



PNH AND THE COMPLEMENT SYSTEM

The cause of hemolysis in PNH can be complicated. This resource will help simplify the process by illustrating how the part of your immune system called the complement system plays a role. Understanding your disease and the complement system can be an important part in feeling more in control.

Learn more on the following page





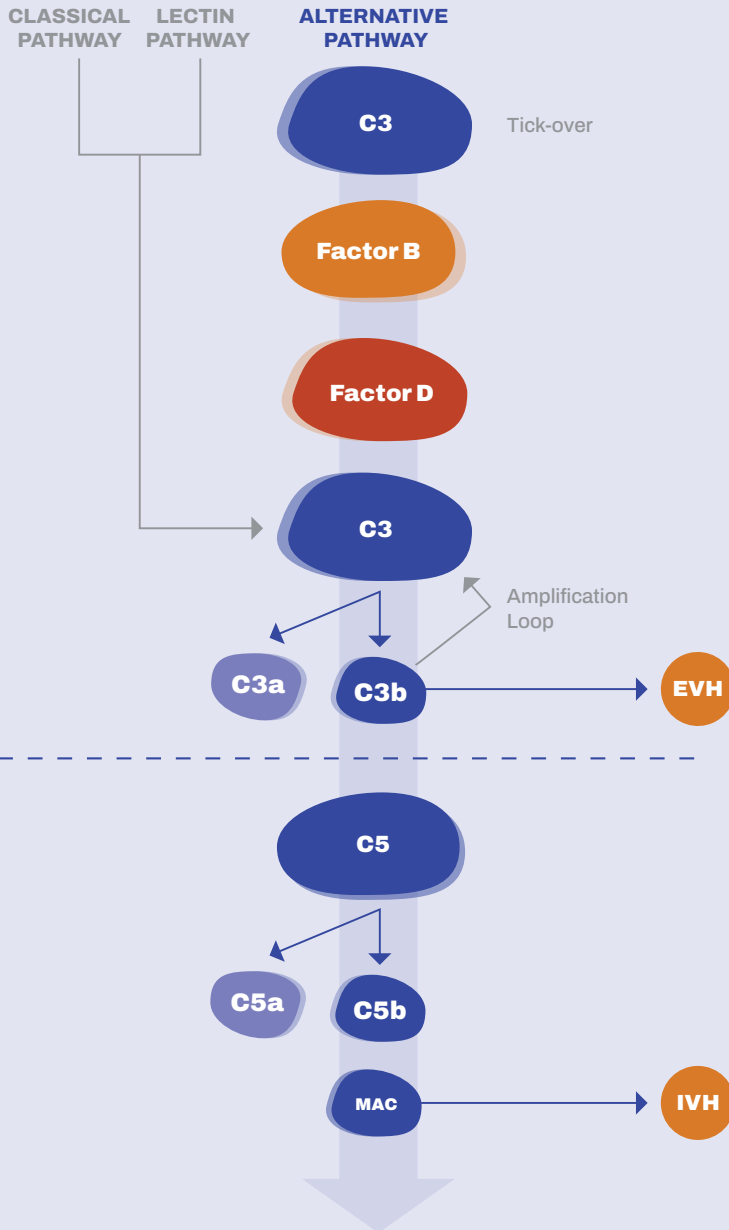
With PNH, some red blood cells (RBCs) are missing two protective proteins, CD55 and CD59

Without their protection, RBCs are attacked and destroyed by a part of your immune system called the **complement system**.

This destruction of RBCs is called hemolysis.

There are three different pathways in the complement system: the lectin, classical, and alternative pathways. The **alternative pathway** plays an important role for hemolysis in PNH.

THE COMPLEMENT SYSTEM



C, complement; MAC, membrane attack complex.

EVH and IVH are two types of hemolysis in PNH.



The upper part of the complement system, specifically the alternative pathway, includes proteins such as Factor B, Factor D, and C3.

Interactions between these proteins drive **EVH**: hemolysis inside the liver and spleen.

EVH, extravascular hemolysis.

The upper part of the complement system leads into the lower part, which drives IVH.



The lower part of the complement system includes many proteins, one being C5, which is important in driving **IVH**: hemolysis inside the blood vessels.

IVH, intravascular hemolysis.

BOTH EVH AND IVH CONTRIBUTE TO PNH SYMPTOMS, INCLUDING ANEMIA AND FATIGUE

