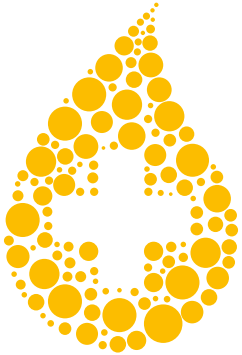
VIII / IX

HAEMOPHILIA IN AFRICA



VIII / IX

HAEMOPHILIA IN AFRICA

MESSAGES AND INFORMATION ABOUT HAEMOPHILIA

IMPORTANT: THE INFORMATION IN THIS NOTEBOOK DOES NOT REPLACE A CONSULTATION WITH A DOCTOR

MESSAGES ABOUT HAEMOPHILIA

VIII / IX

1. HEREDITARY DISEASE

- Transmitted by parents
- Cannot be caught. You are born with haemophilia and have it your entire life.

2. BLEEDING DISEASE

- You bleed longer and the bleeding does not stop
- Joint swelling/haemarthrosis for severe forms
- Spontaneous bleeding or after a surgery, injury or accident
- There are bleeding diseases other than haemophilia

ROLE OF BLOOD COAGULATION TO STOP BLEEDING

BLOOD COAGULATION IS LIKE A SAUCE

VIII / IX

THE SAUCE DOESN'T FORM
WHEN AN INGREDIENT IS MISSING!

WHEN AN INGREDIENT IS MISSING…

Lack of Factor 8 (VIII) Haemophilia A	Lack of Factor 9 (IX) Haemophilia B
Severe : < 1%	Severe : < 1%
Moderate : 1-5%	Moderate : 1-5%
Mild : > 5%	Mild : > 5%

HAEMOPHILIA SEVERITY

VIII / IX

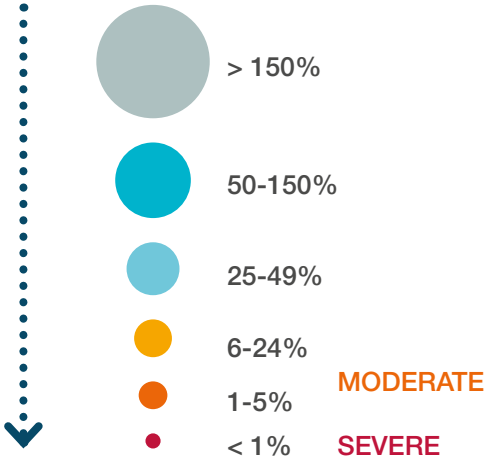
NORMAL
Factor 8 (VIII) and 9 (IX)
50-100%

MILD
Factor 8 (VIII) and 9 (IX)
>5-40%

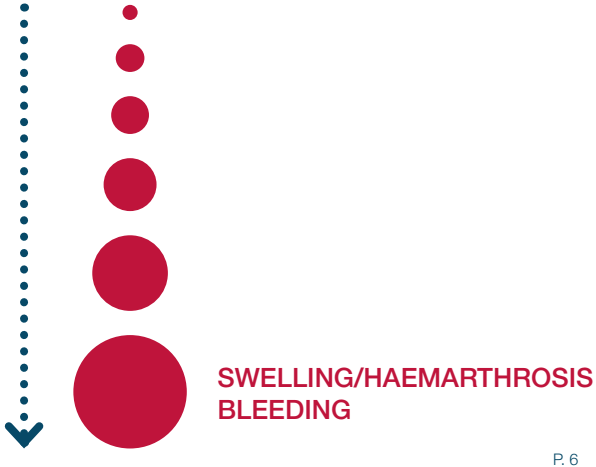
MODERATE
Factor 8 (VIII) and 9 (IX)
1-5%

SEVERE
Factor 8 (VIII) and 9 (IX)
<1%

FACTOR 8 (VIII) OR 9 (IX) LEVELS

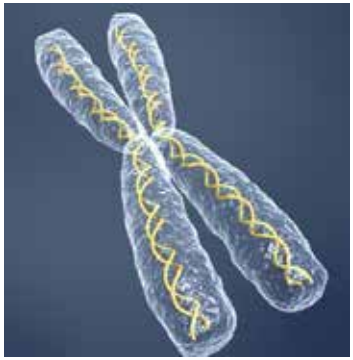


NUMBER OF SWELLING/HAEMARTHROSIS OR BLEEDING



HEREDITY

Chromosome

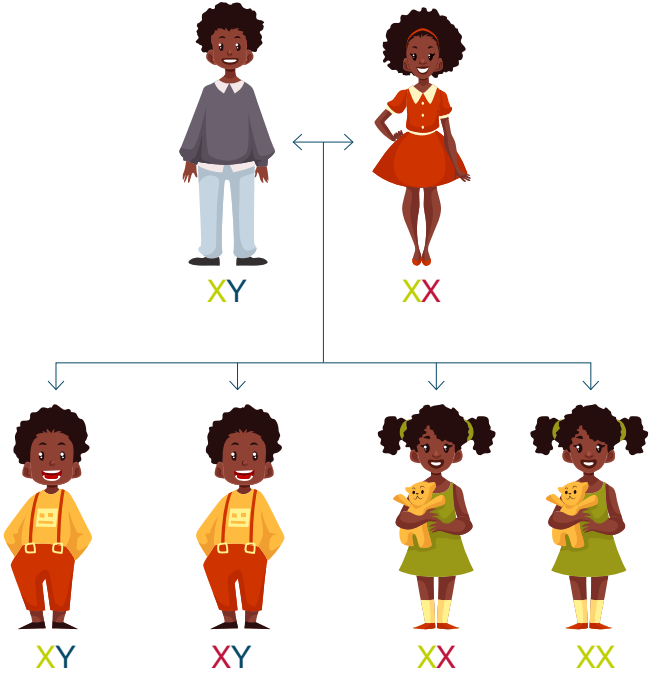


- Heredity information is located in the chromosomes in the form of genes
- Each parent transmits half of their genes to their child (e.g. large ears or large eyes)
- This is random

