

Vasculitis

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THE STORY IN ONE SENTENCE

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“The body's defense army turns on its own blood vessel walls. Sort by vessel size (big, medium, small). Check the ANCA blood test to narrow the type. Calm the immune attack with steroids and other immune-quieting drugs before organs lose their blood supply.”

Normal anatomy — the body's job

Blood vessels are living tubes with three-layer walls. The inner layer (endothelium) is slick so blood flows smoothly. The middle layer has muscle that can tighten or relax to control blood pressure. The outer layer is tough connective tissue that holds the vessel in place. Together, these layers keep blood highways open, strong, and flexible.

BODY'S JOB

Vessels deliver oxygen-rich blood from the heart to every organ and then carry waste-filled blood back. Big arteries (like the aorta) are the main highways. Medium arteries branch out to individual organs (kidneys, gut, limbs). Small arterioles and capillaries are the tiny side streets that reach each tissue cell. Healthy vessel walls let blood flow freely, adjust pressure as needed, and stay sealed so nothing leaks out.

1. Heart pumps oxygen-rich blood into the aorta (the body's biggest artery).
2. Big arteries branch into medium arteries that serve each organ (renal artery to kidney, mesenteric artery to gut, carotid artery to brain).
3. Medium arteries split into small arterioles, then into capillaries (the tiniest vessels where oxygen and nutrients pass into tissue).
4. Capillaries join into venules, then veins, which carry waste-filled blood back to the heart and lungs to pick up fresh oxygen.
5. The three-layer vessel wall stays smooth, strong, and sealed at every step.

What breaks

The immune system attacks the walls of blood vessels as if they were germs. White blood cells swarm the vessel wall, release chemicals that cause swelling (inflammation), and sometimes form clumps called granulomas (balls of angry immune cells). This attack thickens and narrows the vessel, slowing or blocking blood flow. It also weakens the wall so it can leak blood or burst. Which vessels get hit—big, medium, or small—determines which organs suffer.

CLINICAL TERM

Immune-mediated vasculitis. Autoantibodies (such as ANCA in small-vessel disease) or activated T-cells and macrophages invade the vessel wall, triggering an inflammatory cascade. The endothelium is damaged, the media thickens with immune infiltrate and sometimes granulomas, and the vessel lumen narrows (stenosis) or the wall weakens (aneurysm, rupture). Downstream starved blood flow (ischemia) (tissue starvation) and bleeding (hemorrhage) (bleeding) follow.

How the body compensates

The body tries to keep organs alive by raising heart rate and blood pressure to push blood past narrowed vessels, rerouting flow through backup arteries (collaterals), and triggering inflammation signals (fever, weight loss) to call for immune help. These moves can hide how sick the patient really is—vital signs may look okay even as organs are quietly starving. The danger is missing the point when compensation tips into organ failure.

1. {'label': 'A fast heart rate (Tachycardia) and high blood pressure (hypertension)', 'translation': 'The heart beats faster and squeezes harder to push blood through narrowed, inflamed vessels and keep oxygen delivery up.'}
2. {'label': 'Collateral circulation', 'translation': 'Backup side-branch arteries open up to detour blood around a blocked main vessel; this buys time but cannot carry full flow forever.'}
3. {'label': 'Systemic inflammation markers', 'translation': '"The liver pumps out acute-phase proteins (ESR and CRP rise) and the hypothalamus (the brain's thermostat) raises temperature (fever) to fight what it thinks is an invader; weight loss happens because the immune war burns extra calories."}
4. {'label': 'Silent starved blood flow (ischemia)', 'translation': 'Organs may lose half their blood supply before pain or dysfunction appears; by the time symptoms show, damage is often already done.'}

5. {'label': 'The crash point: irreversible organ loss', 'translation': 'Once tissue dies (kidney scarring, nerve death, bowel tissue death from blocked blood flow (infarction), blindness from optic-nerve starved blood flow (ischemia) in giant cell arteritis), it cannot grow back. The window to stop damage with immune-quieting treatment closes fast.'}
6. {'label': 'Trend over snapshot', 'translation': 'A stable blood pressure today means little if creatinine is climbing, rash is spreading, or vision is dimming—the slope (getting worse or better) matters more than one moment in time.'}

