

Pheochromocytoma

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THE STORY IN ONE SENTENCE

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“Pheochromocytoma is a tumor that makes adrenaline-type chemicals. How well you block those chemicals before surgery decides if the patient lives.”

Normal anatomy — the body's job

Chromaffin cells live inside the adrenal medulla (the inner core of each adrenal gland, which sits on top of each kidney) and in a few spots along the spine called paraganglia. Their job is to make and release adrenaline (epinephrine) and noradrenaline (norepinephrine) when the sympathetic nervous system tells them to—like when you are scared, hurt, or need a burst of energy.

BODY'S JOB

When the brain's sympathetic alarm goes off, it sends a nerve signal down to the adrenal medulla. The chromaffin cells respond by dumping adrenaline and noradrenaline into the bloodstream. Those chemicals race to the heart (making it beat faster and harder), the blood vessels (squeezing them to raise blood pressure), the lungs (opening airways), and the liver (releasing sugar for quick fuel). This is the 'fight-or-flight' response. After a few minutes, the surge fades and everything returns to baseline.

1. The brain detects danger or stress.
2. A nerve signal travels down the spinal cord to the adrenal medulla.
3. Chromaffin cells release adrenaline and noradrenaline into the blood.
4. The heart beats faster and harder; blood vessels squeeze; blood pressure rises; airways open; pupils widen; sweat glands turn on.
5. After a few minutes, enzymes (MAO and COMT) break down the catecholamines and the body returns to calm.

What breaks

A tumor grows from chromaffin cells and starts making adrenaline and noradrenaline on its own, ignoring the brain's normal on-off switch. The tumor dumps these chemicals in huge surges (or a constant leak), flooding the body with a fake emergency signal even when the person is resting.

CLINICAL TERM

Pheochromocytoma (tumor in the adrenal medulla) or paraganglioma (tumor in chromaffin tissue outside the adrenal gland, along the sympathetic chain). About 70% are sporadic (random, one-time); about 30% are hereditary, linked to mutations in genes like RET (MEN2A, MEN2B), VHL (von Hippel-Lindau), NF1 (neurofibromatosis type 1), or SDH subunits (familial paraganglioma syndromes). The tumor autonomously secretes catecholamines—mainly norepinephrine, sometimes epinephrine, rarely dopamine—independent of physiologic control.

How the body compensates

The body tries to protect itself from the wild swings in blood pressure and heart rate by downregulating receptors and shifting fluid, but these changes make the patient fragile. Vital signs can look deceptively stable between surges, hiding how close the patient is to a crisis. The most dangerous moment is unblocked surgery: anesthesia or tumor handling can trigger a massive catecholamine surge, causing stroke-level blood pressure or deadly arrhythmias, followed by sudden low blood pressure (hypotension) once the tumor is removed and the catecholamine tap shuts off.

1. {'step': 'Between surges, blood pressure may be normal or only mildly elevated.', 'why': 'Receptor downregulation and lower blood volume keep pressure from staying sky-high all the time, masking the tumor's presence.'}
2. {'step': 'The patient may have orthostatic low blood pressure (hypotension) (pressure drops ≥ 20 mmHg when standing).', 'why': 'Chronic catecholamine exposure shrinks blood volume and exhausts vascular receptors, so the vessels cannot squeeze enough to keep pressure up when gravity pulls blood to the legs.'}
3. {'step': 'Standard blood pressure medicines (ACE inhibitors, ARBs, diuretics, calcium blockers) fail to control pressure.', 'why': 'The tumor keeps pouring out catecholamines that override the drugs' effects. Only alpha-blockers (phenoxybenzamine, doxazosin) can compete with the flood of norepinephrine at the receptor.'}

