Treatment Guidelines for Haemophilia in South Africa

Printing sponsored by
National Bioproducts Institute Ltd

Published by:
The South African Haemophilia Foundation
Medical and Scientific Advisory Council
Treatment Guidelines for Haemophilia in South Africa

These guidelines have been compiled by the South African Haemophilia Foundation Medical and Scientific Advisory Council (SAHF MASAC) to facilitate the appropriate management of people with haemophilia (PWH). The current guidelines are based on the publication noted below.


Note to Healthcare Personnel

This booklet is intended as a guide for healthcare personnel who might not be familiar with haemophilia. People with haemophilia (PWH) and their physicians should be advised by a Comprehensive Haemophilia Treatment Centre staffed by a multidisciplinary team skilled in the care of this uncommon chronic bleeding disorder.

Parents of patients with severe haemophilia are usually trained in home infusion of the clotting Factor when their child is about four years old and self infusion is normally accomplished by 12 - 14 years of age. However, infants and boys with mild haemophilia must rely on a Haemophilia Centre or other medical facility for clotting Factor infusions.

Please contact your nearest Haemophilia Treatment Centre (listed on page 24) if you have any uncertainty regarding management.

Acknowledgement is made of all the past and current members of MASAC, who have produced the previous edition, and reviewed the current edition.
Table of Contents

HAEMOPHILIA OVERVIEW 1
TYPES OF BLEEDING IN HAEMOPHILIA 3
Haemarthrosis 4
Muscle & Soft Tissue Bleeding 5
Head Injury - a Medical Emergency 6
Oral Bleeding - Dental / Gum Bleeding / Epistaxis / Tongue 7
Gastrointestinal Bleeding 8
Genito-urinary Bleeds 9
Management of Patients Undergoing Surgery 10
Management of Chronic Symptoms and Target Joints 11
Management of Haemophilia Carrier and Pregnancy 12
Genetics 14
Prophylaxis 15
Management of Pain 16

TREATMENT OF BLEEDING EPISODES
Factor VIII Treatment Guidelines 17
Factor IX - Treatment Guidelines 18
Factor VIII Inhibitor Management Options 19
Rehabilitation Exercises After Joint or Muscle Bleeds 21
Hepatitis 21
Treatment Guidelines for Other Products 22
Haemophilia Treatment Products Available in South Africa 23

HAEMOPHILIA INFORMATION
Haemophilia Treatment Centres and Haemophilia Treaters 24
South African Haemophilia Foundations 29
Haemophilia is an inherited, x-linked, lifelong bleeding disorder which affects males almost exclusively. Most frequently haemorrhage involves joints or muscles. Bleeding patterns differ with age: infants usually bleed into soft tissues or from the mouth but as the boy grows, characteristic joint bleeding becomes more common.

**Haemophilia A** is the most common form of haemophilia and is due to a deficiency of clotting Factor VIII.  
**Haemophilia B** is due to a deficiency of clotting Factor IX.

### Severity

Haemophilia is classified as severe, moderate, or mild according to the levels of circulating Factor VIII or IX and indicates the expected frequency of bleeding:

- **Severe:** Factor VIII or IX < 1%
  
  Factor VIII or IX replacement is needed several times per month for traumatic or apparently spontaneous bleeding may be on regular prophylactic factor therapy.

- **Moderate:** Factor VIII or IX 1 - 5%

  Less frequent bleeding which usually follows trauma, surgery or dental work.

- **Mild:** Factor VIII or IX > 5 - < 40%

  Occasional bleeding, usually only after severe trauma or surgery
**Factor VIII Inhibitors in Haemophilia**

Inhibitors may develop in 10 - 15% of persons with haemophilia A but are much less common in haemophilia B (1 - 3%). Inhibitors are neutralising antibodies that limit the effectiveness of Factor infusions.