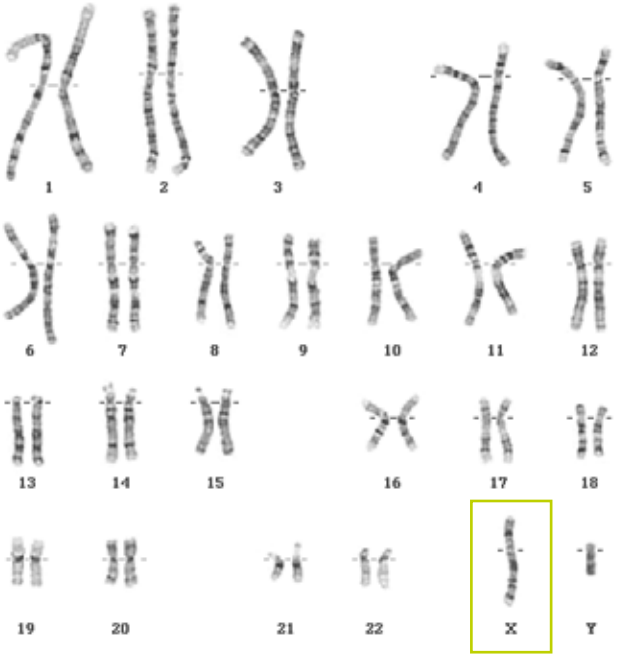
HEREDITARY SIMILARITY →

BOY OR GIRL?

- The father determines the sex of the baby according to the sexual chromosome (X or Y) he transmits.
- The mother always gives an X chromosome to the child.



THE GENE OF FACTORS 8 (VIII) AND 9 (IX) IS LOCATED IN THE X CHROMOSOME



FACTOR 9 (**IX**)

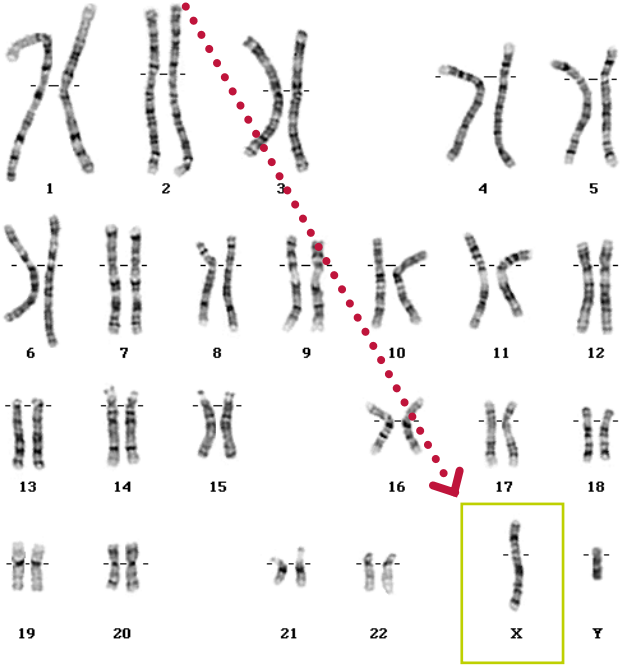
FACTOR 8 (**VIII**)

WHAT CAUSES HAEMOPHILIA?

VIII / IX

Defect
Spelling mistake
Mutation

in the **X** chromosome,
on the factor 8 (VIII) or 9 (IX) gene



IT'S LIKE A RECIPE

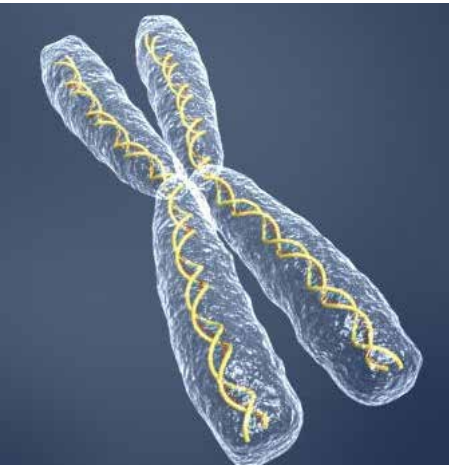
JOLLOF RICE

For the tomato paste :
2 fresh tomatoes, 1 small tatashe (red bell pepper), cored and deseeded, 1 small red onion

For the Jollof rice :
500 g long-grain white rice, 4 tablespoons sunflower or olive oil, 1 red onion, finely chopped, 30 g tomato purée, 500 ml (2 cups) chicken stock or water, 1 tablespoon Ataro Jollof, spice blend, 1/2 teaspoon salt, or to taste, 2 fresh habanero peppers, blended, or to taste, your favourite vegetables for garnish

- If there is an error in your recipe, your dish will taste different.
- If there is an error in the factor 8 (VIII) or 9 (IX) gene, your body will not make enough factor 8 (VIII) or 9 (IX) and you will be affected by haemophilia.

DEFECT
ON THE FACTOR 8 (VIII) OR 9 (IX) GENE



LACK OF
FACTOR 8 (VIII) OR 9 (IX)

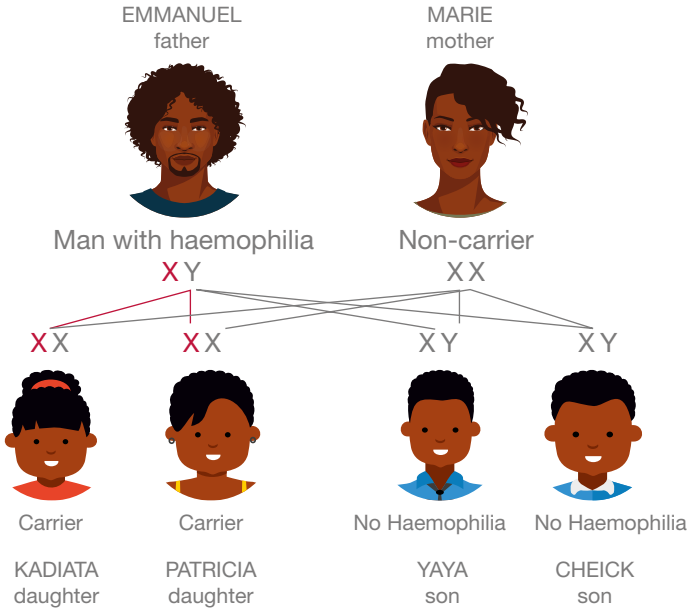
=

HAEMOPHILIA A OR B

HOW
IS HAEMOPHILIA TRANSMITTED?