4. {'step': 'Watch for the crash point: unblocked patient going to surgery.', 'why': 'Anesthesia induction, intubation, or the surgeon touching the tumor can trigger a massive catecholamine dump, spiking blood pressure above 200–300 mmHg and causing stroke, heart attack, or lethal arrhythmias.'}
5. {'step': 'After the tumor is removed, blood pressure can crash (post-resection low blood pressure (hypotension)).', 'why': 'The catecholamine source is suddenly gone, but the blood vessels are still dilated and downregulated, and blood volume is still low. The patient needs IV fluids and sometimes short-term vasopressors to keep pressure stable.'}
6. {'step': 'With high blood pressure (Hypertensive) emergency (pressure >180/120 with organ damage) can happen in untreated patients.', 'why': 'A spontaneous surge or physical stress (trauma, illness, certain drugs) can push pressure high enough to cause stroke, heart failure, aortic dissection, or kidney failure in minutes.'}
7. {'step': 'Look at the trend—how symptoms are changing over days to weeks—not just one moment.', 'why': 'Pheochromocytoma is episodic. A normal blood pressure in the clinic does not rule it out; you need to catch the surges with 24-hour urine or plasma metanephrines (breakdown products of catecholamines).'} }

Domino effect

1. Chromaffin tumor cells dump norepinephrine (and sometimes epinephrine) into the bloodstream in uncontrolled surges. — The tumor has lost normal feedback control, so it releases catecholamines randomly or in response to physical triggers like abdominal pressure, bending, or urination.
2. Norepinephrine binds to alpha-1 receptors on blood vessel walls, squeezing them tight (vasoconstriction). — The squeeze raises resistance in the arteries, shooting blood pressure up—sometimes to 200, 250, even 300 mmHg systolic—in seconds.
3. Epinephrine (if the tumor makes it) binds to beta-1 receptors in the heart, making it beat faster and harder. — The heart pounds (palpitations) and races (tachycardia, often 120–160 bpm), pumping more blood into the already-squeezed vessels, driving pressure even higher.
4. The patient feels sudden severe headache (pounding, throbbing), profuse sweating (soaking clothes), tremor, chest pain or pressure, and intense anxiety or sense of doom. — High pressure stretches pain receptors in blood vessel walls (headache). Catecholamines activate sweat glands all over the body (diaphoresis). Beta stimulation causes muscle tremor. The brain's amygdala senses the body's alarm state and generates fear, even though there is no real danger.
5. Over weeks to months, chronic catecholamine exposure downregulates alpha and beta receptors and shrinks blood volume. — The body tries to protect itself from constant overstimulation by reducing the number and sensitivity of receptors. Blood vessels leak fluid into tissues, lowering total blood volume.

6. The patient develops orthostatic low blood pressure (hypotension) (pressure drops ≥ 20 mmHg when standing, causing dizziness or near-fainting). — When the patient stands, gravity pulls blood to the legs. Normally, vessels squeeze to push it back up. But downregulated receptors and low blood volume mean the vessels cannot squeeze well, so brain blood flow drops and the patient feels faint.
7. Blood pressure becomes resistant to standard medicines; only alpha-blockers (phenoxybenzamine, doxazosin) and sometimes calcium blockers work. — The tumor keeps flooding receptors with catecholamines, overpowering ACE inhibitors, ARBs, and diuretics. Alpha-blockers compete directly at the receptor and win.
8. If the patient goes to surgery without alpha-blockade first, anesthesia or tumor manipulation triggers a catecholamine storm, spiking pressure to stroke or heart-attack levels and causing lethal arrhythmias. — The tumor releases a massive bolus of catecholamines all at once. Without alpha-blockers on board to buffer the surge, blood vessels squeeze violently, and the heart's electrical system can spiral into ventricular a fast heart rate (tachycardia) or fibrillation.
9. Once the tumor is removed, the catecholamine source vanishes and blood pressure can crash (hypotension). — The vessels are still dilated and the receptors still downregulated, but the driving catecholamine surge is gone. The patient needs IV fluids to refill blood volume and sometimes short-term pressors until receptors recover over days.

Symptoms · what the patient feels, and why

- Sudden episodes of pounding headache: The tumor dumps huge bursts of adrenaline and noradrenaline (chemicals that rev up the heart and squeeze blood vessels) into the blood, spiking blood pressure so hard and fast that the vessels in the head throb and ache.
- Heavy sweating that comes in waves: The flood of adrenaline and noradrenaline triggers the sweat glands all over the body as part of the fight-or-flight alarm, even though there is no real danger.
- Fast, pounding heartbeat (palpitations): Adrenaline and noradrenaline make the heart beat harder and faster, like flooring the gas pedal on a car, so you feel every thump in your chest.
- Sudden spikes in blood pressure (hypertensive crisis): The tumor releases huge doses of adrenaline and noradrenaline in bursts, clamping down blood vessels so tightly that pressure can shoot sky-high in seconds.
- Pale skin during an attack: Adrenaline and noradrenaline squeeze the small blood vessels in the skin shut, shunting blood away from the surface and toward the heart and muscles, making the skin look white or gray.
- Shaking or tremor: High levels of adrenaline overstimulate the muscles, causing the fine shake you see when someone is very scared or jittery.

