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## Factor VII Deficiency Frequently Asked Questions

Patient information

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# Dear Patient, Dear Parents,

You or your child have been diagnosed with a deficiency of factor VII (meaning factor 7, with 'VII' the Roman numeral for 7). This diagnosis may have worried you and raised a lot of questions. This booklet intends to give you information about factor VII deficiency and provide answers to your most important questions. Your doctor will be glad to answer any additional questions.



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Doctor's name

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Hospital

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Phone number

Stamp

# Factor VII deficiency

## What is factor VII deficiency?

Factor VII deficiency is a rare hereditary blood clotting disorder, in which a coagulation factor in the blood is reduced. Factor VII (FVII) deficiency is the most common rare inherited bleeding disorder. Severe factor VII deficiency occurs in approximately 1 in 300,000 to 500,000 people.<sup>1-5</sup> FVII deficiency was first described in 1951 by Dr Benjamin Alexander from Boston, USA.<sup>6</sup>

## Is FVII deficiency dangerous?

Many people with FVII deficiency have few problems throughout their lives. Congenital FVII deficiency occurs in various severities: from no signs to a strong bleeding tendency. As bleeding may be heavy in some situations, for instance, when you have an accident or during surgery, it is important for you

to be well informed about your condition and tell your doctor about your FVII deficiency immediately. This is also the reason why you are issued an emergency health card, which you should carry with you at all times.

## What is a coagulation factor?

When a person sustains a bleeding injury, the coagulation system begins to work in order to close the bleeding site and avoid further blood loss. For this purpose, the human blood contains various substances (consisting of protein) known as coagulation factors. The coagulation factors can promote or prevent coagulation.

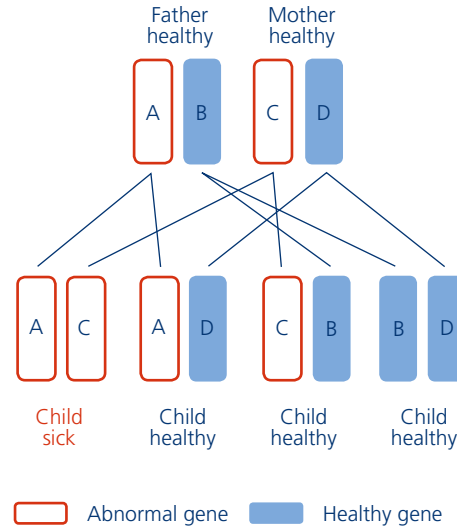
# Degrees of severity and inheritance

## Are there different forms or degrees of severity of FVII deficiency?

Yes. A distinction is made between mild, moderate and severe FVII deficiency based on the amount of FVII present in the blood and its efficiency, referred to as FVII residual activity. In mild and moderate cases, the affected individual may in fact never have any problems with bleeding.

## How do people get FVII deficiency?

FVII deficiency is an inherited disorder that is passed on from parents to their children. FVII deficiency affects men and women in equal numbers.<sup>7</sup> For a child to be born with FVII deficiency, both parents need to carry the abnormal gene. As shown in the figure, for these individuals, the risk of a child inheriting factor FVII deficiency is 25%.<sup>8</sup>



# Bleeding tendency

## When do I have an increased tendency to bleed?

The tendency to bleed is dependent on 'coagulation activity', meaning how well the blood clots. If 'coagulation activity' is reduced, there will be a tendency to develop bruises (haematomas) without any obvious cause and an increased risk of bleeds with heavy blood loss. A strong bleeding tendency is also given in the case of accidents and surgery as well as for the mother during childbirth and for women during menstruation.<sup>1,9</sup>

## Do women have a higher bleeding tendency?

In young women with FVII deficiency, the first menstrual period may be longer and heavier than normal. This holds also true for menstrual periods in general. Please note that the regular, heavy blood loss may gradually lead to iron deficiency resulting in anaemia. It is therefore important to ensure that your iron intake is adequate. If not, you may need to take an iron supplement.<sup>1</sup>

## Which types of bleeds occur frequently?

In addition to the heavy menstrual bleeding and bleeding during surgery already mentioned, the most common types

of bleeding occurring in people with severe FVII deficiency are mucosal bleeding (bleeding from the nose or gums), bleeding into the muscles and joints, bleeding in the gastrointestinal tract, blood in the urine and bleeding in the central nervous system.<sup>1,9</sup>

## What should pregnant women be aware of?

Pregnancies in women with FVII deficiency are often trouble-free. However, heavy bleeding may occur during childbirth in women with pronounced FVII deficiency, placing the baby and the mother at risk. Depending on the severity of the condition, it may be necessary to administer an FVII-containing product before or during delivery. If bleeding occurs during pregnancy, it is also possible to reduce the bleeding tendency with prophylactic administration of the coagulation factor.

The decision as to whether an FVII-containing product should be administered to a patient with FVII deficiency during pregnancy must always be taken by an experienced physician (coagulation disorder specialist). The benefit of treatment in terms of a reduced bleeding tendency must be weighed carefully against a possibly increased risk of developing blood clots (thrombosis) during pregnancy.



