BARE YOUR RARE

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PATIENT & CAREGIVER GUIDE

POEMS Syndrome

A rare disease where early treatment can reverse disability.

> A comprehensive, plain-language reference drawn from peer-reviewed literature and the world's leading POEMS registries. Designed to be read before — and brought to — your next specialist appointment.

P	O	E	M	S
Polyneuropathy	Organomegaly	Endocrinopathy	M-protein	Skin changes

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Contents

A roadmap through the guide. Skip to what matters most right now.

SECTION 01

What POEMS Syndrome Is

A rare, multi-system disease driven by a small group of abnormal plasma cells. It disables by stealth — and, often, responds dramatically to the right treatment.

POEMS syndrome is a rare disorder caused by a tiny population of abnormal plasma cells in the bone marrow. Those cells release an aberrant antibody fragment (the **M-protein**) and flood the body with inflammatory messenger molecules — most importantly **vascular endothelial growth factor (VEGF)**. The result is damage across many organ systems at once: nerves, organs, hormones, blood, skin, eyes, lungs, and blood vessels.

The disease is named for five of its most common features — **P**olyneuropathy, **O**rganomegaly, **E**ndocrinopathy, **M**-protein, and **S**kin changes. But that acronym, coined in 1980, captures only part of the picture. Pulmonary hypertension, thrombosis, kidney involvement, and eye findings are now known to be common and clinically important.

POEMS has also been called *Crow–Fukase syndrome*, *Takatsuki disease*, *PEP syndrome*, and *osteosclerotic myeloma*. In 2018 it was formally reclassified under the umbrella of **Monoclonal Gammopathy of Clinical Significance (MGCS)** — a framing that matters because it emphasizes what POEMS really is: a disease in which a small, biologically quiet plasma cell clone causes massive systemic harm through what it *signals*, not through what it destroys.

> **THE REASON THIS GUIDE EXISTS**
>
> POEMS is one of the rare diseases where early, accurate diagnosis and effective treatment can **reverse** disability that would otherwise be permanent. People who arrived at treatment using a wheelchair have walked out years later. That outcome depends on one thing: the right doctor recognizing what this is, fast enough.

SECTION 02

The Acronym — Unpacked

Five letters, five clinical fingerprints. Here's what each one actually means in a person's body.

P — Polyneuropathy

The neuropathy of POEMS is the feature that drives most patients to a doctor. It typically begins as numbness and tingling in both feet, then spreads upward and into the hands, becoming weakness that makes stairs, gripping, and balance progressively harder. It is **symmetrical**, **distal** (feet and hands first), and combines **sensory and motor** loss. Unlike in many other neuropathies, the pattern on nerve conduction testing is a **uniform demyelination** — a subtle but important clue that distinguishes POEMS from CIDP. Cranial and autonomic nerves are usually spared. Pain can be severe, especially at night.

O — Organomegaly

Up to half of patients have an enlarged liver, spleen, or lymph nodes. Enlargement is usually mild and causes no direct symptoms, but it is an important diagnostic clue — especially lymphadenopathy, because when a lymph node is biopsied it may show **Castleman disease**, a separate condition that overlaps with POEMS in ways that matter for treatment.

E — Endocrinopathy

Most patients have at least one hormonal abnormality, and many have several at once. The most common pattern is **hypogonadism** — low testosterone, erectile dysfunction, loss of libido, gynecomastia in men; menstrual irregularity or early menopause in women. Thyroid dysfunction, adrenal insufficiency, type 2 diabetes, hyperprolactinemia, and elevated parathyroid hormone are all reported. Diabetes and thyroid disease alone don't count toward the diagnostic criteria because they're common in the general population — but *multiple* endocrinopathies together strongly support POEMS.

M — Monoclonal protein

A small clone of plasma cells produces an abnormal antibody fragment — the M-protein. In POEMS, this is almost always **IgA or IgG with lambda light chains**. The level is usually low — so low, in fact, that in **30–46%** of patients the standard serum protein electrophoresis (SPEP) misses it entirely. Immunofixation catches more, but still misses an estimated 15–25%. In some patients the M-protein is only detectable in a **24-hour urine study** or through careful bone marrow examination. Repeat testing is often necessary.

S — Skin changes

Skin is affected in roughly half of patients and often earlier than people realize. The most common findings are **hyperpigmentation** (darkening of the skin, especially extensor surfaces), **hypertrichosis** (coarse hair growth on the face, arms, chest), **glomeruloid hemangiomas** (small firm red-purple bumps — considered nearly specific to POEMS), **white nails**, and **scleroderma-like skin thickening**. Sweating changes, Raynaud phenomenon, clubbing of the fingers, and leg edema are also common.