VIII / IX

TRANSMISSION BY FATHER

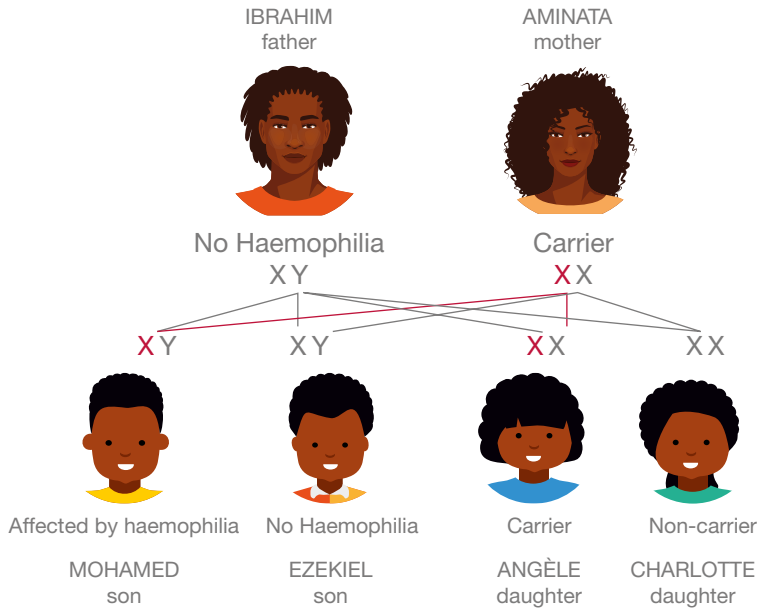


- **ALL** daughters of a man with haemophilia are carriers
- The son of a man with haemophilia son is **NOT** affected by haemophilia

HOW
IS HAEMOPHILIA TRANSMITTED?

VIII / IX

TRANSMISSION BY MOTHER



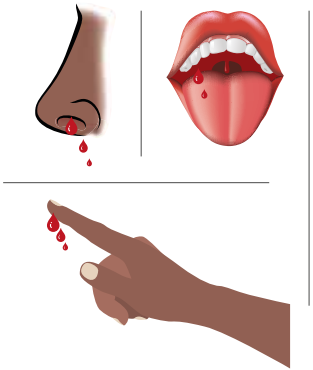
- The son of a carrier has a 50 % chance of being affected by haemophilia
- The daughter of a carrier has a 50 % chance of being a carrier

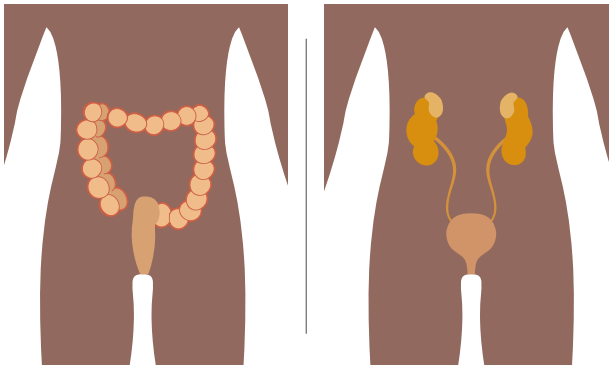
SIGNS OF HAEMOPHILIA

VIII / IX

- Swelling of joints/haemarthrosis
- Swelling in a muscle (haematoma)
- Bleeding from the nose, mouth (gums-teeth), in the urine, mark/bumps on the skin (haematoma)
- Internal/deep bleeding (in the belly, head, etc.)

SWELLING / HAEMARTHROSIS - BLEEDING





CONSEQUENCES OF HAEMOPHILIA

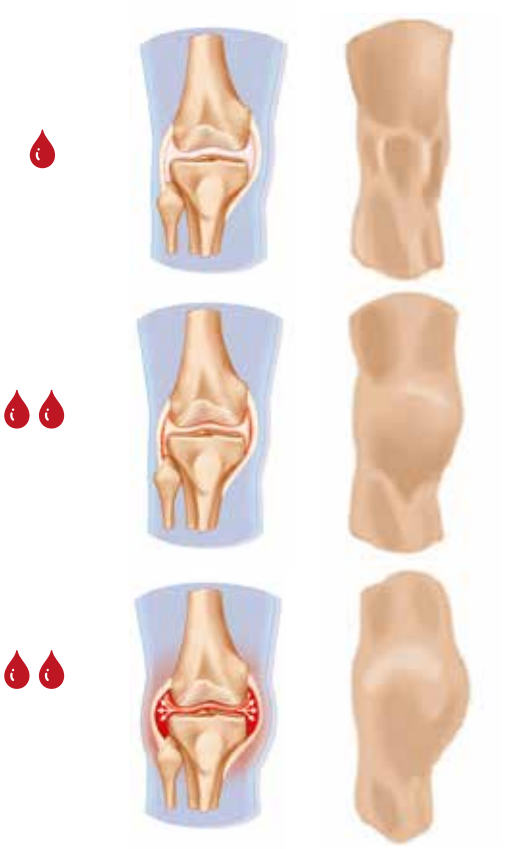
VIII / IX

- REPEATED SWELLING AND HAEMARTHROSIS WILL DESTROY THE JOINTS AND DAMAGE THE MUSCLES
- EXCESSIVE BLEEDING IN CASE OF SURGERY (INCLUDING CIRCUMCISION!)
- ANAEMIA, LACK OF BLOOD
- SOMETIMES DEATH (BLEEDING INTO THE HEAD, AFTER CIRCUMCISION, ETC.)