## Emergency situation

### Does every patient with FVII deficiency get an emergency health card?

Yes. Even if your FVII deficiency is mild, you will receive an emergency health card. In many cases, the bleeding tendency is not clearly foreseeable. A potential increased bleeding tendency depends on the type and severity of the planned procedure, on the residual FVII activity, as well as on personal particulars such as the type of inherited FVII gene mutation and the patient's history. Therefore, it is important for you to carry your emergency health card with you and inform your attending physician about your FVII deficiency.

European emergency health card ►





# Diagnosis



## How can FVII deficiency be diagnosed?

Congenital FVII deficiency is often a random finding. This means that conspicuous laboratory blood findings are observed as part of a general check-up or before an operation, without the presence of bleeding. Other patients first notice a tendency for bleeding or bruising that leads to a doctor identifying an FVII deficiency.<sup>4</sup>

## What type of blood test is used to determine FVII deficiency?

FVII deficiency is suspected when a specific laboratory value is out of normal range (prothrombin time (PT) is prolonged or 'Quick' value is too low). A DNA test used to analyse the FVII genetic material allows the underlying genetic mutation to be determined.<sup>10,11</sup>





### What does an analysis of FVII genetic material reveal?

In patients with FVII deficiency, bleeding tendency can vary widely. On the one hand, genetic analysis is used to find the genetic mutation responsible for the FVII deficiency. On the other hand, genetic testing of many people affected and the accompanying documentation of bleeds in these patients can lead to a better understanding of the disease and eventually, one day, to the predictability of a bleeding tendency due to specific gene mutations.<sup>10–12</sup>



# Treatment

## Is there any treatment for my FVII deficiency?

Yes, there is. Treatment entails replacing the missing coagulation substance, a treatment known as replacement therapy.<sup>4</sup> The coagulation substances must be injected into a vein; they cannot be taken orally. Depending on the residual activity, coagulation factor may have to be administered, possibly as a prophylactic treatment, if bleeding is anticipated (surgery). Some patients with FVII deficiency will never need coagulation factor VII replacement throughout their lives.

## What should I do in the case of bleeding?

You should always go to hospital.

## What kind of substances can be used to treat FVII deficiency?

There are two options for replacing the missing FVII. The first is a product prepared by means of genetic engineering. Such products are referred to as 'recombinant'. They are free from components from human-donated blood and thus safe

from transmitting infections with foreign viruses or bacteria. The second possibility is the production from human blood donations; the product is plasma-derived.<sup>4,13</sup>

## Are there any risks associated with the administration of coagulation substances?

Two important risks must be kept in mind: on the one hand, the risk that coagulation will be too strong, leading to an undesired blood clot (thrombosis), and on the other hand, the risk of developing antibodies to the coagulation factor administered, known as inhibitors. Both the risk of thrombosis and the possibility of inhibitors developing are very low in patients with FVII deficiency, however.<sup>2,4,13,14</sup>

# Managing everyday life



## Can a child with FVII deficiency take part in preschool and school activities?

Yes, they can. FVII deficiency does not usually interfere with everyday life. Nevertheless, you should inform your child's teachers about your child's FVII deficiency, so that they can react properly in the case of an emergency. Your child can also participate in school sports and do sports in general.

## What should be kept in mind during second dentition or dental extraction?

If an individual is known to have a strong bleeding tendency in the case of injuries, prophylactic administration of a product containing FVII should be considered before dental work is performed. If a child with FVII deficiency has frequent bleeds during second dentition (emergence of permanent ('adult') teeth), prophylactic treatment with a coagulation factor should be discussed with a coagulation expert.

## Can I take my medication along when travelling?

Ask your doctor for a customs certificate, which will enable you to take your medication with you without trouble when travelling.



# CUSTOMS CERTIFICATE

for medication

GB

To whom it may concern,

The following patient has haemophilia and can only control his/her bleeding with the help of factor concentrates.

Mr/Ms

Patient name

who has

e.g. Haemophilia A or B

is travelling to

The above-named person cannot start his/her journey without taking factor concentrates. He/She is carrying sufficient medication for autoinfusion and additional quantities for emergencies. Accidental confiscation/seizure of the factor concentrates could put the life of the above-named person at risk because this medication is essential for him/her.

Thank you for your co-operation.

Yours sincerely

Signature, name and title of the attending physician

# MEDICAL CERTIFICATE

for presentation to the authorities

GB

Mr/Ms

Is suffering from the blood disease called:

i.e. a clotting factor deficit,

I know the patient personally. In case of bleeding he/she responds well to prompt autoinfusions of factor concentrates. Preferably of:

Name of the concentrate

You can safely follow the instructions given by Mr/Ms

Name of the patient

concerning when and what quantity of factor concentrates are to be administered.

Yours sincerely

Signature, name and title of the attending physician

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Changing haemophilia. Improving quality of life.

Nutrition  
Physiotherapy  
Training  
Pain  
Travel

Novo Nordisk established HaemCare™ to support people with haemophilia, their families, as well as their attending physicians, carers and therapists.

HaemCare™ is part of the international Changing Haemophilia® initiative.

With HaemCare™, we offer a comprehensive range of services and are available to answer your questions or provide suggestions at any time.

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**Novo Nordisk Pharma GmbH, Brucknerstraße 1, 55127 Mainz, Germany**

Tel.: +49 (0) 6131-9030, Fax: +49 (0) 6131-903 1370, [www.novonordisk.de](http://www.novonordisk.de)

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