Domino effect

1. Immune cells attack vessel walls everywhere. — In vasculitis, the immune system makes antibodies (like ANCA—antineutrophil cytoplasmic antibodies that tag white blood cells to attack vessels) or activates T-cells (the immune system's attack squad) that mistake vessel-wall proteins for foreign invaders. White blood cells swarm into the wall, release inflammatory chemicals (cytokines like TNF-alpha and interleukin-1, plus reactive oxygen species that burn tissue), and damage the endothelium (slick inner lining) and muscle layers.
2. Fever and weight loss appear first. — The widespread immune battle raises the body's thermostat (the hypothalamus in the brain) to create fever, which is meant to slow germs but here just signals inflammation. The fight also burns extra calories and suppresses appetite (cytokines like IL-1 and TNF-alpha turn down hunger signals), so weight drops.
3. Red or purple spots (purpura) appear on skin. — Small skin vessels leak red blood cells when their walls break from immune attack; the leaked blood shows as tiny red or purple dots (petechiae) or larger bruise-like patches (purpura) that do not blanch (turn white) when you press them.
4. Blood appears in urine (hematuria). — In the kidney, inflammation punches holes in the glomerulus (the tiny filter ball made of capillaries), so red blood cells spill through into the urine. Protein leaks too (proteinuria), making urine foamy.
5. Coughing up blood (hemoptysis) signals lung involvement. — Small vessels in the lung's air sacs (alveoli—the tiny balloon-like pockets where oxygen enters the blood) rupture when vasculitis hits them; blood leaks into the airway and is coughed out. This is common in granulomatosis with polyangiitis (GPA) and microscopic polyangiitis (MPA).
6. Numbness, tingling, or weakness in hands and feet (mononeuritis multiplex). — Vasculitis blocks the tiny arteries (vasa nervorum—the little supply vessels that feed peripheral nerves). Starved of oxygen, the nerve fibers malfunction—first tingling, then numbness, then weakness. Multiple separate nerves fail in a patchy pattern (not symmetric like diabetic neuropathy).

7. Severe belly pain and bloody stools. — When vasculitis clogs the arteries feeding the intestines (mesenteric vessels), sections of bowel lose their blood supply. The gut lining dies (ischemia, then tissue death from blocked blood flow (infarction)—tissue death), causing intense pain, bleeding into the stool, and risk of perforation (a hole that spills gut contents into the belly, leading to life-threatening infection).
8. Sudden vision loss or severe headache in giant cell arteritis. — Giant cell arteritis inflames the temporal artery (the vessel you can feel pulsing at your temple) and branches that feed the optic nerve (the cable carrying signals from the eye to the brain). If the optic artery clogs, the nerve dies within hours and blindness is permanent. Jaw pain when chewing (jaw claudication—the jaw muscles cramping because their blood supply is choked) is another clue that the chewing-muscle arteries are starved.

Symptoms · what the patient feels, and why

- Fever that won't go away: The inflamed blood vessel walls release chemicals called cytokines (alarm signals) that travel to the hypothalamus (the brain's thermostat) and reset your body temperature higher, like turning up a heater to fight what looks like an infection.
- Weight loss you didn't try for: Ongoing inflammation burns extra energy — like leaving a car engine running all the time — and the cytokines also kill your appetite, so you eat less and your body starts using its own stores.
- Purple or red spots on the skin (purpura): Tiny blood vessels in the skin get inflamed and leak red blood cells into the surrounding tissue, like a garden hose with holes spraying water into the dirt, leaving behind purple or red marks.
- Numbness, tingling, or weakness in hands or feet: The small arteries feeding the nerves in your arms and legs get inflamed and narrow, starving the nerves of oxygen and nutrients — like a pinched phone charger that stops electricity flowing — so the nerves can't send signals properly.
- Blood in the urine (hematuria): Inflammation hits the tiny blood vessels in the kidney's filters (glomeruli), making them leaky so red blood cells spill into the urine, turning it pink, red, or cola-colored.
- Jaw pain when chewing (jaw claudication): In giant cell arteritis, the arteries feeding the jaw muscles get inflamed and narrowed, so when you chew and the muscles need more oxygen, they don't get enough — like a cramp from running with a blocked airway — and they ache or tire quickly.
- New, severe headache (often at the temples): The temporal arteries (the blood vessels on the sides of your head) swell and their walls become tender, pressing on nearby nerves and causing a throbbing, aching pain.