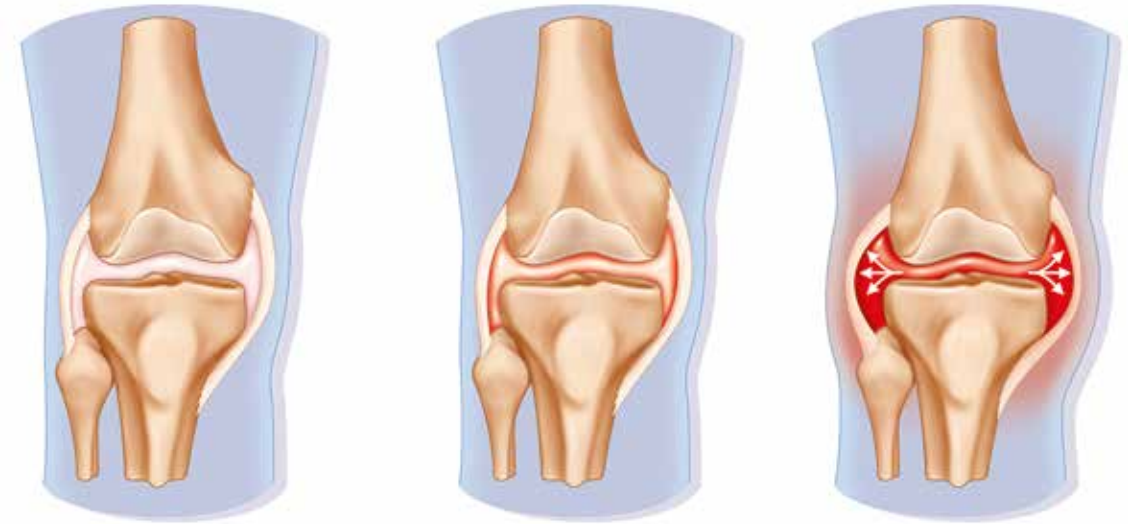
JOINT SWELLING / HAEMARTHROSIS

VIII / IX

- TINGLING
- PAIN
- SWELLING
- BLOCKAGE
- HEAT

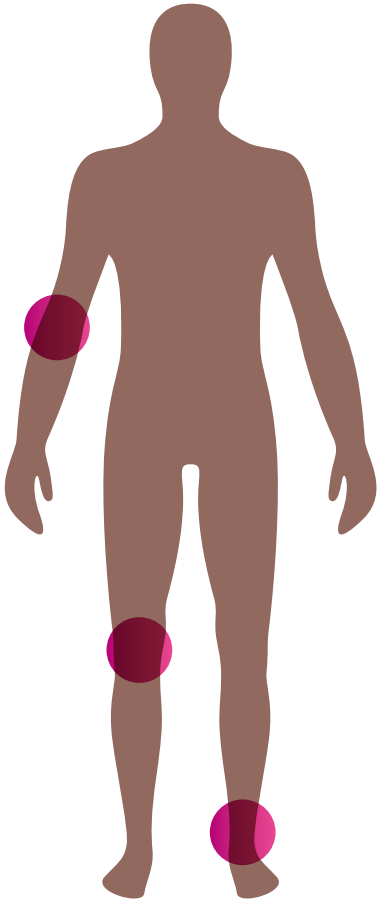


REPEATED SWELLING/HAEMARTHROSIS WILL LEAD TO JOINT DESTRUCTION



THE MOST AFFECTED JOINTS

VIII / IX



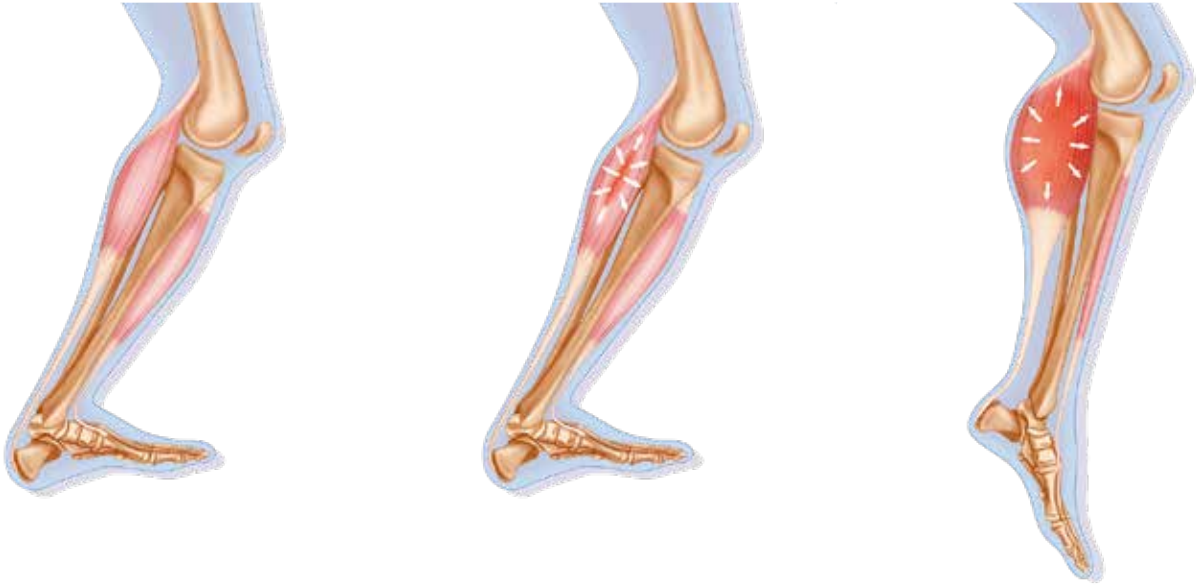
- KNEES
- ANKLES
- ELBOWS

CONSEQUENCES OF JOINT SWELLING/HAEMARTHROSIS

VIII / IX

- PAIN
- DIFFICULTY IN BENDING OR STRAIGHTENING A JOINT
- DISABILITY - LOSS OF MOBILITY

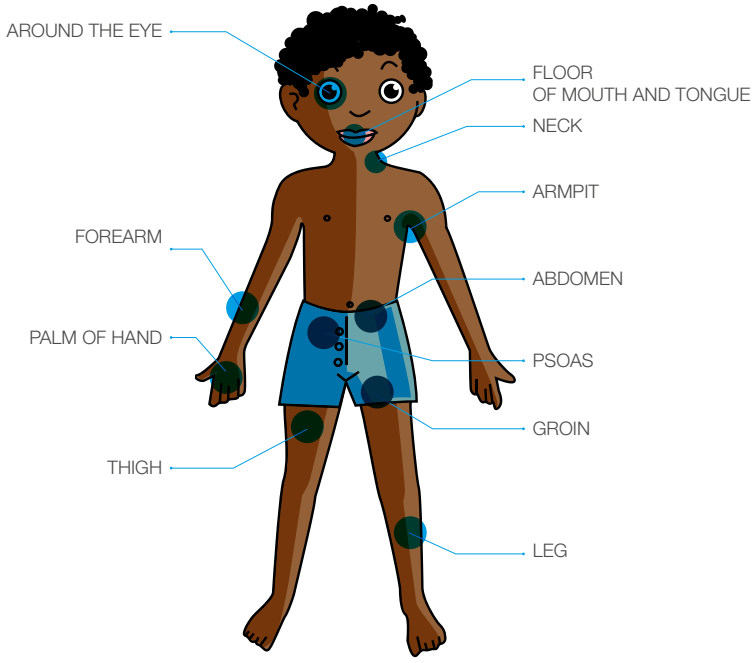
SWELLING OF MUSCLES
MUSCLE HAEMATOMAS

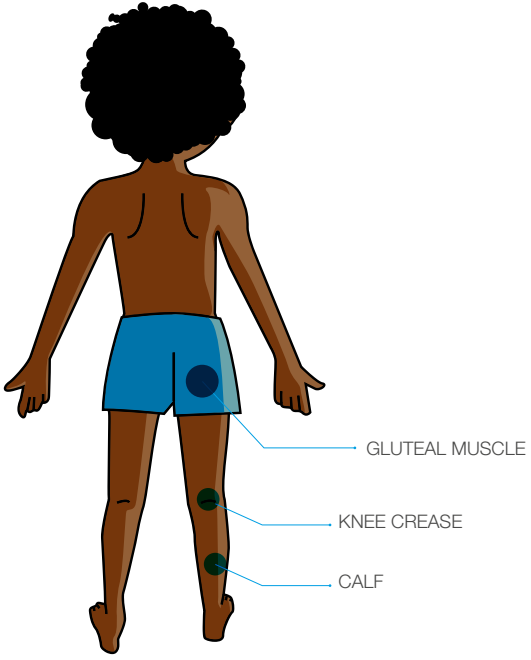


MUSCLE HAEMATOMAS

VIII / IX

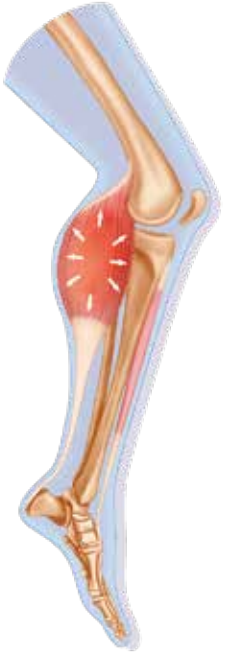
PAIN • SWELLING • MARKS ON THE SKIN



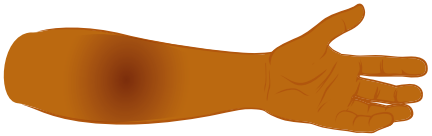


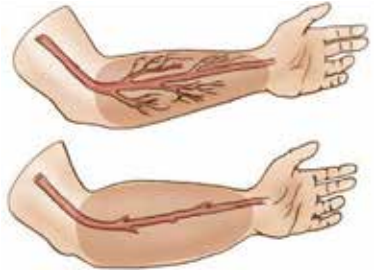
CONSEQUENCES OF MUSCLE HAEMATOMAS PSOAS HAEMATOMA

CONSEQUENCES OF MUSCLE HAEMATOMAS LOSS OF FLEXIBILITY/MUSCLE RETRACTION



CONSEQUENCES OF MUSCLE HAEMATOMAS COMPRESSION OF NERVES AND PARALYSIS





CEREBRAL HAEMORRHAGE

VIII / IX

- HEADACHE
- LOSS OF CONSCIOUSNESS
- PARALYSIS
- TROUBLE SPEAKING/LOSS OF SPEECH
- VOMITING
- DEATH

MAY OCCUR SPONTANEOUSLY
OR