Risk Factors for the development of inhibitors:

- severe haemophilia
- family history of inhibitor development
- more frequent in black patients

If an individual is going to develop an inhibitor, this usually happens within the first 50 exposure days after starting Factor VIII replacement therapy.

Inhibitors titres are measured in Bethesda units (BU)

- Low Responders: titre remains below < 5 BU
- High Responders: titre above > 5 BU. The level may increase markedly and rapidly after Factor VIII infusion (may have rapid anamnestic response in 3 days)

**Rules for Inhibitor Management**

1. **Monitor all patients every 3 - 6 months for the development of inhibitors.**
   
   This is particularly important and should be done more frequently in newly diagnosed black patients with severe haemophilia A, who are at greater risk.

2. **Never undertake a surgical procedure or joint aspiration in a person with haemophilia without checking for inhibitors.**

3. **If there is no response to appropriate replacement therapy, test for inhibitors.**

4. **Call a Haemophilia Treatment Centre for advice on patient management.**

Refer to page 19 for Factor VIII inhibitor management options.
TYPES OF BLEEDING IN HAEMOPHILIA

**Major bleeding episodes**
- Central nervous system
- Gastrointestinal
- Neck/throat
- Severe injury
- Hip or iliopsoas
- Advanced joint/muscle
- Forearm compartment

**Important**
- May cause death or deformity. Advice should be sought from a Haemophilia Treatment Centre physician.
- Start appropriate Factor replacement urgently. Hospitalisation is usually required to maintain adequate Factor levels.
- If the patient has an inhibitor, the Haemophilia Treatment Centre must be consulted for major bleeding problems.

**Minor bleeding episodes**
- Joint (early)
- Muscle/soft tissue
- Mouth/gums
- Epistaxis
- Painless haematuria

**Important**
- Although considered minor bleeds, complications may occur. If there are uncertainties about medical management, consult a Haemophilia Treatment Centre.
- Treat early to avoid long term complications.
Haemarthrosis

Commonly affected joints
Knees, elbows, ankles, (shoulder, hip and other joint bleeds are rare).

- After one or several haemarthrosis with synovitis, a joint may become ‘targeted’ for recurrent bleeding and damage.
- These patients should be referred for synovectomy

Symptoms & Signs
- Tingling sensation (early)
- Stiffness
- Pain
- Limited range of motion
- Swelling
- Limp or refusal to use limb

- Do not delay treatment.
- Early bleeding can be felt by the patient before signs are apparent.
- Obvious joint swelling is a late sign of bleeding.

Treatment Guidelines
Replace missing Factor, see: page 17 haemophilia A page 18 haemophilia B

- Treat early.
- Repeat infusions in 12 to 24 hours.
- Ice packs may reduce bleeding:
  Apply 5 minutes on 10 minutes off.
- No circumferential casting.
- X-ray not indicated - only if fracture is suspected.
- Rest the affected joint/limb
  - posterior splint
  - sling for arm
  - no weight bearing
Muscle & Soft Tissue Bleeding

**Dangerous Areas**
- Quadrats femoris (thigh)
- Forearm compartment
- Neck/throat
- Iliopsoas/retroperitoneal
- Popliteal
- Gastrocnemius (calf)

- Aggressive treatment may be indicated. Consult a Haemophilia Treatment Centre for advice.
- Hospitalisation may be required.

**Symptoms & Signs**
- Muscle tightness (early)
- Pain
- Swelling
- Limited range of motion
- Bruising
- Warmth
- Refusal to use limb (young child)

- Bruising may be absent with deep muscle bleeding.

- **Muscle compartment** bleeding (calf, forearm) may cause nerve damage or vascular compromise.

**Treatment**
Replace missing Factor, refer to:
- page 17 haemophilia A
- page 18 haemophilia B
- Ultrasound or CT scan to confirm diagnosis and get baseline clot size.