- Sudden vision loss or double vision: Inflamed arteries near or behind the eye cut off blood flow to the optic nerve (the cable that sends pictures from your eye to your brain), starving it of oxygen — this is an emergency because the nerve can die and leave you blind in that eye.
- Cough, wheezing, or shortness of breath: In some vasculitides — especially eosinophilic granulomatosis with polyangiitis (EGPA) — blood vessels in the lungs become inflamed and the airways tighten, like asthma, because extra eosinophils (a type of white blood cell) flood the tissue and trigger allergic-like swelling.
- Joint pain and swelling (arthralgia or arthritis): Inflamed blood vessels in or near the joints leak fluid and immune cells into the joint space, causing swelling, warmth, and pain — the same process that happens in many autoimmune diseases where the body mistakes its own tissue for an invader.

Focused exam

- Fever and weight loss over days to weeks: When the immune system attacks blood vessels all over the body, the constant inflammation burns energy and raises temperature. These general signs tell you something is inflaming many organs at once, not just one spot.
- Palpable purple bleeding blotches (purpura) (raised purple or red spots on the skin, usually on legs): Tiny blood vessels in the skin are so swollen and damaged that blood leaks out into the tissue. These spots do not blanch when you press them because the blood is stuck outside the vessels. This means small-vessel vasculitis is happening right under the skin where you can see it.
- Blood in the urine (Hematuria) (blood in the urine, often visible or microscopic) or new facial or ankle swelling: The kidneys filter blood through millions of tiny vessels. When vasculitis inflames these glomerular capillaries, they leak red blood cells and protein into the urine, and fluid backs up into the body. Kidney vasculitis can destroy filtering units fast, so catching this early protects kidney function.
- Coughing up blood (Hemoptysis) (coughing up blood) or shortness of breath: Inflamed lung capillaries can bleed into the air sacs, making breathing hard and oxygen low. When lung bleeding happens at the same time as kidney damage (called pulmonary-renal syndrome), it points to ANCA-associated vasculitis and needs emergency treatment to stop irreversible organ loss.
- New foot drop, wrist drop, or numbness in a glove-and-stockings pattern (mononeuritis multiplex): Nerves depend on a steady blood supply from tiny feeding vessels called vasa nervorum. When vasculitis blocks or inflames these vessels, the nerve starves and stops conducting signals. Sudden weakness in one hand or foot that does not match a stroke pattern is a red flag for nerve-vessel damage.

- Severe new headache, jaw pain with chewing, or sudden vision loss in one eye (especially in someone over 50): Giant cell arteritis inflames the medium and large arteries in the head, especially the temporal artery and the vessels feeding the optic nerve. Jaw claudication (pain when chewing because the jaw muscles are not getting enough blood) and vision loss mean the vessels are so swollen they are blocking blood flow. Vision loss can become permanent in hours if not treated with high-dose steroids immediately.

Diagnostics · every test answers a question

- Red blood cell (Erythrocyte) sedimentation rate (ESR) and C-reactive protein (CRP) → Is there active widespread inflammation in the body right now?
- ANCA (antineutrophil cytoplasmic antibodies): PR3-ANCA and MPO-ANCA → Are there antibodies attacking the walls of small blood vessels?
- Urinalysis with microscopy → Are the kidneys being damaged by inflamed glomerular capillaries?
- Complete blood count (CBC) with differential → Is there anemia, low platelets, or high eosinophils that point to a specific vasculitis type?
- Serum creatinine and estimated glomerular filtration rate (eGFR) → How much kidney function has been lost, and how fast?
- Tissue biopsy (kidney, skin, lung, sural nerve, or temporal artery depending on the presentation) → Can we see inflamed vessel walls under the microscope and confirm the diagnosis?
- Hepatitis B and C serology, cryoglobulins, complement levels (C3, C4), antinuclear antibodies (ANA) → Is there an underlying infection or autoimmune disease causing secondary vasculitis instead of primary vasculitis?
- Chest X-ray or high-resolution CT chest (if lung symptoms or suspicion of GPA/MPA/EGPA) → Are there nodules, infiltrates, or bleeding in the lungs that confirm lung involvement?