AFTER A BLOW OR FALL TO THE HEAD

HOW DO I KNOW IF I AM AFFECTED BY HAEMOPHILIA?

VIII / IX

- Visit to the doctor at the haemophilia treatment centre
- Track your family history (family tree)
- Blood test to measure the factor 8 (VIII) or 9 (IX) levels
- The blood test can be done at birth

WHEN IS THERE REASON TO SUSPECT HAEMOPHILIA?

VIII / IX

- **IN A BOY WHO HAS**
  - A Bleeding/haemorrhage after circumcision, surgery, dental care, etc.
  - Joint swellings, repeated bleeding (mouth, nose, digestive system, urine, etc.)
  - One or more known person with haemophilia in the family (from the maternal side) (brother, uncle, grandfather, cousin)
  - One or more deaths from bleeding (especially after circumcision) in the family

HAEMOPHILIA TREATMENT

VIII / IX

INJECTION OF CLOTTING FACTOR WITHOUT DELAY IN CASE OF SWELLING/HAEMARTHROSIS/ HAEMATOMA OR BLEEDING

TREAT RAPIDLY
TO AVOID COMPLICATIONS

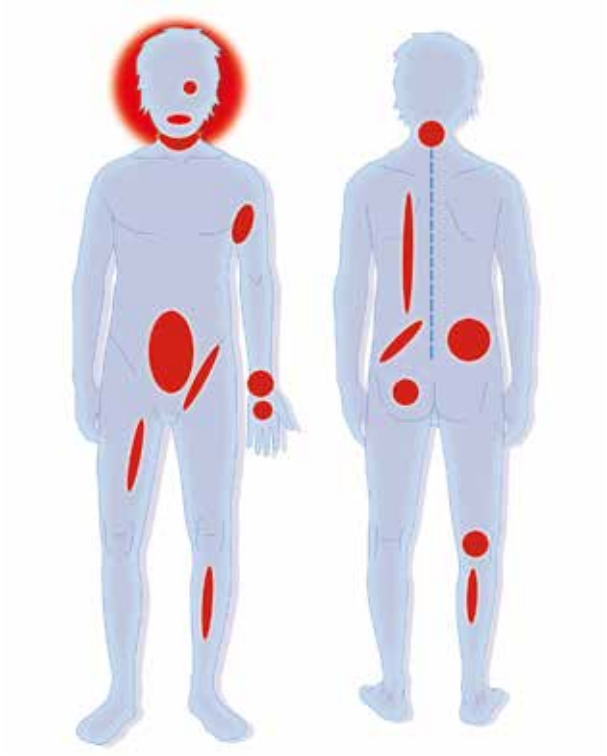
RECOGNISING SIGNS OF SWELLING

VIII / IX

- PAIN
- TINGLING
- HEAT
- SWELLING
- DIFFICULTY IN MOVING A JOINT/LIMB
- CRYING IN CHILDREN

RECOGNISING DANGEROUS SWELLING/BLEEDING

CAUTION! CONTACT THE DOCTOR IMMEDIATELY!