- Treat early.
- **Repeat infusions** in 12 to 24 hours.
- Ice packs 5 minutes on, 10 minutes off.
- No circumferential casting.
- Rest affected joint/limb
  - posterior splint
  - sling
  - no weight bearing
Head Injury - a Medical Emergency

History
Minor head trauma can lead to CNS bleeding. Unrecognised trauma is common in children. “Spontaneous” bleeds can occur.

• Treat as a major bleed.
• Toddlers and young children are prone to head injury.

Symptoms & Signs
(onset may be delayed)
• Headache
• Vomiting
• Irritability/convulsions
• Lethargy/drowsiness
• Vision disturbance
• Focal neurologic deficits
• Ataxia
• Loss of consciousness
• Paralysis

Any of these with or without a history of trauma is highly suggestive of CNS bleeding. These are late features secondary to mass effect or brain irritation.

NB: Symptoms may mimic a viral infection.

Treatment
1. Urgent Factor replacement - measure level and maintain above 80 - 100% for 7 days. Then maintain plasma factor level at 50% for haemophilia A and 30% for haemophilia B for a further 14 days.
2. CT scan of head
3. Call a Haemophilia Treatment Centre

• If in doubt - treat
• Admit to hospital
• Factor replacement for possible subarachnoid bleeding is indicated even with a normal CT scan.
• For proven intracranial bleeding treatment is required for 21 days.
• Anti-epileptic medication as soon as bleed is confirmed.
Oral Bleeding
Dental / Gum Bleeding / Epistaxis / Tongue

• Bleeding can be profuse
• Swallowing blood:
  - vomiting blood
  - dark tarry stools
• May need Hb and HCT

Treatment

Replace missing Factor, as per minor bleed, refer to:
page 17 haemophilia A
page 18 haemophilia B

- Tranexamic acid
  (Refer to page 22).
- Local pressure.
- Ice 5 minutes on, 10 minutes off.
- If tooth extracted use deep silk or Vicryl suture.
- Red cell transfusion if necessary.

- Torn frenulum (upper lip) bleeding is problematic in the young child.
- Dental injection or extraction requires prior therapy to raise Factor levels.

• Cool, soft, or liquid diet following gum bleeding or dental extraction.
• Tranexamic acid solution:
  give 5 - 10 ml (500 mg / 5 ml) 6-hourly, holding in mouth for 2 minutes before swallowing. Tranexamic acid tablets can also be crushed in warm water before swallowing.
• Continue factor infusion and tranexamic acid until bleeding stops.
Gastrointestinal Bleeding

Potential emergency
- Bleeding can be profuse
- Blood / coffee-ground emesis
- Dark/tarry stools

Abdominal pain - regard as a bleed until proven otherwise: infuse Factor first, then investigate as for all GIT bleeds.

Treatment
- Immediate Factor replacement refer to: page 17 haemophilia A
  page 18 haemophilia B
as per major bleed

- Consult a Haemophilia Treatment Centre.
- Check BP, Hb & HCT.
- Concomitant tranexamic acid therapy.

- Hospitalise to maintain Factor level and monitor ongoing blood loss.
- Investigate for site of bleeding.
- May need red cell transfusion.
Genito-urinary Bleed

- Usually spontaneous

**Signs & Symptoms**
- May have renal angle tenderness
- Red or dark urine
- Usually no dysuria

**Treatment**
- Increase fluid intake (1 glass/hour)
- Bed rest
- If not resolved in 24 hours treat with Factor replacement

Refer to:
- page 17 haemophilia A
- Page 18 haemophilia B
- Continue to increase fluids and bed rest for 7 days

- Persistent or recurrent haematuria should be investigated

- Tranexamic acid is contraindicated
Management of Patients undergoing Surgery

Types of surgical interventions
• Minor surgery, which includes endoscopy, skin biopsy, bronchoscopy, lumbar puncture, dental procedures, etc.
• Major surgery, which includes laparotomy, arthroplasty.

