia (Ti)  
Blood coagulation disorders, inherited (MJR)

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(("immune tolerance induction"[Title/Abstract] OR "tolerance induction"[Title] OR "immune tolerance"[Title]) AND ("hemophilia"[title] OR "haemophilia"[title] OR "Blood Coagulation Disorders, Inherited"[MeSH Major Topic]) AND "english"[Language])

## ***Hemophilia – immune tolerance***

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### **Embase.com (Elsevier) September 2009**

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Immune tolerance induction (TiAb) AND Hemophilia (Ti)  
Tolerance induction (Ti) Haemophilia (Ti)  
Immune tolerance (Ti) Blood clotting disorders (Exp,MJR)

Limits: English (LA)

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(('immune tolerance induction':ti,ab OR 'tolerance induction':ti OR  
'immune tolerance':ti) AND ('hemophilia':ti OR 'haemophilia':ti OR 'blood  
clotting disorder'/exp/mj) AND [english]/lim AND [embase]/lim)

## ***Hemophilia – immune tolerance***

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### **CENTRAL (Wiley) September 2009**

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Immune tolerance induction (TiAb) AND Hemophilia (Ti)  
Tolerance induction (Ti) Haemophilia (Ti)  
Immune tolerance (Ti) Blood coagulation disorders (Me)

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- #1 (immune tolerance induction):ti,ab OR (tolerance induction):ti OR (immune tolerance):ti
- #2 (hemophilia):ti OR (haemophilia):ti
- #3 MeSH descriptor Blood Coagulation Disorders explode all trees
- #4 MeSH descriptor Blood Coagulation Disorders explode all trees
- #5 (#1 AND (#2 OR #3 ))

## **Abbreviations**

*	Wildcard indicating a variable number of characters (including none)
/CO	Complications (MeSH Subheading or EMTREE disease subheading)
/DM	Disease management (EMTREE disease subheading)
/DT	Drug therapy (MeSH Subheading or EMTREE disease subheading)
/PC	Prevention and control (MeSH Subheading) or prevention (EMTREE disease subheading)
/SI	Side effect (EMTREE disease subheading)
/TH	Therapy (MeSH Subheading or EMTREE disease subheading)
De	Descriptor (EMBASE)
Exp	Explode (EMBASE)
LA	Language
Me	Medical Subject Headings, MeSH (PubMed)
MJR	MeSH Major Topic (PubMed) OR EMTREE Major Focus (EMBASE)
NLM	National library of medicine
PT	Publication type
SB	Subset
Ti	Title
TiAb	Title/Abstract