> **THE PEST EXTENSION**
>
> Some clinicians use a second acronym — **PEST** — to capture features that the POEMS name misses: **P**apilledema (swelling of the optic disc, in up to 30%), **E**xtravascular volume overload (edema, pleural effusion, ascites), **S**clerotic bone lesions, and **T**hrombocytosis or polycythemia. PEST features are formally part of the diagnostic criteria.

SECTION 03

Who Gets It

POEMS is rare, under-diagnosed, and more common than the published numbers suggest.

~0.3	50s	2:1
cases per 100,000 per year (estimated)	typical age at onset	male-to-female ratio

POEMS has been reported globally, with the largest cohorts published in Japan, the United States (Mayo Clinic), China (Peking Union Medical College), and the United Kingdom (UCLH Registry). Japanese cohorts suggest a slightly higher prevalence than Western ones, though this may reflect diagnostic vigilance rather than a true biological difference.

The **time from first symptom to diagnosis is typically 13 to 18 months** — and frequently longer. Multiple studies have found that patients see three or more specialists, and are often treated for chronic inflammatory demyelinating polyradiculoneuropathy (CIDP), diabetic neuropathy, Guillain-Barré, or another plasma cell disorder before the correct diagnosis is made.

Because POEMS is rarely fatal in the short term but frequently disabling, and because its early neuropathy is easily attributed to more common conditions, the published incidence almost certainly underestimates the true number of affected people.

SECTION 04

How It Causes Disease

A tiny clone, enormous downstream chaos. Understanding the mechanism explains why treatment works.

POEMS is a **paraneoplastic syndrome** — that is, the damage to the body is not caused by the tumor itself invading tissue, but by what the tumor *secretes*. The tumor in POEMS is a small clonal population of plasma cells, often so small that a bone marrow biopsy looks nearly normal. What matters is not the bulk of the clone but its biological output.

VEGF — the central driver

Vascular endothelial growth factor (VEGF) is a signaling protein that normally regulates blood vessel growth and leakage. In POEMS it is dramatically elevated — typically 5 to 10 times higher than in healthy people or in other neuropathies. The excess VEGF produces most of the characteristic features:

- **Endoneurial edema** — fluid leakage into and around nerves, disrupting signal conduction and driving demyelination.
- **Extravascular volume overload** — peripheral edema, pleural effusions, ascites.
- **Skin changes** — hyperpigmentation, hypertrichosis, vascular lesions.
- **Pulmonary vascular remodeling** — leading in some patients to pulmonary hypertension.
- **Papilledema** — swelling of the optic disc.

Other cytokines

VEGF acts alongside a cluster of pro-inflammatory cytokines — **interleukin-1β, interleukin-6, and TNF-α** — that are also consistently elevated. This mixture drives the systemic inflammation, the endocrine disruption, and the thrombocytosis and erythrocytosis seen in many patients.

Why this matters for treatment

Because the damage is *signaled* rather than *structural*, eliminating the small plasma cell clone turns off the signal. When that happens, VEGF levels fall rapidly — often within weeks — and the body begins to recalibrate. Peripheral nerves remyelinate. Edema resolves. Pulmonary pressures drop. Endocrine function partially recovers. This is the biological basis for one of the most remarkable features of POEMS: **reversibility**.

"Shut down the clone and the system recalibrates. A tiny cause. A vast collapse. A profound recovery — if caught in time."

The central clinical truth of POEMS

SECTION 05

Diagnostic Criteria

The Mayo Clinic / Dispenzieri criteria (updated 2023) are the global standard for POEMS diagnosis. A formal diagnosis requires meeting all three groups below.

Mandatory Criteria — BOTH Must Be Present

- Polyneuropathy (typically demyelinating)
- Monoclonal plasma cell proliferative disorder (almost always lambda light-chain restricted)

Other Major Criteria — At Least ONE Required

- Sclerotic bone lesions
- Elevated VEGF (often >1,000 pg/mL — but patients at ~850 pg/mL should not be excluded without further workup)
- Castleman disease

Minor Criteria — At Least ONE Required

- Organomegaly (splenomegaly, hepatomegaly, or lymphadenopathy)
- Extravascular volume overload (edema, pleural effusion, ascites)
- Endocrinopathy (excluding diabetes and thyroid disease alone)
- Skin changes (hyperpigmentation, hypertrichosis, glomeruloid hemangiomas, white nails)
- Papilledema
- Thrombocytosis or polycythemia

The Castleman Disease Variant

A recognized separate entity. It occurs **without** a clonal plasma cell disorder — lymph node biopsy shows Castleman histology, multiple POEMS minor criteria are present, but the M-protein and/or the polyneuropathy may be absent. These patients are classified and treated differently and have a somewhat different prognosis. The youngest reported case was age 6.

