CONDITION

Phlegmasia cerulea dolens

Phlegmasia cerulea dolens is a severe deep-vein blood clot that causes sudden painful swelling and blue-purple skin in a limb.

WHAT IT IS
Limb-threatening blockage of deep veins by extensive blood clots

BODY SYSTEM
Heart and circulation

MAIN SYMPTOMS
Sudden swelling, severe pain, blue-purple skin, numbness

WHAT CAUSES IT
Massive deep-vein blood clot blocking limb drainage

HOW IT IS DIAGNOSED
Duplex ultrasound, with CT or catheter venography when needed

MAIN TREATMENT
Emergency anticoagulation and often clot-removal procedures

CAN IT BE CURED?
Yes - prompt treatment can restore venous drainage

IS IT AN EMERGENCY?
Yes - blue, rapidly swollen painful limb

LONG-TERM OUTLOOK
Often good with prompt care; serious risks remain

Overview

Phlegmasia cerulea dolens is a rare, severe form of deep vein thrombosis, or DVT. A DVT is a blood clot in a deep vein. In this condition, extensive clotting blocks most or all of the veins carrying blood out of a limb, usually a leg.

Blood enters the limb through arteries but cannot drain normally through the blocked veins. Pressure builds quickly, causing major swelling, severe pain and blue or purple skin. "Cerulea" refers to this blue colour.

The swelling can eventually reduce the flow of oxygen-rich arterial blood into the limb. Without urgent treatment, this can damage nerves, muscles and skin. It may lead to tissue death, called venous gangrene. The clot can also break loose and travel to the lungs, causing a pulmonary embolism.

Phlegmasia cerulea dolens is therefore a medical emergency. It is more serious than an uncomplicated DVT and usually requires treatment in hospital.

Symptoms

Symptoms commonly begin in one leg and may worsen over hours. The whole leg can become involved, from the foot to the groin.

Typical symptoms include:

The swelling may be firm and tense. Fluid-filled blisters can appear on the skin in advanced cases.

Numbness, tingling, weakness or increasing difficulty moving the foot can mean that pressure and poor blood flow are affecting nerves and muscles. The foot may become cool, and pulses may be difficult to feel. These are particularly serious signs.

An earlier stage, called phlegmasia alba dolens, causes major swelling and pale skin without the same blue discolouration. It can progress to phlegmasia cerulea dolens.

Symptoms of a pulmonary embolism may occur at the same time. These include sudden shortness of breath, chest pain that may worsen with breathing, coughing up blood, fainting or a very fast heartbeat.

- Sudden, extensive swelling
- Severe pain, tenderness or a feeling of tightness
- Blue, purple or dusky skin
- A leg that feels heavy and difficult to move

Causes and risk factors

The direct cause is an extensive DVT that blocks the limb's major veins and smaller connecting veins. It often involves the iliac veins in the pelvis and the femoral veins in the thigh. These are sometimes described together as an iliofemoral DVT.

Blood clots are more likely when blood flow is slow, blood vessels are injured or the blood has an increased tendency to clot. Factors that can contribute include:

Some people have compression of a pelvic vein, such as May-Thurner syndrome. This can slow blood flow and make a large left-leg DVT more likely.

Not everyone has an obvious trigger. Having one or more risk factors does not mean that you

will develop this condition.

- Cancer
- Recent major surgery or serious injury
- A long hospital stay or prolonged lack of movement
- A previous DVT or pulmonary embolism
- Pregnancy and the weeks after giving birth
- Oestrogen-containing medicines
- Inherited or acquired blood-clotting conditions
- Severe infection or critical illness
- A catheter placed in a major vein

Diagnosis

Diagnosis begins with the symptoms and an urgent examination of the affected limb. The healthcare team will assess its colour, temperature, swelling, sensation, movement and arterial pulses. They will also check for signs of a pulmonary embolism or circulation problems affecting the whole body.

Duplex ultrasound is usually the first imaging test. It combines standard ultrasound with a measurement of blood flow and can confirm a DVT in many of the deep veins.

