

Hughes syndrome

Hughes syndrome, or antiphospholipid antibody syndrome (APS), is an autoimmune condition that causes thickening of the circulating blood. The immune system produces abnormal blood proteins called antiphospholipid antibodies, which cause blood platelets to clump together.

Hughes syndrome is sometimes called 'sticky blood syndrome' because people with this condition are more likely to form clots in blood vessels (thromboses). People with certain autoimmune diseases such as systemic lupus erythematosus (SLE) are at increased risk of having Hughes syndrome.

Without medical treatment, blood clots may lodge in veins or arteries and cause dangerous complications. Any organ or limb can be affected, depending on the site of the blood clot.

Hughes syndrome is one of the more common autoimmune conditions, yet it is not as well known as some other diseases of the immune system. It is a lifelong condition and the causes are unclear.

There is no cure, but medical treatment can ease symptoms and reduce the risk of complications.

Antiphospholipid antibodies explained

The antiphospholipid antibody is an abnormal type of blood protein that is usually found only in people who have certain diseases such as Hughes syndrome or systemic lupus erythematosus. While most people with Hughes syndrome don't have lupus, about one in every two people with lupus has Hughes syndrome. The reason for this overlap is unclear.

Antiphospholipid antibodies bind to molecules in the blood called phospholipids (a phospholipid is a fat or 'lipid' bound with phosphorus). This autoimmune reaction results in platelets sticking together. Clumping of platelets causes the build-up of clots. It is not clear what triggers the production of antiphospholipid antibodies.

It is thought that about two in every 100 healthy people have mild but detectable levels of antiphospholipid antibodies following illnesses such as hepatitis or malaria. Some drugs such as cocaine, quinine and antibiotics can also cause the blood to make antiphospholipid antibodies. In most of these cases, the temporary production of this abnormal blood protein doesn't cause any harm.

Symptoms of Hughes syndrome

Without treatment, Hughes syndrome can cause many symptoms and complications, including:

- Migraine headaches
- Mottled skin tone (livedo reticularis)
- Low blood platelet count (thrombocytopenia)
- Vein thrombosis
- Deep vein thrombosis (DVT)
- Arterial thrombosis
- Heart attack
- Stroke
- Catastrophic antiphospholipid syndrome.

Complications of pregnancy

Hughes syndrome has been identified as a major cause of recurrent miscarriage. During pregnancy, the placenta provides the growing baby with oxygen and nutrients from the mother's bloodstream. The blood of a woman with Hughes syndrome may be too sticky to pass through the tiny blood vessels of the placenta.

Complications can include:

- Miscarriage
- Fetal death
- Premature labour.

Catastrophic antiphospholipid syndrome

This is a rare but extremely life-threatening complication of Hughes syndrome. Typically, a healthy patient with Hughes syndrome suddenly develops multiple clots in various vital organs, including the brain, heart, lungs or liver. The reason for this is unclear. However, studies show that a percentage of patients have had a recent viral or bacterial infection, which leads some medical researchers to believe that infection may be a trigger.

Primary and secondary Hughes syndrome

The two types of Hughes syndrome include:

- **Primary** – the disease occurs on its own, not in association with an autoimmune condition such as systemic lupus erythematosus. Most people with Hughes syndrome have the primary type.
- **Secondary** – the disease occurs in association with a health problem somewhere else in the body, such as systemic lupus erythematosus.

Diagnosis of Hughes syndrome

Tests used in the diagnosis of Hughes syndrome include:

- Medical history
- Physical examination
- Blood tests.

Treatment for Hughes syndrome

There is no cure for Hughes syndrome. Treatment aims to ease symptoms and reduce the risk of complications. Options include:

- Medications to stop platelets from clumping together, such as low-dose aspirin
- Medications to thin the blood, such as heparin
- Cortisone drugs to control the inflammation associated with autoimmune diseases such as lupus
- Drug therapy to control other health problems, such as hypertension (high blood pressure) or diabetes, that may increase the risk of complications including stroke or heart attack
- Regular medical check-ups to monitor the medications and reduce the risk of side effects: for example, blood thinners may cause uncontrolled bleeding in some cases
- Lifestyle changes such as quitting cigarettes, eating a healthy diet and exercising regularly.

Ongoing medical treatment allows most people with Hughes syndrome to live long and healthy lives. However, you may need to take medications for long periods of time, perhaps for the rest of your life. Some people are troubled by complications, despite the best of care.

Where to get help

- Your doctor, who can refer you to a specialist such as a haematologist, obstetrician or rheumatologist
- In an emergency, always call triple zero (000)

- Arthritis Victoria Tel. (03) 8531 8046 or 1800 011 041
- NURSE-ON-CALL Tel. 1300 60 60 24 – for expert health information and advice (24 hours, 7 days)

Things to remember

- Hughes syndrome is thickening of the circulating blood caused by an abnormal immune system.
- Complications include heart attack, stroke and recurrent miscarriage.
- Treatment includes medications to thin the blood and prevent platelets from clumping together.

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