- A family history of pheochromocytoma or related tumors: About one in three cases are caused by inherited gene mutations (such as in the RET, VHL, NF1, or SDH genes), so if a parent or sibling has it, you are at higher risk.
- Finding a lump (mass) on the adrenal gland by accident on a scan (adrenal incidentaloma): A CT or MRI done for another reason may show a tumor on the adrenal gland; any unexpected adrenal mass should be tested for pheochromocytoma because even a silent tumor can release dangerous bursts of hormones.

Focused exam

- Blood pressure: sudden spikes over 180/110 or high blood pressure that will not come down even with several medicines: The tumor dumps adrenaline and noradrenaline (the body's fight-or-flight chemicals) in bursts. Each burst squeezes blood vessels tight and drives pressure sky-high. This wild swing is the number-one clue.
- Drenching sweat during an attack, even when sitting still in a cool room: Adrenaline and noradrenaline turn on sweat glands hard. The patient looks like they just ran a mile, but they have not moved. The tumor is flooding the body with chemicals, not exercise.
- Fast heart rate (often over 100) or feeling the heart pounding in the chest: Adrenaline and noradrenaline tell the heart to beat faster and harder. During a surge from the tumor, the patient feels every thump and cannot slow it down by resting.
- Pounding headache that comes on suddenly during an attack: The spike in blood pressure stretches vessels in the brain. The headache is the brain feeling the pressure wave from the chemical dump.
- Shaking hands or sudden fear and panic during attacks: Adrenaline and noradrenaline flip the nervous system into alarm mode. The body trembles and feels terrified even though there is no real threat nearby.
- Lightheadedness or near-fainting when standing up quickly: The body gets used to constant high chemical levels and turns down its receptors. Between surges, blood pressure drops too low when standing, because the system cannot tighten vessels fast enough.

Diagnostics · every test answers a question

- Plasma free metanephrines (blood test drawn after the patient has rested lying down for 30 minutes) → Is the tumor making too much adrenaline and noradrenaline right now?
- 24-hour urine collection for fractionated metanephrines and catecholamines → Over a full day, is the body breaking down more adrenaline-type chemicals than normal?
- CT scan of the abdomen and pelvis with contrast, or MRI if CT contrast is risky → Where exactly is the tumor sitting?

- Genetic testing panel for MEN2, von Hippel-Lindau, NF1, and SDH mutations → Is this tumor part of an inherited syndrome that will cause more tumors or cancers later?
- Functional imaging with ^{123}I -MIBG scan or ^{68}Ga -DOTATATE PET scan → Is the tumor in more than one spot, or has it spread to lymph nodes or distant organs?

Treatment logic

- Alpha-blocker (phenoxybenzamine 10 mg twice daily, increased every 2-3 days to 20-40 mg twice daily, or doxazosin 2-8 mg daily) started at least 7-14 days before surgery → Blocks adrenaline and noradrenaline from squeezing blood vessels. Blood pressure stops spiking into dangerous ranges, and the heart does not have to fight against tight pipes. This prevents a with high blood pressure (hypertensive) crisis when the surgeon touches the tumor.
- Beta-blocker (propranolol 10-40 mg three times daily or metoprolol 25-100 mg twice daily) started only AFTER alpha-blockade is working well → Slows the heart rate and makes each beat gentler, so the heart does not burn out from racing all day. Giving beta-blockers first would leave vessels squeezed with no outlet, causing a stroke or heart attack, so alpha MUST come first.
- Surgical removal of the tumor (laparoscopic adrenalectomy for small single tumors under 6 cm; open surgery for larger, bilateral, or malignant tumors) → Cuts out the cells that are leaking adrenaline and noradrenaline. Once the tumor is gone, chemical surges stop, blood pressure normalizes, sweating and headaches disappear, and the patient is cured if the tumor was benign.
- Generous IV normal saline (1-2 liters before surgery and continued after tumor removal) to expand blood volume → Refills the blood volume that was squeezed down by months of high chemical levels. This prevents a dangerous blood pressure crash when the tumor is removed and the squeezing suddenly stops.
- Calcium channel blocker (nifedipine extended-release 30-90 mg daily or amlodipine 5-10 mg daily) added if blood pressure stays high despite alpha- and beta-blockade → Relaxes blood vessel muscle through a different chemical door (blocks calcium entry into vessel walls). Brings down stubborn high pressure when adrenaline blockade alone is not enough.
- Lifelong annual biochemical screening (plasma or urine metanephrines) and imaging every 1-2 years after surgery → Catches tumor recurrence early (10-15 percent come back) or new tumors in hereditary cases before symptoms start. Early detection means simpler, safer repeat surgery.