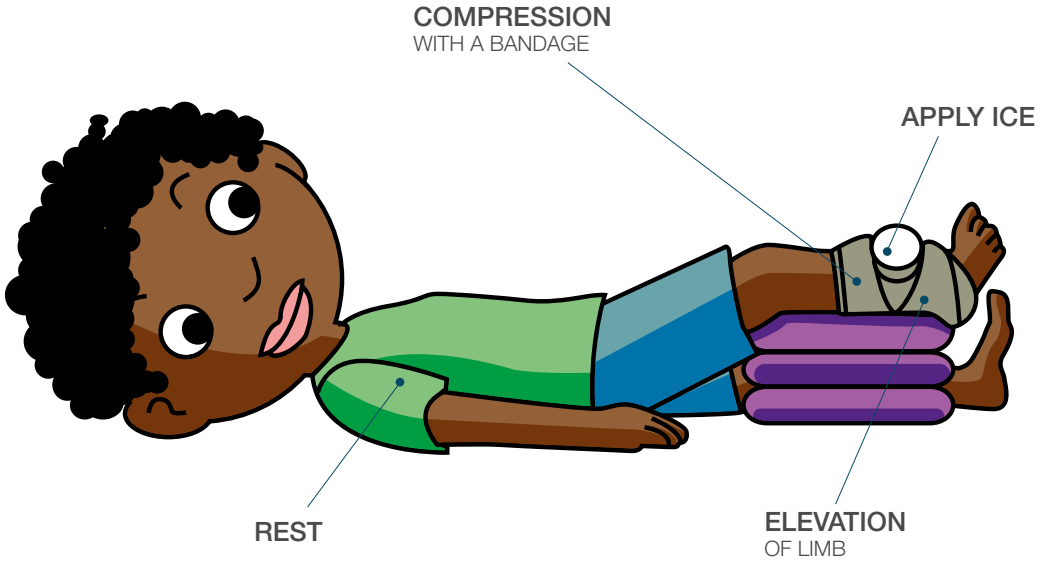
- HEAD (FALL OR BLOW TO THE HEAD)
- NECK
- AROUND OR IN THE EYE
- SPINE
- GROIN - PSOAS
- FRACTURE
- VOMITING BLOOD
- BLOOD IN STOOL

TREATMENT OF JOINT SWELLING/HAEMARTHROSIS

VIII / IX

- IF POSSIBLE, RAPIDLY INJECT CLOTTING FACTORS
- REST, ICE, COMPRESSION, ELEVATION OF LIMB
- PHYSIOTHERAPY (AFTER A FEW DAYS OF REST)
- PAINKILLERS
  - PARACETAMOL/ACETAMINOPHEN
  - COX-2 INHIBITORS (E.G. CELEBREX® OR ARCOXIA® OR EXXIB®)

TREATMENT
OF JOINT SWELLING/HAEMARTHROSIS



PHYSIOTHERAPY

1. Rest for a few days
2. Gradual resumption of movement
   (first without weight-bearing - with a crutches)
3. Physiotherapy sessions

TO FACILITATE WALKING AND REDUCE PAIN

- CRUTCHES
- SPLINT
- ORTHOPAEDIC SHOES
- ORTHOPAEDIC SOLES
- ORTHOTICS

OTHER HAEMOPHILIA TREATMENTS

VIII / IX

- **TRANEXAMIC ACID - EXACYL®** : to be swallowed or used in mouthwash
- **DDAVP - DESMOPRESSIN - MINIRIN®**
- **PAINKILLERS**
  - PARACETAMOL + ACETAMINOPHEN
  - COX-2 INHIBITORS (E.G. CELEBREX® OR ARCOXIA® OR EXXIB®, ETC.)

Buy the medicines in the pharmacy!

HOW CAN YOUR TREATMENT BE IMPROVED?

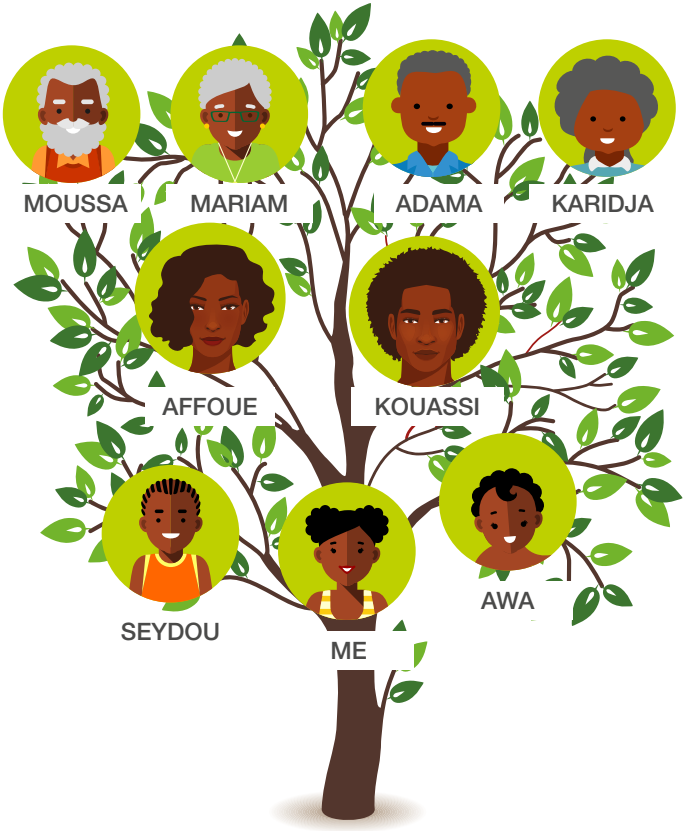
VIII / IX

- Have clotting factors at home
- Inject clotting factors without any delay
- Inject the dose recommended by the doctor
- Fill in your patient diary
- Contact the haemophilia treatment centre quickly if there is no improvement
- Parent or patients are able to inject themselves coagulation factors

REGULAR FOLLOW-UP AT HAEMOPHILIA TREATMENT CENTRE

- Physical examination
- Look for other cases of haemophilia in the family (screening)
- Check of joint condition by the physiotherapist
- Treatment recommendations
- Inform the haemophilia centre if a baby boy is born in the family

FAMILY TREE → SCREENING



HAEMOPHILIA TREATMENT

VIII / IX

→ PREVENTION

- Have a haemophilia identification card
- Fill in your patient diary
- Do not hide swelling or bleeding (children)
- Inform the haemophilia centre if a baby boy is born in the family

WFH
WORLD FEDERATION OF HEMOPHILIA

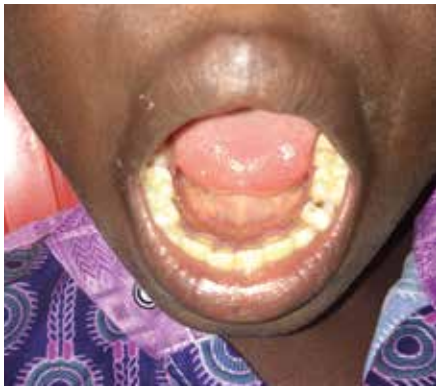
HAEMOPHILIA TREATMENT

→ PREVENTION

- No surgery, circumcision or dental care without treatment
- No intra-muscular injection (subcutaneous vaccine)
- No aspirin
- No anti-inflammatory drugs (except COX-2 inhibitors, such as Arcoxia®, Celebrex® and Exxib®)

→ PREVENTION

- Visit to the dentist
- Dental hygiene (brush your teeth every day!)