Preoperative assessment and preparation
• Consultation between surgeon, haematologist and blood centre.
• Check FBC, liver function, renal function and inhibitor level.
• Do Factor recovery studies.
• Prepare a written management plan and communicate this to all stakeholders.

Treatment goals
• Raise Factor level to 50 - 80% for minor surgery and 80 - 100% for major surgery.
• Maintain Factor level at 50% for major surgery for at least 7 - 14 days.
• Avoid intraoperative and postoperative blood loss.

Treatment approach
• Haemophilia A:
  • for major surgery, give 40 - 50 IU/kg FVIII
  • for minor surgery give 20 - 40 IU/kg FVIII, 30 minutes before surgery, 6 hours postoperatively and then 12-hourly thereafter.
• Haemophilia B:
  • for major surgery, give 60 - 80 IU/kg FIX
  • for minor surgery 20 - 40 IU/kg, 30 minutes before surgery. Repeat the same dose 6 hours postoperatively and then daily thereafter.
• Factor infusion for major surgery should continue for 7 - 14 days. Venous thromboembolism (VTE) prophylaxis using elastic stockings should be considered in all high-risk surgery.
• Keep peak maintenance Factor level at 50% until healing has started.
• Introduce postoperative rehabilitation and mobilisation gradually under Factor prophylaxis.
• Continuous infusion of Factor with a pump may be used.
• Use of antibiotics postoperatively is mandatory.
• Ensure that patient receives adequate analgesia - NB avoid intramuscular analgesia.
Management of Chronic Synovitis and Target Joints

- Synovitis is the inflamed state of the synovium
- Blood vessels proliferate.
- These vessels are more prone to recurrent bleeding and target joint occurs.

- Chronic synovitis is a condition that persists for 6 months or more.
- There is a predilection for large joints

Treatment

- Secondary prophylaxis (refer to page 15).
- Raise plasma level above 5%.
- Exercises to strengthen joint and muscle.
- Ablation of the synovium using Yttrium Synoviorthesis (only at HCCC).

Procedure for Synoviorthesis:

- To prevent cycle of bleeding commence secondary prophylaxis
- Raise Factor level to 50% prior to procedure
- Give Factor daily x 3 days thereafter
- Intra-articular injection of local anesthetic and steroids given at the same time
- Immobilize for 3 days
Management of the Haemophilia Carrier and Pregnancy

• Check haemophilia carrier baseline Factor level
• Symptomatic carriers managed according to severity of symptoms
• May be treated with DDAVP, Tranexamic acid or Factor replacement

Pre-Pregnancy
• Provide pre-pregnancy genetic counselling to all carriers
• Establish gene abnormality

At Pregnancy
• Take family and personal bleeding history
• Plan management with obstetrician and haematologist
• Measure Factor level at 28 and 34 weeks
• If bleeding do not use DDAVP

• Level done to establish bleeding risk
• Symptomatic carriers should wear MedicAlert® bracelets
• Menorrhagia can be controlled using hormonal, haemostatic or surgical methods

• If not sure of carriers status check sex of fetus
Treatment Guidelines for Haemophilia

**At delivery**
- Plan for vaginal delivery
- Avoid scalp monitor
- Avoid vacuum and forceps
- Take cord blood for Factor levels
- Avoid heel pricks
- Give oral Vit K
- If bleed use Factor replacement

- Watch for bleeding in mother and child
- FIX assay unreliable in new born

**Post Partum**
- Watch for post partum bleeding
- Treat with Factor replacement or DDAVP

- Factor levels fall to baseline in first week
Genetics

Genetic testing for haemophilia A and B is important for:

- Definitive carrier testing
- Prenatal counselling and testing

**Definitive carrier testing:**
All females who are at risk of being haemophilia carriers (mother, sisters, maternal aunts and maternal aunts’ daughters of a person with haemophilia) should be offered genetic counselling and testing, so that their carrier status can be determined definitively. This can be done in early childhood, so that pre-emptive management is possible, but with appropriate consent and genetic counselling.