Treatment logic

- High-dose corticosteroids (e.g., prednisone 1 mg/kg/day, usually 60–80 mg daily, or IV methylprednisolone 500–1000 mg daily for severe organ-threatening disease) → Steroids shut down the immune cells that are releasing inflammatory signals and attacking vessel walls. This stops new inflammation quickly (within hours to days) and allows damaged vessels to begin healing. In giant cell arteritis, high-dose steroids prevent permanent vision loss. In ANCA vasculitis, they halt kidney and lung destruction.

- Cyclophosphamide (IV or oral) or rituximab (anti-CD20 monoclonal antibody) for severe ANCA-associated vasculitis (GPA, MPA, EGPA with organ threat) → These drugs knock down or wipe out the B cells and other immune cells making the antibodies (ANCA) that attack vessel walls. Rituximab depletes CD20+ B cells; cyclophosphamide kills rapidly dividing immune cells. Both induce remission (stop active disease) and work deeper and longer than steroids alone, preventing relapse. Current guidelines favor rituximab over cyclophosphamide for induction in most cases because it has similar efficacy with a better long-term safety profile.
- Plasma exchange (plasmapheresis, PLEX) for severe pulmonary bleeding (hemorrhage) or rapidly progressive glomerulonephritis with dialysis-level kidney failure → PLEX physically removes circulating ANCA antibodies and inflammatory proteins from the blood right away, before steroids and immunosuppressants have time to work. This can rescue kidney function and stop lung bleeding in the most severe cases. Some studies show benefit when creatinine is very high (e.g., > 5.7 mg/dL) or the patient needs dialysis at presentation.
- Low-dose aspirin (75–100 mg daily) for giant cell arteritis → Aspirin inhibits platelet clumping, making blood less likely to clot inside inflamed arteries. In giant cell arteritis, inflamed vessel walls can trigger clots that block the ophthalmic artery (causing blindness) or cerebral arteries (causing stroke). Aspirin reduces this blood-starved (ischemic) complication risk.
- Tocilizumab (IL-6 receptor blocker, subcutaneous injection weekly or every other week) for giant cell arteritis → Tocilizumab blocks interleukin-6, a key inflammatory signal that drives giant cell arteritis. Adding it to steroids lets patients taper steroids faster and stay in remission longer with lower cumulative steroid doses, reducing steroid side effects like osteoporosis, diabetes, and weight gain. FDA-approved for giant cell arteritis based on the GiACTA trial.
- Azathioprine, methotrexate, or mycophenolate mofetil for maintenance therapy after remission induction → Once rituximab or cyclophosphamide plus steroids have stopped active vasculitis, a gentler immunosuppressant is used long-term (often 18–24 months or longer) to keep the disease quiet and prevent relapse. These drugs have fewer severe side effects than cyclophosphamide and allow further steroid tapering.
- Prevention (Prophylaxis) against *Pneumocystis jirovecii* pneumonia (PCP) with trimethoprim-sulfamethoxazole during intense immunosuppression → High-dose steroids plus cyclophosphamide or rituximab severely weaken the immune system, creating high risk for opportunistic infections. Preventive (Prophylactic) antibiotics prevent PCP, a life-threatening pneumonia in immunosuppressed patients. Guidelines recommend PCP prevention (prophylaxis) whenever prednisone ≥ 20 mg/day is combined with another immunosuppressant.

Medications

Prednisone (corticosteroid) · Glucocorticoid (steroid)

Prednisone quiets down the immune system's attack on blood vessel walls. It works like a dimmer switch: it turns down the production of inflammatory chemicals (cytokines like TNF-alpha and IL-1) that white blood cells release when they're fired up. By cooling the inflammation, it stops the swelling and damage inside the arteries and veins so blood can flow normally again.

1. Prednisone enters immune cells and binds to a receptor inside the cell.
2. This receptor travels into the cell's control center (nucleus) and turns off the genes that make inflammatory chemicals like TNF-alpha, IL-1, and IL-6.
3. Fewer inflammatory chemicals means fewer white blood cells swarming the vessel wall.
4. The swelling in the vessel wall goes down, the tissue heals, and blood flow improves.
5. Symptoms like fever, pain, and organ damage start to reverse within days to weeks.