HAEMOPHILIA TREATMENT

→ PREVENTION

- Sport or appropriate activity
- Regular physiotherapy

IN CASE OF SURGERY

VIII / IX

- **Visit to the haemophilia treatment centre BEFORE surgery to:**
  - Confirm the haemophilia diagnosis if necessary
  - Ensure the absence of inhibitor
  - Establish a treatment plan with clotting factors
    - Product type
    - Dose
    - Duration
- **Close and specialised follow-up AFTER SURGERY**

IN CASE OF CIRCUMCISION

- Visit to the haemophilia treatment centre and confirmation of diagnosis BEFORE circumcision
- Circumcision at the hospital
- Circumcision while using a coagulation factor

INHIBITORS

VIII / IX

- The body sometimes produces antibodies that make the clotting factors ineffective or less effective
- These antibodies are called inhibitors and are detected with a blood test
- Treatment of bleeding is difficult and requires management by the haemophilia treatment centre

PATIENTS ASSOCIATION OF HAEMOPHILIA AND OTHER BLOOD DISEASES

WFH
WORLD FEDERATION OF HEMOPHILIA

- IDENTIFICATION CARD
- REGISTER
- INFORMATION
- ADVICE
- SUPPORT

VIII / IX

CARRIERS OF HAEMOPHILIA

BEING A CARRIER MEANS…

VIII / IX

- Raising one or more sons affected by haemophilia
- Perhaps having heavy/abnormal bleeding
- Knowing that there is a risk of having a son affected by haemophilia and having a daughter who is a carrier

IMPORTANT FOR CARRIERS

- Screen for carriers in the family of a person with haemophilia
- Inform carriers about the transmission of haemophilia
- Educate mothers to best treat their son(s) affected by haemophilia
- Identify and treat carriers at risk of bleeding

VIII / IX

TESTING CARRIERS

WHO ARE THE CARRIERS?

VIII / IX

MANDATORY

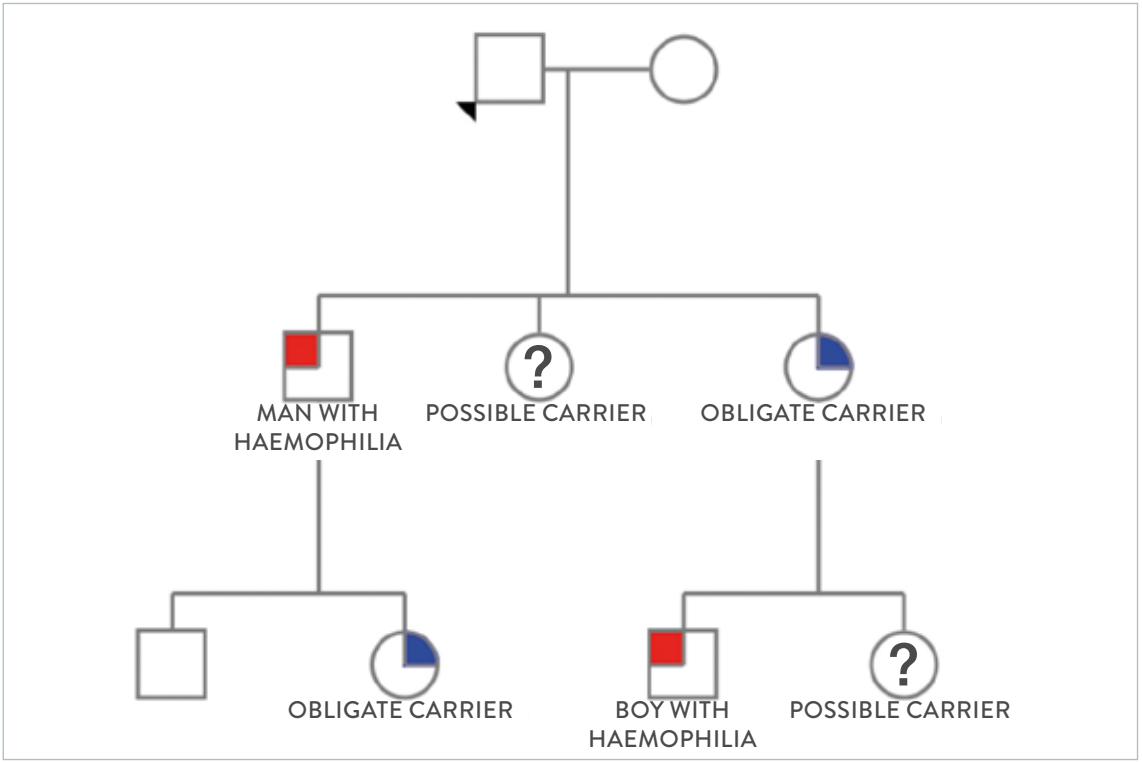
- The daughter of a man with haemophilia
- The mother who has
  - Two sons with haemophilia

    OR
  - One son with haemophilia and another person with haemophilia in the family on the mother's side (brother, uncle, cousin, etc.)

POSSIBLE

- The sister of a person with haemophilia
- The sister of a carrier
- The mother of a person with haemophilia without other known haemophilia cases in the family

FAMILY TREE EXAMPLE



HOW ARE CARRIERS IDENTIFIED?