- Females who are shown to be carriers or high-risk can then be managed appropriately for bleeding complications
- Females who are non-carriers or at low risk would be at very low risk of bleeding complications

**Prenatal counselling and testing:**
Females who are shown to be carriers or high-risk should be offered genetic counselling when they reach child-bearing age to discuss their risks and options for prenatal testing and pregnancy management

**Genetic testing**
- Is complex
- May be done by direct mutation analysis or gene tracking (linked marker) analysis
- May require blood samples from a number of family members (including unaffected individuals)
- Consult with a Genetics Centre to determine from which family members samples are required
## Prophylaxis

- Primary prophylaxis is aimed at stopping spontaneous bleeds in people who have severe haemophilia.
- Secondary prophylaxis is given when there is a high requirement for on demand treatment.
- Single dose prophylaxis is given prior to an event known to cause bleeding.
- Rationale for prophylaxis is to maintain Factor activity above 1% converting a bleeding pattern from severe to mild / moderate.
- Target joints are a high requirement and secondary prophylaxis is used.

### Treatment

- **Haemophilia A:**
  - give 25 - 40 IU/kg
  - 2 - 3 times per week

- **Haemophilia B:**
  - give 25 - 40 IU/kg
  - twice per week
Management of Pain in Haemophilia

- The most effective pain management is early appropriate Factor replacement.
  - Avoid giving analgesic agents for every bleed

Pain may be caused by
- Joint capsular stretching as a result of haemarthroses
- Haemophilia arthropathy
- Compartment syndrome
  - Aim to relieve pain without risk of bleeding

Treatment
- COX-2 inhibitors are favoured due to favourable side effects, good analgesic effects, anti-inflammatory and anti angiogenic effects
  - Aspirin and other antiplatelet agents must be avoided
- Other agents know to relieve pain without increasing bleeding risk are:
  - opiates
  - paracetamol.
  - Analgesia requiring intra muscular injections must be avoided
TREATMENT OF BLEEDING EPISODES

Factor VIII Treatment Guidelines
Refer to page 23 for the products available in South Africa
- Always refer to the Haemophilia Treatment Centre physician’s instructions
- Treatment products may change: always read the package insert
- Patients with inhibitors require special treatment

Factor VIII replacement for Haemophilia A, no inhibitor

Dose depends on bleeding severity
**Minor bleed:** 20 - 40 IU/kg
**Major bleed:** 40 - 50 IU/kg

Expected response: 1 IU/kg = 2% rise in Factor VIII level

**Half life** Factor VIII: 8-12 hr

For serious bleeding **Factor VIII assay** may be required to monitor the response to the infusion.

If there is no response to appropriate replacement therapy, test for inhibitors.

- **The Haemophilia Treatment Centre** physician chooses the most suitable product for each patient. Please follow these recommendations.
- **Plasma-derived Factor VIII** is treated with heat or solvent/detergent to inactivate viruses.
- **Round off** dose to the nearest vial; do not discard excess Factor VIII but rather infuse it.
- **Repeat doses** may be required depending upon the severity of bleeding: always needed for major bleeds every 12 - 24 hours.
Factor IX Treatment Guidelines

Refer to page 24 for the products available in South Africa

- Always refer to the Haemophilia Treatment Centre physician’s instructions
- Treatment products may change: always read the package insert
- Patients with inhibitors require special treatment

Factor IX replacement for Haemophilia B, no inhibitor

Dose depends on bleeding severity

Minor bleed: 20 - 40 IU/kg
Major bleed: 60 - 80 IU/kg

Expected response: 1 IU/kg = 1% rise in Factor IX level

Half life Factor IX: 16-24 hr

For serious bleeding Factor IX assay may be required to monitor the response to the infusion.

If there is no response appropriate replacement therapy, test for inhibitors.