SAFETY WATCH

High blood sugar (can trigger or worsen diabetes) · Weight gain and increased appetite · Bone thinning (osteoporosis)—patients on long courses need calcium, vitamin D, and sometimes a bisphosphonate · Higher risk of infections (bacterial, fungal, viral) · Mood swings, trouble sleeping, or anxiety · High blood pressure · Stomach irritation or ulcers—take with food · Cataracts and glaucoma with prolonged use · Adrenal gland suppression—NEVER stop suddenly; taper slowly under medical guidance

Cyclophosphamide (Cytoxan) · Alkylating agent (immunosuppressant chemotherapy)

Cyclophosphamide shuts down the immune cells (B cells and T cells) that are attacking the blood vessels. It works by damaging the DNA inside rapidly dividing immune cells so they can't multiply. Because the immune army can't keep growing, the attack on the vessel walls stops and inflammation fades.

1. Cyclophosphamide is converted in the liver into an active form (phosphoramidate mustard).
2. The active drug enters immune cells and attaches to their DNA strands.
3. The DNA breaks, so the cell can't copy itself or make the proteins it needs to survive.
4. B cells (which make antibodies, including the harmful ANCA antibodies in some vasculitis) and T cells stop multiplying.
5. Over weeks, the immune attack weakens and vessel inflammation decreases.

SAFETY WATCH

Low white blood cell count (increases infection risk)—regular blood tests required · Bleeding in the bladder (hemorrhagic cystitis)—drink lots of water and may need a protective drug called mesna · Nausea and vomiting · Hair thinning or loss · Infertility (can be permanent)—discuss egg or sperm banking before starting · Increased risk of certain cancers (bladder cancer, blood cancers) with long-term use · Liver enzyme changes—monitor labs · Suppressed bone marrow—watch blood counts closely

Rituximab (Rituxan) · Monoclonal antibody (B-cell-depleting agent)

Rituximab targets and removes B cells, the immune cells that make harmful antibodies (like ANCA) that tag blood vessels for attack. It works like a smart eraser: it sticks to a marker (CD20) on the surface of B cells and flags them for destruction. Once the B cells are gone, the production of damaging antibodies stops and inflammation in the vessels quiets down.

1. Rituximab is given by IV infusion; it travels through the bloodstream.
2. It binds to CD20, a protein found only on B cells.
3. Once attached, rituximab marks the B cell for destruction by the body's cleanup crew (natural killer cells and complement proteins).
4. B cells die off within hours to days; levels stay low for months.
5. Without B cells making ANCA antibodies, the immune attack on vessel walls fades and inflammation decreases.

SAFETY WATCH

Infusion reactions (fever, chills, rash, low blood pressure) during or right after the IV—premedicate with acetaminophen and antihistamine · Increased infection risk, especially viral and fungal infections · Low antibody levels over time—may need immunoglobulin (IVIG) replacement if severe · Rare risk of progressive multifocal leukoencephalopathy (PML), a serious brain infection—watch for new confusion, weakness, or vision changes · Hepatitis B reactivation—screen before starting · Low white blood cell or platelet counts · Heart rhythm problems during infusion in patients with heart disease

Methotrexate · Antimetabolite (immunosuppressant)

Methotrexate slows down immune cells by blocking an enzyme (dihydrofolate reductase) they need to build DNA. Think of it like cutting off the supply of bricks to a construction crew—without enough building blocks, immune cells (especially T cells) can't multiply as fast. This

calms the immune attack on blood vessel walls and keeps inflammation low after the initial flare is controlled.

1. Methotrexate enters immune cells and blocks dihydrofolate reductase, the enzyme that recycles folate (a B vitamin needed to make DNA bases).
2. Without enough folate, cells can't build new DNA strands.
3. Immune cells stop dividing as quickly, so the attack on vessel walls weakens.
4. Inflammation decreases and the risk of relapse drops.
5. The drug also raises levels of adenosine, a natural anti-inflammatory chemical, which further quiets the immune system.

SAFETY WATCH

Mouth sores (stomatitis)—take folic acid daily to reduce this · Nausea and upset stomach—often better if taken with food or at bedtime · Low blood counts (white cells, red cells, platelets)—requires regular blood tests · Liver damage—check liver enzymes every 8 to 12 weeks · Lung inflammation (rare but serious)—report new cough or shortness of breath immediately · Birth defects—absolutely avoid pregnancy; use reliable contraception · Avoid alcohol (increases liver risk) · Increased sensitivity to sunlight—use sunscreen

Azathioprine (Imuran) · Purine analog (immunosuppressant)

Azathioprine tricks immune cells by mimicking a building block (purine) they need to copy their DNA. When immune cells (especially T and B cells) try to use azathioprine as a brick, they can't finish building new DNA, so they stop multiplying. This keeps the immune attack on blood vessels under control and helps prevent relapses after the initial treatment.