VIII / IX

- **FAMILY TREE**

  Easy - Free

- **FACTOR 8 (VIII) OR 9 (IX) ASSAY**

  Laboratory specialised in blood clotting (at the haemophilia treatment centre)

- **GENETIC TESTING**

  Very expensive - Laboratory specialised in genetics

MEASURING FACTORS 8 (VIII) AND 9 (IX) IN CARRIERS

VIII / IX

- **WHY?**
  - Some carriers have less factor 8 (VIII) or factor 9 (IX) and may bleed more
    - After surgery, childbirth or an accident
    - Heavy menstrual periods
    - Bleeding of the nose, gums, etc
  - If factor 8 (VIII) and 9 (IX) levels are known in the carriers, precautions can be taken to prevent bleeding

PREVENTION OF BLEEDING

VIII / IX

- **WHEN?**
  - During menstruation
  - In case of bleeding/haemorrhage (nose, gums, etc.)
  - In case of surgery
  - At time of delivery or miscarriage
  - Dental care

- **HOW?**
  - TRANEXAMIC ACID (Exacyl®)
  - DDAVP - DESMOPRESSIN - Minirin®
    - Only for carriers of haemophilia A
    - Test to confirm the efficacy of DDAVP
  - Sometimes injection of clotting factor

DDAVP Test

MODE OF HAEMOPHILIA TRANSMISSION

VIII / IX

- The son of a carrier has a 50 % chance of having haemophilia
- The daughter of a carrier has a 50 % chance of being a carrier
- The risk is the same with each new pregnancy.
- In doubt, a potential carrier is considered as being a carrier

PREGNANCY IN CARRIERS

- Inform the gynaecologist
- Contact the haemophilia treatment centre
- Hospital delivery
- Forceps and vacuum extraction should be avoided

IF THE BABY IS A BOY

VIII / IX

Take a blood test to check for haemophilia (BEFORE circumcision)

- To avoid bleeding and death after circumcision
- To treat swelling/haemarthrosis and other bleeding from a young age
- To avoid complications

TAKING CARE OF A SON WITH HAEMOPHILIA

VIII / IX

- Recognise the signs of swelling/haemarthrosis/bleeding
- Treat quickly to avoid complications
- Regular follow-up at the haemophilia treatment centre
- Visit to the dentist
- No surgery - circumcision without advice of the haemophilia treatment centre

SCHOOL AND HAEMOPHILIA

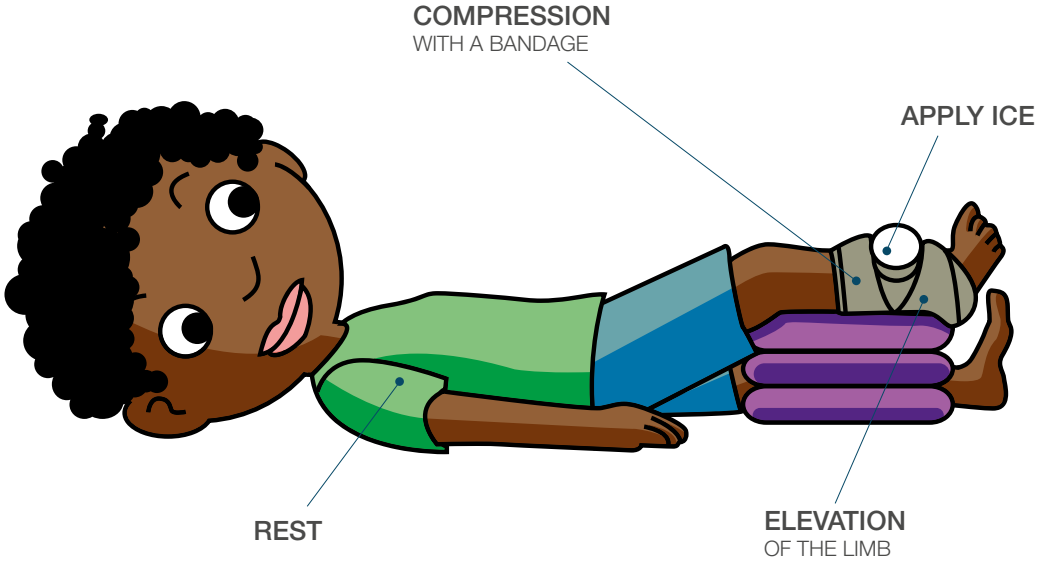
- Children with haemophilia have normal intellectual skills
- They should attend school regularly
- Notify the teacher, professor or school director about haemophilia
- Do not systematically exclude the child from participating in all activities
- Appropriate activities, non-violent games

IMPORTANT INFORMATION

VIII / IX

IF A JOINT SWELLING/HAEMARTHROSIS OCCURS

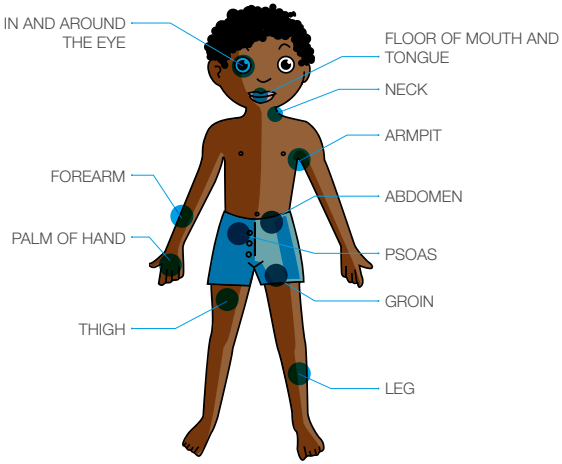
- The clotting factors should be administered without any delay

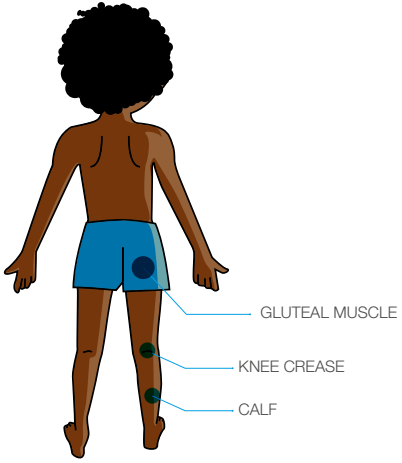


IMPORTANT INFORMATION

VIII / IX

- Swelling or bleeding in these areas may be dangerous
- Clotting factors should be injected and the doctor should be contacted immediately





This booklet was written by Dr Catherine Lambert in collaboration
with the Yopougon University Hospital Haematology and Physical Medicine Department

Thanks to the patients for participating in this project!

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