- The Haemophilia Treatment Centre physician chooses the most suitable product for each patient. Please follow these recommendations.
- Plasma-derived Factor IX concentrates are treated with solvent/detergent to inactivate viruses.
- Factor IX Complex [Prothrombin complex concentrate (PCC)] also contain Factors II, VII and X (can reverse the effects of warfarin).
- NB: thrombosis or disseminated intravascular coagulation may occur with frequent or large doses of PCC.
Factor VIII or IX Inhibitor Management Options
Refer to page 24 for the products available in South Africa

Haemophilia A

1. Acute bleeding episodes
   - Ice/cold pack - 5 minutes on, 10 minutes off
   - Immobilise joint with a splint

Low Responder (< 5 BU)
- Give Factor VIII at 2 - 3 times the normal dose
- Monitor response clinically
- Frequent factor recovery levels

High Responder (> 5 BU)
Both APCC and rVIIa are effective for treatment of acute bleeding episodes in patients with Factor VIII inhibitors.
- **Activated Prothrombin Complex Concentrate (APCC)**
  - Dose: 50 - 100 IU/kg q12 - 24h for 3 days or until clinical improvement
  - Infuse at 2 IU/kg/min
  - Do not exceed a single dose of 200 IU/kg
  - **Do not** use antifibrinolytic drugs (e.g. tranexamic acid) concurrently because of the risk of thromboembolism

- **Recombinant Factor VIIa (rFVIIa)**
  - 90 µg per kg q2 - 3 h or by continuous infusion (at 20 µg/kg/hr) until clinical improvement. Factor VIIa activates Factor X and leads to the formation of a haemostatic plug.
  - New single dose of 270 µg/kg may be used
- **Tranexamic acid** 15 - 25 mg/kg/dose po/IV q6- 8h may be used concurrently with recombinant Factor VIIa.

2. Long term Management - Immune tolerance (IT)
- IT should be initiated at a Haemophilia Treatment Centre.
- Successful therapy (eliminating the inhibitor) may take months.
- Several regimens are effective - the Dutch regime (25 IU Factor VIII/kg 3 times per week) is the most affordable.
Haemophilia B

Treatment of haemophilia B with inhibitors

- An aPCC should be carefully monitored for anaphylaxis and anamnestic reaction. Therefore patients with haemophilia B and inhibitors are best treated with rFVIIa, the only bypassing agent that does not contain FIX.

- There is no evidence to guide tolerisation procedures in patients with haemophilia B and inhibitors. Plasma-derived FIX may be used for tolerisation with careful monitoring of anaphylactic reactions

Treatment with rFVIIa:

- Give dose of 90 - 120 µg/kg IV every 2 - 3 hours as bolus or 20 IU/kg/hour as continuous infusion. Single dose of 270 µg/kg may be used.

- Antifibrinolytic can be given concurrently with rFVIIa.
Rehabilitation Exercises After Joint or Muscle Bleeds

Rehabilitation after a bleed is essential to maintain strength and range of motion.

When to start rehabilitation exercises?
As soon as the pain is gone.

What exercises?
1. Static exercise.
2. 3 days after resolution of the bleed: free active exercises where the only resistance is gravity.
3. 10 days after the resolution of the bleed: weight bearing exercises to build up muscle strength and bulk.

Hepatitis

- Any person with haemophilia and related bleeding disorders may have transfusion acquired infection.
  - Test annually for HAV, HBV, HCV.
  - Antibody/antigen negative patients should be immunised, and response assessed.
  - Active infection should be excluded in positive patients.
  - Patients with chronic active hepatitis should be referred to a Hepatologist for management.

NB: Patients with hepatic dysfunction may have other Factor deficiencies (test PT or INR) or a low platelet count.
## Treatment Guidelines for Other Products

### Do not give aspirin

- Aspirin impairs platelet function which may compound an existing bleeding disorder.