1. Azathioprine is broken down in the body into 6-mercaptopurine, the active form.
2. This active drug enters immune cells and gets mistaken for a normal DNA building block (a purine).
3. When the cell tries to copy its DNA, the fake block jams the machinery.
4. T cells and B cells can't divide normally, so fewer immune cells are made.
5. The immune attack on vessel walls weakens, inflammation stays low, and relapse risk drops.

SAFETY WATCH

Low white blood cell count (leukopenia)—increases infection risk; requires regular blood tests · Low platelet count—watch for easy bruising or bleeding · Liver enzyme elevations—monitor liver tests every 8 to 12 weeks · Nausea and vomiting—take with food · Increased risk of certain cancers (skin cancer, lymphoma) with long-term use—use sunscreen and get regular skin checks · Pancreatitis (rare but serious)—stop immediately if severe abdominal pain occurs · Genetic test (TPMT enzyme level) recommended before starting—some people break down the drug too slowly and need a lower dose

Tocilizumab (Actemra) · Monoclonal antibody (IL-6 receptor blocker)

Tocilizumab blocks a specific inflammatory signal called interleukin-6 (IL-6), which is one of the main alarm bells that keep the immune attack going in giant cell arteritis. It works by attaching to the IL-6 receptor on immune cells, so IL-6 can't deliver its 'keep fighting' message. Without that signal, the inflammation in large arteries (especially the temporal artery and aorta) cools down and symptoms improve.

1. Tocilizumab is given by IV infusion or under-the-skin injection; it travels through the bloodstream.
2. It binds to IL-6 receptors on the surface of immune cells and vessel-lining cells.
3. IL-6 (the inflammatory signal) can no longer lock onto its receptor and send the 'attack' message.
4. Immune cells produce fewer inflammatory chemicals, and the swelling in artery walls decreases.
5. Patients can often taper their steroid dose faster and lower, reducing steroid side effects.

SAFETY WATCH

Increased infection risk (bacterial, viral, fungal)—screen for tuberculosis before starting · Elevated liver enzymes—monitor liver tests regularly · Low white blood cell or platelet counts—check blood counts before each dose · High cholesterol—may need a statin · Gastrointestinal perforation (very rare but serious)—report severe new belly pain immediately · Infusion reactions (rash, itching, dizziness) if given IV · Can hide fever—report any new symptoms even without a fever

Aspirin (low-dose, 81 mg daily) · Antiplatelet agent

Low-dose aspirin keeps platelets (the sticky blood cells that form clots) from clumping together too easily. It works by blocking an enzyme (cyclooxygenase-1, or COX-1) inside platelets that makes thromboxane A2, a chemical that tells platelets to stick and form a plug. By preventing

clots, aspirin lowers the risk of stroke and heart attack in patients with inflamed arteries (especially in giant cell arteritis).

1. Aspirin enters platelets and permanently binds to the COX-1 enzyme.
2. COX-1 can no longer make thromboxane A₂, the signal that tells platelets to stick together.
3. Platelets become less sticky and less likely to form dangerous clots.
4. Blood flow through inflamed arteries (like the temporal artery in giant cell arteritis) stays smoother.
5. The risk of stroke (from a clot in a brain artery) or heart attack drops.

SAFETY WATCH

Stomach irritation, heartburn, or ulcers—take with food or a proton pump inhibitor if needed · Increased bleeding risk (nosebleeds, bruising, blood in stool or urine)—report any unusual bleeding · Do NOT take if you have active stomach ulcer or recent bleeding · Allergic reactions (rare)—rash, wheezing, swelling · Avoid in children with viral infections (risk of Reye syndrome, a rare brain-liver problem)

Plasma Exchange (PLEX, plasmapheresis) · Procedure (not a drug—removes harmful antibodies from blood)

Plasma exchange is like doing a partial oil change on the blood. A machine filters the patient's blood, removing the liquid part (plasma) that contains harmful antibodies (like ANCA) and inflammatory proteins. The blood cells are mixed with fresh donor plasma or a replacement fluid and returned to the body. This quickly lowers the level of antibodies attacking the kidneys and lungs in severe ANCA-associated vasculitis.