Diagnostic Workup — What Should Be Ordered

- Complete blood count with differential (looking for thrombocytosis, erythrocytosis)
- Comprehensive metabolic panel, liver function, serum albumin, eGFR
- Serum protein electrophoresis (SPEP) **plus immunofixation** (one without the other misses cases)

- 24-hour urine protein electrophoresis and immunofixation
- Free light chain assay
- Serum VEGF (order specifically — not part of standard panels)
- Bone marrow biopsy with flow cytometry
- Full skeletal survey **plus whole-body CT or PET/CT** (most POEMS lesions are <1 cm and best seen on CT bone windows)
- Nerve conduction studies / electromyography
- Echocardiogram (including estimated sPAP for pulmonary hypertension screening)
- Endocrine panel — testosterone (men), FSH/LH, prolactin, TSH, morning cortisol, HbA1c, PTH, vitamin D
- Ophthalmology referral for papilledema and macular edema
- Consider lymph node biopsy if adenopathy is present — rules in/out Castleman disease

> **KEY POINT**
>
> **A single negative SPEP does not rule out POEMS.** Up to 46% of patients have the M-protein missed on first testing. If clinical suspicion is strong, proceed to immunofixation, urine studies, bone marrow, and serum VEGF.

SECTION 06

The Misdiagnosis Problem

Most POEMS patients spend more than a year being treated for the wrong condition. The costs — in disability, time, and money — are not small.

The single most common misdiagnosis is **chronic inflammatory demyelinating polyradiculoneuropathy (CIDP)**. The neuropathies look similar on casual examination and on routine nerve conduction studies, and both involve demyelination. The distinguishing features are subtle:

Feature	POEMS Syndrome	CIDP
Demyelination pattern on NCS	Uniform along the nerve	Patchy, segmental
Motor vs sensory	Motor often predominates, distal	More balanced, proximal and distal
Response to IVIG or steroids	Minimal or none	Often substantial
M-protein	Present (IgA/IgG lambda); may be low-level	Absent
VEGF	Elevated, often dramatically	Normal
Systemic features	Endocrine, skin, organomegaly, edema	Typically absent

Other common misdiagnoses include **diabetic neuropathy** (especially in patients who also have the endocrinopathy of POEMS-induced diabetes), **Guillain-Barré syndrome**, **monoclonal gammopathy of undetermined significance (MGUS)** with incidental neuropathy, and **amyloidosis**.

RED FLAGS — WHEN TO RE-INVESTIGATE FOR POEMS

A patient being treated for CIDP, MGUS neuropathy, or diabetic neuropathy who has **not improved** with first-line therapy — especially if they have *any* of: unexplained endocrinopathy, papilledema, skin hyperpigmentation, new pleural effusion or ascites, thrombocytosis, or sclerotic bone lesions incidentally noted on imaging — deserves a dedicated POEMS workup.

SECTION 07

Beyond the Acronym — The Other Complications

The POEMS acronym was coined in 1980. Forty-plus years of research has revealed several complications just as clinically important as the original five — and often missed because they aren't in the name.

Pulmonary Hypertension — 27% to 48%

In the largest reported cohort (154 patients, Peking Union Medical College), 27% had pulmonary hypertension (estimated sPAP ≥50 mmHg) at diagnosis. Other series have reported up to 48%. PH significantly worsens survival: median overall survival of 54 months with PH versus not reached without PH. The good news: PH in POEMS is often **reversible** with successful treatment of the underlying disease. Every new POEMS diagnosis warrants an echocardiogram.

Thrombosis — 27% to 42%

A major and under-recognized complication. In the UCLH POEMS Registry (103 patients), 30% experienced at least one thrombotic event. In a Mayo Clinic cohort of 230 patients, the rate was 27%. Unlike most plasma cell disorders, **arterial events are slightly more common than venous**:

- **Stroke accounts for 26% of all thrombotic events** and 53% of arterial events
- Other arterial events: peripheral arterial occlusion, myocardial infarction, microvascular disease, limb ischemia, bowel ischemia, Budd-Chiari syndrome
- Venous events: deep vein thrombosis, pulmonary embolism, PICC-associated DVT during ASCT
- **Most thromboses occur before treatment begins**, during active disease — they are not primarily a treatment side effect
- Hyperprolactinemia has been newly identified as a venous thrombosis risk factor in POEMS

> **ANTICOAGULATION — A DIFFICULT BALANCE**
>
> Most expert centers use **prophylactic anticoagulation** during periods of active disease, peri-transplant, and in patients with multiple risk factors (thrombocytosis, polycythemia, hyperprolactinemia, low albumin). Decisions are individualized. Bleeding risk from thrombocytopenia during chemotherapy must be weighed against thrombotic risk. This discussion belongs at an expert center.