### Tranexamic acid

- Antifibrinolytic - prevents clot breakdown.
- Indicated for mucous membrane bleeding.
- Contraindicated in haematuria or with concurrent use of Factor IX complex, or activated PCC.

- 15-25 mg/kg/dose po/IV q6 or 8 hr.

### DDAVP

- Moderate or mild haemophilia A and von Willebrand disease.
- Releases stored Factor VIII and vWF into circulation.
- Less effective with lower baseline Factor VIII level.
- Tachyphylaxis may occur with repeat doses.

- 0.3 µg/kg IV in normal saline over 20-30 minutes
- 0.4 µg/kg SC

NB: Beware of fluid retention and syndrome of inappropriate ADH secretion
Monitor weight and baseline U+E
Restrict fluid as necessary

### Childhood Immunisation

- Avoid other intramuscular injections in haemophilia.

Following the injections:
- press on area > 5 min.
- Apply ice.
- Subcutaneous injection is safer.
# Haemophilia Treatment Products Available in South Africa

## Plasma-derived Factor VIII Products:

<table>
<thead>
<tr>
<th>Product Name</th>
<th>Company</th>
<th>Contact Details</th>
</tr>
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<tbody>
<tr>
<td>Haemosolvate Factor VIII</td>
<td>National Bioproducts Institute NPC</td>
<td>Ronnie Ramphal 031 714 6700 083 229 5339</td>
</tr>
<tr>
<td>Virally Inactivated Factor VIII</td>
<td>Western Province Blood Transfusion Service</td>
<td>021 507 6300</td>
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## Factor IX Complex Products (Prothrombin Complex Concentrates [PCC]):

<table>
<thead>
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<th>Product Name</th>
<th>Company</th>
<th>Contact Details</th>
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<tbody>
<tr>
<td>Haemosolvex Factor IX</td>
<td>National Bioproducts Institute NPC</td>
<td>Ronnie Ramphal 031 714 6700 083 229 5339</td>
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## Activated Prothrombin Complex Concentrate (APCC):

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<tbody>
<tr>
<td>FEIBA</td>
<td>Adcock Ingram Critical Care</td>
<td>Janine Blackensee 011 494 8496 076 538 6532</td>
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## Recombinant Factor VIIa:

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<tr>
<th>Product Name</th>
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<tbody>
<tr>
<td>NovoSeven</td>
<td>Novo Nordisk</td>
<td>Piletso Maniza 083 255 8299</td>
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## Recombinant Factor VIII:

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<tr>
<th>Product Name</th>
<th>Company</th>
<th>Contact Details</th>
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<tbody>
<tr>
<td>Kogenate FS Antihaemophilic Factor 250 IU, 500 IU, 1000 IU</td>
<td>Bayer Schering Pharma</td>
<td>Tracey Tingle 011 921 5633 082 324 9211</td>
</tr>
</tbody>
</table>
Individuals with haemophilia and their physicians should be advised by a Comprehensive Haemophilia Treatment Centre staffed by a multidisciplinary team consisting of a Haematologist or Physician, Paediatrician, Nurse, Geneticist, Physiotherapist, orthopaedic surgeon, dentist, pharmacist and laboratory scientist, who are skilled in the care of this uncommon bleeding disorder.

### Haemophilia Treatment Centres and Haemophilia Treaters

<table>
<thead>
<tr>
<th>Contact person</th>
<th>Hospital</th>
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<tbody>
<tr>
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<td>082 896 3833</td>
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### Pretoria

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<td>Dr A Dlova</td>
<td>Dr George Mukhari</td>
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### Cape Town

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## Treatment Guidelines for Haemophilia

### Haemophilia Treatment Centres and Haemophilia Treaters continued . . .

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### S A Haemophilia Foundations

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Published by:
The South African Haemophilia Foundation
Medical and Scientific Advisory Council

Printing sponsored by
NATIONAL BIOPRODUCTS INSTITUTE Ltd.