Acquired haemophilia

Patient brochure

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Dear patient,

This brochure deals with acquired haemophilia. The clinical picture relates to one of the rare disorders associated with blood clotting and as such, it has to be treated acutely as soon as bleeding occurs. Special medications are available for rapid haemostasis. In the long run, the aim of the treatment is to completely eliminate the cause of the disease, the coagulation factor inhibitor. On the following pages you will learn more about the cause of the disease, its diagnosis and treatment options.

We would like to thank **Dr. med. Ute Scholz**, at the Zentrum für Blutgerinnungsstörungen Leipzig, as well as MVZ Labor Dr. Reising-Ackermann and colleagues for their professional support.

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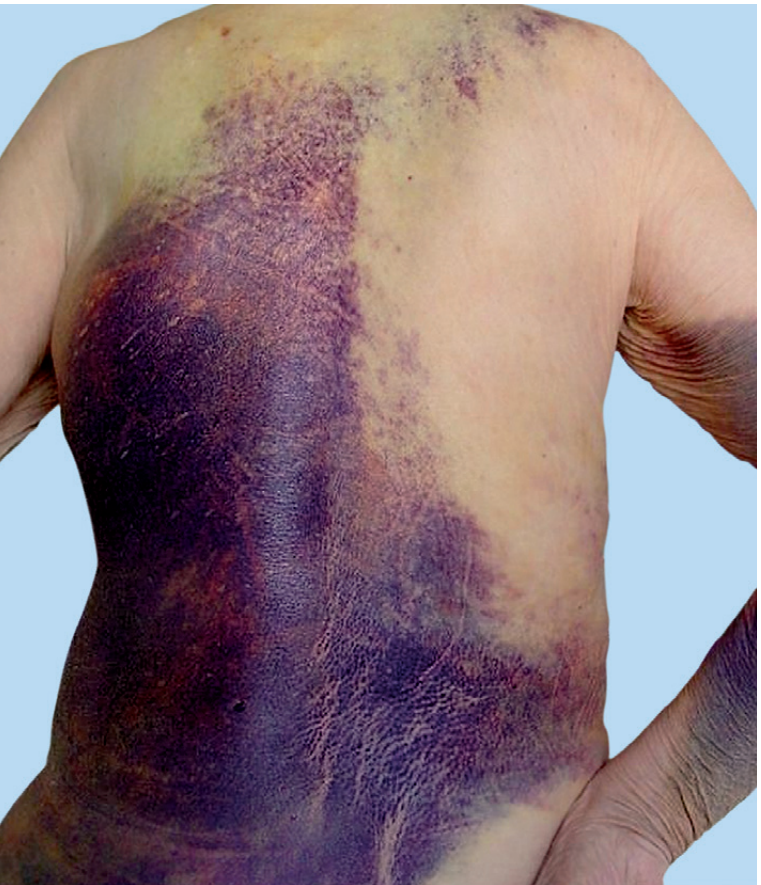
Acquired haemophilia

What is the coagulation system?
The human body is able to both stop bleeding wounds and prevent the clogging of blood vessels (thrombosis or embolism). This is due to the platelets and various proteins, the so-called coagulation factors. Among other things, these are referred to as Roman figures I to XIII.

What is an inhibitor?
If the immune system is no longer able to distinguish between structures within and outside the body, it increasingly produces inhibitors (= antibodies) that incorrectly attack structures within the body. This is also the case with acquired haemophilia: Inhibitors are formed against the body's own coagulation factors – these are most often against factor VIII. As a result, their effect is overridden and blood clotting is blocked.

The development of inhibitors due to an uncontrolled immune system may occur in connection with certain diseases or circumstances, e.g. rheumatological diseases, tumours, inflammatory bowel diseases, severe infections or pregnancy.

Diagnosis



What are the typical signs of the disease?
The disease manifests itself in the tendency to bleed in different ways. Extensive bleeding in the skin and muscles is particularly common, with no apparent cause. Unexplained bleeding can also occur after surgery. Very often, patients need to receive bottled blood. If diagnosis and treatment are not performed quickly, the disease can lead to serious complications.

Which laboratory findings confirm the diagnosis?

In the event of unexplained bleeding and the occurrence of prolonged aPTT*, the physician must assume an inhibitor against FVIII – unless the prolonged aPTT has other causes (medicines for blood clotting, e.g. heparin or special antiphospholipid antibodies).

Confirmation is carried out following a diagnosis of the reduced coagulation factor by means of a special test for an antibody against factor VIII. Various laboratory tests are available for this purpose. The activity of the inhibitor in the blood is usually indicated in special units (Bethesda units).

* aPTT (activated partial thromboplastin time) is a laboratory screening for congenital or acquired coagulation disorders.

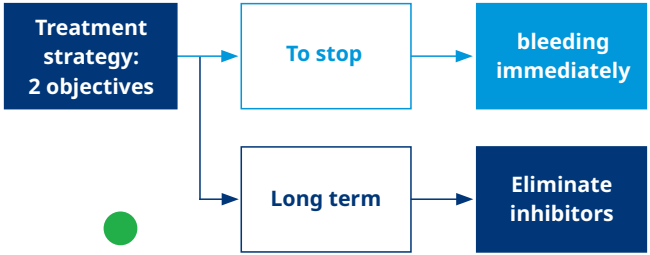
Therapy

How is bleeding treated?

When bleeding occurs, everyone involved must act quickly. This is because the patient needs to receive fast-acting medication. Various substances are available to restore the coagulation function. Coagulation factor preparations are usually used.

What are the options for eliminating inhibitors?

The aim of the therapy is to completely remove the inhibitor to factor VIII. This can be done in different ways, e.g. through medication to suppress the immune system, by washing the antibody out of the blood or by administering a preparation to destroy the inhibitor. If treatment is not successful, a combination of other treatment options is considered.



In short

1. Acquired haemophilia is a rare disease.
2. If bleeding occurs, apply emergency therapy immediately.
3. The aim of the treatment is the complete removal of the inhibitor.
4. Men and women can be equally affected by acquired haemophilia.

What's the latest?

haemcare.de is your portal for everything you need to know about blood coagulation disorders, sports, nutrition, travel … and what's happening right now.

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