Medications

Phenoxybenzamine · Non-selective alpha blocker

Blocks alpha receptors (tiny switches on blood vessel walls that tighten the vessel when hit by adrenaline or noradrenaline). By sitting on those switches, it stops the tumor's flood of catecholamines from squeezing blood vessels shut, so blood pressure stays down and blood flow stays smooth.

1. The pheochromocytoma tumor dumps huge waves of noradrenaline and adrenaline into the bloodstream.
2. Those catecholamines normally lock onto alpha receptors on blood vessel walls, making the muscle squeeze and blood pressure spike.
3. Phenoxybenzamine binds tightly (irreversibly) to both alpha-1 and alpha-2 receptors, blocking the catecholamines from landing.
4. Without the catecholamine signal getting through, the blood vessels relax, blood pressure drops, and the wild swings calm down.
5. This pre-surgery alpha blockade also lets the body refill its blood volume — the high catecholamines had been squeezing vessels so tight that total blood volume shrank; blocking that constriction lets fluid shift back into the bloodstream.

SAFETY WATCH

Must start 7–14 days before surgery to give blood volume time to recover and blood pressure time to stabilize. · Watch for orthostatic low blood pressure (hypotension) (dizziness when standing up) — the body was used to super-high pressure, so blocking alpha suddenly can make standing pressure drop. · Nasal congestion and fatigue are common because alpha receptors also help keep nasal blood vessels tight and help you stay alert. · NEVER give a beta blocker first — if you block the heart's beta receptors while alpha receptors are still wide open to catecholamines, the unopposed alpha constriction can cause a with high blood pressure (hypertensive) crisis.

Doxazosin · Selective alpha-1 blocker

Blocks only alpha-1 receptors (the switches on blood vessel muscle that tighten the vessel). By covering those switches, it stops the tumor's adrenaline and noradrenaline from spiking blood pressure, but it leaves alpha-2 receptors alone so some feedback brakes stay on.

1. The pheochromocytoma releases surges of catecholamines that hit alpha-1 receptors on blood vessel walls.
2. Doxazosin sits on those alpha-1 receptors (reversibly), so the catecholamines cannot bind and squeeze the vessels.

3. Blood vessels relax, resistance drops, and blood pressure comes down.
4. Unlike phenoxybenzamine, doxazosin does NOT block alpha-2 receptors, which means the body's natural feedback loop (alpha-2 on nerve endings that tells the nerves to release less noradrenaline) stays partly intact.
5. Volume expansion happens as vessels relax and the body shifts fluid back into the bloodstream.

SAFETY WATCH

Start 7–14 days before surgery to allow blood pressure and volume to stabilize. · First-dose low blood pressure (hypotension) can happen — the first pill may drop pressure more than later doses, so take it at bedtime. · Orthostatic dizziness is common as the body adjusts to lower pressure. · NEVER start a beta blocker before adequate alpha blockade — unopposed alpha stimulation during a catecholamine surge can cause stroke-level blood pressure spikes.

Propranolol or metoprolol (beta blocker) · Non-selective (propranolol) or selective beta-1 (metoprolol) blocker

Blocks beta receptors on the heart (beta-1) and sometimes the lungs and blood vessels (beta-2 if non-selective). This slows the heart rate and cuts the force of each heartbeat, calming the pounding and racing caused by the tumor's catecholamine flood. MUST be started only AFTER alpha blockade is solid.