1. Blood is drawn from a vein and passed through a machine (plasmapheresis device).
2. The machine spins or filters the blood to separate plasma (the liquid) from blood cells.
3. The plasma (which contains ANCA antibodies and inflammatory chemicals) is discarded.
4. The blood cells are mixed with replacement fluid (donor plasma or albumin solution) and returned to the patient.
5. Over several sessions (usually 5 to 7 exchanges in 10 to 14 days), antibody levels drop and the immune attack on kidneys and lungs lessens.

SAFETY WATCH

Low blood pressure during the procedure—fluids and slower flow rates help · Tingling or muscle cramps from low calcium (citrate in replacement fluid binds calcium)—may need calcium supplements · Infection risk (requires a large IV line or catheter) · Bleeding risk (procedure uses blood thinners to prevent clotting in the machine) · Allergic reactions to donor plasma (rare) · Fatigue after sessions—schedule rest time

Red flags

- Sudden vision loss or new severe headache in someone over 50 (possible giant cell arteritis — needs urgent steroids to save sight)
- Blood in the urine plus swelling or high blood pressure (kidney damage can spiral quickly)
- Coughing up blood or fast-worsening shortness of breath (lungs may be bleeding or severely inflamed)
- Numbness, weakness, or stroke-like symptoms (brain blood vessels may be involved)

Nursing priorities

1. Check vision in each eye separately every 4 hours and report any new blurry vision, blind spots, or vision loss to the provider immediately — Giant cell arteritis can cause anterior blood-starved (ischemic) optic neuropathy (blood flow loss to the optic nerve), leading to permanent blindness within hours if steroids are delayed
2. Monitor oxygen saturation continuously and listen to lung sounds every 2 hours; report oxygen saturation below 92 percent or new crackles — Pulmonary bleeding (hemorrhage) (lung bleeding) from ANCA-associated vasculitis can progress rapidly to respiratory failure and death; early detection allows time for plasma exchange and mechanical ventilation if needed
3. Measure urine output every hour and check urine color; report output less than 0.5 mL per kilogram per hour or dark urine — Rapidly progressive glomerulonephritis (fast kidney inflammation) in vasculitis can destroy nephrons (kidney filtering units) within days, causing permanent dialysis dependence if immunosuppression is not started quickly
4. Administer high-dose corticosteroids exactly on time (usually prednisone 40 to 60 mg daily or IV methylprednisolone 1000 mg daily for severe cases) and never hold the dose without provider order — Corticosteroids stop the inflammatory cascade (immune attack on blood vessel walls) and prevent organ damage; even one missed dose can allow vasculitis to progress and cause stroke, blindness, or kidney failure

5. Check blood pressure in both arms and both legs at admission and then every 4 hours; report differences greater than 10 mmHg between arms — Unequal blood pressures indicate narrowing (stenosis) (narrowing) or blockage (occlusion) (blockage) of large arteries from Takayasu arteritis or giant cell arteritis, which changes imaging and treatment plans
6. Monitor blood glucose (finger-stick) every 6 hours and give insulin per sliding scale if glucose is above 180 mg/dL — High-dose corticosteroids cause high blood sugar (hyperglycemia) (high blood sugar) by increasing gluconeogenesis (liver sugar production) and insulin resistance; uncontrolled glucose delays wound healing and increases infection risk
7. Assess skin, nose, and extremities for new purple bleeding blotches (purpura) (purple spots), ulcers, or black discolored areas (necrosis); photograph and measure any skin changes — Palpable purple bleeding blotches (purpura) (raised purple spots) indicates leukocytoclastic vasculitis (small-vessel inflammation), while digital starved blood flow (ischemia) (finger or toe blood flow loss) suggests medium-vessel involvement; skin findings help classify vasculitis type and guide biopsy decisions
8. Test hand grip strength and foot dorsiflexion (ability to lift toes up) every shift and ask about new numbness or tingling — Mononeuritis multiplex (nerve damage in multiple separate areas) or peripheral neuropathy from vasculitis affecting vasa nervorum (tiny arteries feeding nerves) can cause permanent disability if immunosuppression is delayed

Sources

- ACR/EULAR 2022 Classification Criteria for Vasculitis
- Chapel Hill Consensus Conference (CHCC) 2012 Nomenclature
- Cochrane systematic reviews on vasculitis treatment
- UpToDate clinical guidelines (Rheumatology)

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