Ultrasound may not show pelvic veins clearly. CT venography or MR venography can help identify how far the clot extends and whether a vein is being compressed. Catheter venography, in which contrast dye is injected directly into a vein, may be performed as part of a clot-removal procedure.

Blood tests cannot confirm phlegmasia cerulea dolens by themselves. Tests may be used to check blood counts, kidney function, blood clotting and readiness for treatment. A D-dimer blood test is not a substitute for urgent imaging when symptoms strongly suggest a large, limb-threatening clot.

Imaging should not unnecessarily delay emergency treatment when the clinical signs are clear.

Treatment and management

Treatment aims to stop the clot from growing, restore venous drainage, protect the limb and

prevent a pulmonary embolism. Care usually involves specialists in vascular surgery, interventional radiology, haematology or critical care.

Anticoagulant medicine is started promptly unless there is a reason it cannot be used. Anticoagulants like warfarin or apixaban are often called blood thinners, although they do not literally thin the blood. They reduce further clotting while the body begins to break down the existing clot.

The affected limb may be raised and closely monitored. Fluids, oxygen, pain relief and treatment for low blood pressure may be needed. These measures support circulation but do not replace treatment of the clot.

Because this condition threatens the limb, anticoagulation alone may not be enough. Procedures that may be considered include:

Clot-dissolving treatment can cause serious bleeding, so the team weighs this risk against the immediate threat to the limb. The choice of procedure also depends on how long symptoms have been present, the location of the clot and the person's overall health.

If an underlying pelvic-vein narrowing contributed to the blockage, a stent may sometimes be placed to hold the vein open after the clot has been treated.

A fasciotomy may be needed if severe swelling raises pressure within the muscle compartments of the leg. This operation releases the pressure. If tissue has already died and cannot recover, amputation may occasionally be necessary to prevent further harm.

Anticoagulation generally continues after the emergency treatment. How long it is needed depends on factors such as what caused the clot, whether the trigger remains and the risk of bleeding or another clot.

- Catheter-directed thrombolysis: A thin tube delivers clot-dissolving medicine into the clot
- Mechanical thrombectomy: A catheter-based device removes or breaks up the clot
- Surgical thrombectomy: An operation removes the clot when other methods are unsuitable or unavailable

Complications

The main immediate concern is permanent damage to the affected limb. Severe venous pressure can reduce arterial circulation, depriving tissues of oxygen. This can cause skin breakdown, muscle and nerve injury, compartment syndrome or venous gangrene.

A pulmonary embolism occurs when part of the clot travels to the lungs. A large embolism can place major strain on the heart and circulation.

Extensive swelling can also shift fluid out of the bloodstream. In severe cases, this contributes to low blood pressure, shock and kidney injury. Restoring blood flow to badly affected tissue can sometimes trigger further swelling or chemical changes that require close monitoring.

Longer-term complications include another DVT and post-thrombotic syndrome. Post-thrombotic syndrome results from lasting damage or blockage in the veins. It can cause ongoing swelling, aching, heaviness, skin discolouration and, in severe cases, slow-healing leg ulcers.

Prevention

Phlegmasia cerulea dolens cannot always be prevented. It may develop despite appropriate care, and some contributing factors, such as inherited clotting tendencies or the need for cancer treatment, cannot be removed.

Reducing the risk of DVT also reduces the risk of this severe complication. During hospital stays or after surgery, preventive measures may include anticoagulant medicine, leg-compression devices and getting moving as soon as it is medically safe.

If you have been prescribed an anticoagulant, taking it as directed is important. Do not stop it because symptoms improve without discussing this with the prescribing team.

During long periods of sitting, such as travel, regular leg movement and walking when possible can reduce prolonged immobility. People with previous blood clots, active cancer or other major risk factors may need advice tailored to their situation.

Prompt assessment of new one-sided leg swelling or pain may allow a DVT to be treated before it becomes more extensive.

Living with it and what to expect

The first days focus on preserving the limb and preventing a pulmonary embolism. Swelling and skin colour may begin to improve once venous drainage is restored, but recovery is not always immediate. Pain, weakness and reduced mobility can continue after the hospital stay.