1. Alpha blockade is already in place, so blood vessels are relaxed and volume is refilled.
2. The tumor's catecholamines still hit beta receptors on the heart, making it race and pound.
3. The beta blocker covers those beta receptors, so adrenaline and noradrenaline cannot speed up the heart or make it squeeze harder.
4. Heart rate and contractility drop, lowering the work the heart does and smoothing out the dangerous swings in blood pressure and pulse.

SAFETY WATCH

Start ONLY after 7–14 days of alpha blockade and adequate volume expansion. · Watch for a slow heart rate (bradycardia) (heart rate too slow) or heart block if the dose is too high. · In patients with asthma or COPD, non-selective beta blockers (propranolol) can trigger bronchospasm — use a selective beta-1 blocker (metoprolol) carefully or avoid if lung disease is severe. · NEVER start before alpha blockade — unopposed alpha stimulation during a catecholamine surge can cause stroke or heart attack.

Amlodipine or nifedipine (calcium channel blocker) · Dihydropyridine calcium channel blocker

Blocks calcium channels in the smooth muscle of blood vessel walls. Calcium is the key that lets muscle cells squeeze; blocking it keeps the vessels relaxed. Used when alpha and beta blockers alone do not control blood pressure swings.

1. Even with alpha and beta blockade, some patients still have blood pressure spikes because catecholamine surges can activate other pathways.
2. Calcium channel blockers prevent calcium from entering smooth muscle cells in vessel walls.
3. Without calcium, the muscle cannot contract tightly.
4. Vessels stay dilated, resistance drops, and blood pressure falls further.

SAFETY WATCH

Can cause ankle swelling (peripheral swelling (edema)) because dilated vessels leak a bit of fluid into tissues. · Headache and flushing are common as vessels dilate. · Watch blood pressure closely — stacking blockers can sometimes drop pressure too low.

Intravenous normal saline (liberal fluids and salt) · Volume expander (supportive therapy)

Refills the bloodstream with salt water. The tumor's chronic high catecholamines squeezed blood vessels so hard that total blood volume shrank (the body sensed high pressure and dumped fluid). Alpha blockade relaxes vessels, and the body needs extra volume to fill the newly opened space and avoid a pressure crash during surgery.

1. High catecholamines cause intense vasoconstriction, which tricks the body into thinking blood volume is too high.
2. The kidneys respond by dumping salt and water, so total blood volume drops.
3. When you start alpha blockade, vessels relax and the vascular space suddenly gets bigger.
4. Giving IV saline (and encouraging salty foods and fluids by mouth) refills that space so blood pressure stays stable and organs get enough flow.
5. This volume loading is critical in the 7-14 days before surgery to prevent shock when the tumor is removed and catecholamine levels plummet.

SAFETY WATCH

Monitor for fluid overload (swelling, shortness of breath) if the patient has heart failure or kidney disease, though this is rare in typical pheochromocytoma patients. · Check blood pressure sitting and standing — the goal is to see orthostatic drop shrink as volume refills.

Red flags

- Blood pressure spiking suddenly to dangerously high levels (hypertensive crisis), which can cause stroke or heart attack
- Chest pain or trouble breathing during an attack, suggesting the heart is being overworked
- Severe headache with confusion, vision changes, or weakness, which can mean bleeding in the brain from very high pressure
- Episodes triggered by pressing on the belly, surgery, or certain medicines (beta blockers alone), which can provoke a life-threatening surge

Nursing priorities

1. Monitor blood pressure every 15 to 30 minutes during with high blood pressure (hypertensive) episodes and notify the provider immediately if systolic pressure goes above 180 or diastolic above 120. — Catecholamine surges cause sudden, severe vasoconstriction (blood vessel narrowing) that can push systolic pressure over 200 mmHg. This level of high blood pressure (hypertension) can cause stroke (brain bleed), heart attack (myocardial infarction) (heart attack), or aortic dissection (tear in the main artery). Early detection and rapid treatment with IV phentolamine (an alpha-blocker) or nitroprusside can prevent organ damage.
2. Verify that the patient is on alpha-blocker medication like phenoxybenzamine or doxazosin for at least 7 to 14 days before any surgery is scheduled, and confirm beta-blockers were started only after alpha-blockade. — Alpha-blockade must happen first to prevent unopposed alpha-receptor stimulation. If you give a beta-blocker first, it blocks the beta-2 receptors (which cause blood vessel relaxation), leaving only alpha receptors active. This causes extreme vasoconstriction and with high blood pressure (hypertensive) crisis. Adequate preoperative alpha-blockade prevents intraoperative (during surgery) blood pressure swings that can be fatal.
3. Keep the patient in a calm, quiet environment and avoid sudden position changes, abdominal examining by touch (palpation), or any procedure that puts pressure on the abdomen. — Physical pressure or manipulation of the tumor causes mechanical catecholamine release, triggering a with high blood pressure (hypertensive) spell. The adrenal glands sit on top of the kidneys in the back of the abdomen, so even deep examining by touch (palpation) (pressing) during an exam can squeeze the tumor. Sudden position changes can also shift abdominal organs and compress the mass. Keeping the environment calm reduces endogenous (internal) catecholamine surges from stress.

4. Administer IV normal saline at the rate ordered (often 2 to 3 liters over 24 hours) before surgery and continue liberal fluids postoperatively for at least 48 hours. — Chronic catecholamine excess causes vasoconstriction that shrinks intravascular volume (the amount of fluid in blood vessels). Patients are often volume-depleted (low on fluid) even though their blood pressure is high. After the tumor is removed, blood vessels suddenly relax (vasodilation), and without enough fluid on board, blood pressure can crash (severe low blood pressure (hypotension)). Preloading with saline expands blood volume and prevents postoperative shock.
5. Monitor continuous cardiac telemetry (heart rhythm monitor) and watch for arrhythmias like atrial fibrillation (an irregular, quivering heartbeat), ventricular a fast heart rate (tachycardia), or premature ventricular contractions. — Excess catecholamines have direct toxic effects on the myocardium (heart muscle) and increase electrical irritability, causing dangerous arrhythmias. High norepinephrine (a catecholamine) also increases myocardial oxygen demand while constricting coronary arteries (the heart's blood supply), which can lead to starved blood flow (ischemia) (not enough oxygen) and arrhythmias. Telemetry allows instant detection so we can treat life-threatening rhythms with antiarrhythmics or cardioversion (shock to reset rhythm).
6. Check blood glucose every 4 to 6 hours and treat high blood sugar (hyperglycemia) as ordered, knowing that glucose levels may drop suddenly after tumor removal. — Catecholamines inhibit (block) insulin release from the pancreas and increase gluconeogenesis (making new glucose in the liver), causing high blood sugar (hyperglycemia) (high blood sugar). After surgery removes the source, insulin production rebounds and glucose can plummet (reactive low blood sugar (hypoglycemia)), especially in the first 24 to 48 hours. Monitoring prevents both hyperglycemic complications before surgery and dangerous low blood sugar after.
7. Teach the patient to avoid tyramine-rich foods like aged cheese, cured meats, and red wine, and to tell all providers about this diagnosis before any procedure or new medication. — Tyramine (found in aged or fermented foods) can trigger catecholamine release and precipitate (cause) a with high blood pressure (hypertensive) crisis in pheochromocytoma patients. Many medications—including certain anesthetics, opioids like morphine, and metoclopramide (a nausea drug)—can also stimulate catecholamine release or interact dangerously with alpha-blockers. Patient education prevents accidental triggers and ensures safe care across all settings.

8. Coordinate genetic counseling and testing for hereditary syndromes like MEN2, von Hippel-Lindau, NF1, and SDH mutations, and document family history for screening recommendations. — Because 30 to 40 percent of pheochromocytomas are hereditary, finding a germline mutation (inherited gene change) changes management for the patient (lifelong surveillance for recurrence or other tumors) and identifies at-risk family members who need screening. For example, MEN2 patients need thyroid monitoring for medullary thyroid cancer, and von Hippel-Lindau patients need kidney and pancreas imaging. Early detection in relatives saves lives.

Sources

- Endocrine Society Clinical Practice Guideline on Pheochromocytoma and Paraganglioma 2014
- National Comprehensive Cancer Network (NCCN) Neuroendocrine and Adrenal Tumors guideline
- UpToDate Clinical Overview of Pheochromocytoma
- American Heart Association (AHA) on Secondary Hypertension

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