Over the following weeks and months, anticoagulation lowers the chance that the clot will grow or recur. Follow-up may include blood tests, review of bleeding risk and imaging when there is concern about continuing blockage or a treated vein.

Some people recover with little lasting disability. Others have persistent swelling, aching or skin changes because the veins and their one-way valves were damaged. Rehabilitation,

gradual return to activity and treatment of ongoing swelling may help. Compression garments are sometimes used after circulation is stable, but they should be fitted and recommended by the treating team rather than started during an acutely threatened limb.

The risk of another clot depends on why the first one occurred. A temporary trigger, such as surgery, is different from an ongoing risk such as active cancer or a persistent vein abnormality. Follow-up is used to decide how long anticoagulation should continue and whether further treatment is needed.

When to get medical help

Call emergency services now for a leg or arm that becomes rapidly swollen and severely painful, especially when the skin is blue, purple, dusky, blistered, numb or cold. New weakness or difficulty moving the limb also requires emergency care.

Call emergency services for possible pulmonary embolism symptoms, including sudden shortness of breath, chest pain, coughing up blood, fainting or severe light-headedness.

If you are taking an anticoagulant, seek urgent advice for heavy or uncontrolled bleeding, vomiting blood, black stools, a major head injury or a sudden severe headache.

Routine follow-up is appropriate for stable, gradually improving swelling or discomfort after treatment. Contact your regular healthcare team sooner if swelling begins increasing again, a new sore develops, medicine is difficult to take, or you notice unusual bruising or minor bleeding that continues.

Common questions

Is phlegmasia cerulea dolens the same as a DVT?

It is an unusually extensive and severe form of DVT. An ordinary DVT may block one or more deep veins. Phlegmasia cerulea dolens blocks so much venous drainage that the limb becomes massively swollen, painful and blue, with a risk of tissue damage.

Why does the leg turn blue or purple?

Blood enters through the arteries but cannot leave normally through the blocked veins. Oxygen-poor blood collects in the limb, and pressure rises in small blood vessels. This produces the blue or purple colour.

Can the leg be saved?

Often it can, particularly when treatment starts before permanent tissue damage occurs. The

outcome is influenced by how much of the venous system is blocked, how long circulation has been impaired and whether complications such as gangrene or shock have developed.

Will I need surgery?

Not everyone needs open surgery. Many people receive anticoagulation and a catheter-based procedure to dissolve or remove the clot. Open thrombectomy, fasciotomy or amputation is reserved for particular circumstances.

Can phlegmasia cerulea dolens come back?

It can recur, especially when an ongoing risk factor remains. Anticoagulation and follow-up care help reduce this risk. New one-sided swelling, pain or colour change should be assessed promptly.

How long does recovery take?

The emergency phase is treated immediately, but swelling and mobility may take weeks or months to improve. Some people develop lasting symptoms from vein damage. Recovery is shaped by the extent of the clot, the treatment required and the health of the limb after circulation is restored.

Sources

- Chinsakchai K, Ten Duis K, Moll FL, de Borst GJ. Trends in management of phlegmasia cerulea dolens . Vascular and Endovascular Surgery . 2011;45(1):5-14.
- Kesieme EB, Kesieme CN, Jebbin NJ, Irekpita E, Dongo A. Deep vein thrombosis: a clinical review . Journal of Blood Medicine . 2011;2:59-69.
- Kakkos SK, Gohel M, Baekgaard N, et al. European Society for Vascular Surgery 2021 clinical practice guidelines on the management of venous thrombosis . European Journal of Vascular and Endovascular Surgery . 2021;61(1):9-82.
- Ortel TL, Neumann I, Ageno W, et al. American Society of Hematology 2020 guidelines for management of venous thromboembolism: treatment of deep vein thrombosis and pulmonary embolism . Blood Advances . 2020;4(19):4693-4738.
- Vedantham S, Goldhaber SZ, Julian JA, et al. Pharmacomechanical catheter-directed thrombolysis for deep-vein thrombosis . New England Journal of Medicine . 2017;377:2240-2252.