Renal Involvement

Kidney disease is present in roughly 15% of patients and more often under-recognized. The pathology is typically **membranoproliferative-glomerulonephritis-like (MPGN-like)**, driven by endothelial injury from VEGF excess and from M-protein deposition. Unlike AL amyloidosis, POEMS kidney disease usually does *not*

cause heavy proteinuria — the presentation is more often declining eGFR with bland urine. Importantly, renal involvement is *excluded* from formal diagnostic criteria, but reduced eGFR is a **negative prognostic factor** and should be monitored.

Cardiac Involvement

Beyond pulmonary hypertension, patients may have **left ventricular hypertrophy**, subclinical **biventricular dysfunction** on strain echocardiography, and **sleep-disordered breathing** driven by volume overload, pulmonary issues, and endocrine changes. Systolic dysfunction (cardiomyopathy) is reported though less common. Baseline echocardiography and consideration of a sleep study are reasonable in many patients.

Eye Findings

Papilledema (swelling of the optic disc) is present in up to 30% — sometimes without symptoms, sometimes causing headache or visual changes. Other findings include cystoid macular edema, serous macular detachment, and venous sinus thrombosis. Every new POEMS diagnosis should prompt an ophthalmology referral.

Hematologic Changes

Thrombocytosis (high platelets) is present in roughly half of patients and contributes to thrombotic risk. **Polycythemia** (elevated red cells) is less common but also reported. Anemia is relatively uncommon at presentation, in contrast to multiple myeloma.

SECTION 08

Risk Stratification & Prognosis

POEMS is not a uniformly dangerous disease — but it is disabling if untreated. Prognosis depends on specific, measurable factors.

Mayo Clinic and European groups have identified a list of prognostic factors that guide clinical decision-making. Risk factors associated with **worse overall survival** include:

- Low serum albumin (<3.2 g/dL)
- Reduced eGFR
- Pulmonary hypertension
- Pleural effusion or significant extravascular volume overload
- Age over 50
- Fingernail clubbing
- Reduced DLCO on pulmonary function testing

Patients with none of these features can expect long survival with treatment, often exceeding 14 years median. Patients with three or more risk factors have a meaningfully shorter median survival and should be managed at a dedicated expert center.

Risk stratification in POEMS is currently based on **clinical phenotype**, not on molecular markers — unlike multiple myeloma, there is no FISH or cytogenetic panel in routine use.

SECTION 09

Treatment — First-Line Options

Treatment aims to eliminate the plasma cell clone, shut down the cytokine signal, and allow the body to recover. Choice of therapy depends on whether the disease is localized or disseminated.

Localized Disease — Radiation Therapy

For patients with a **single or a few sclerotic plasmacytomas** and no disseminated bone marrow involvement, **radiation therapy at curative doses (approximately 40–50 Gy)** to the affected lesion(s) is the first-line treatment. This approach can produce durable remission and sometimes cure. About half of radiation-treated patients achieve long-term disease control.

Disseminated Disease — High-Dose Chemotherapy with ASCT

For patients with diffuse bone marrow involvement, multiple lesions, or progression after radiation, **high-dose melphalan with autologous stem cell transplantation (ASCT)** is the standard of care in medically fit patients. This is the treatment with the most robust long-term data in POEMS.

~90%	~80%	>70%
hematologic response rate with ASCT	5-year overall survival with ASCT	of patients show neurologic improvement

A critical point: patients should be evaluated carefully and **stabilized before transplant**. Engraftment syndrome and capillary leak can be dangerous in patients with uncontrolled disease. Some centers use **induction therapy** (for example, a short course of lenalidomide-dexamethasone, bortezomib-dexamethasone, or daratumumab-based therapy) before proceeding to ASCT, specifically to reduce VEGF and stabilize volume status.

Patients Not Eligible for ASCT

For older patients or those with comorbidities that preclude transplant, options include:

- **Low-dose melphalan with dexamethasone** — an older but effective regimen
- **Lenalidomide with dexamethasone (Rd)** — generally well tolerated, good neurologic response
- **Bortezomib-based regimens** — effective, but peripheral neuropathy risk requires careful monitoring given the pre-existing POEMS neuropathy
- **Daratumumab-based regimens** — increasingly used (see next section)

THALIDOMIDE — A CAUTIONARY NOTE

The J-POST randomized controlled trial confirmed that thalidomide lowers VEGF and can help control POEMS. However, **23% of patients developed new or worsening peripheral neuropathy** on thalidomide. Lenalidomide is generally preferred for this reason.

BEVACIZUMAB — DO NOT USE OUTSIDE RESEARCH SETTINGS

Because VEGF is elevated, early investigators tried the anti-VEGF monoclonal antibody bevacizumab. Results were poor and **at least three patient deaths have been reported**. Bevacizumab is no longer recommended and should not be used outside of carefully designed clinical trials.

SECTION 10

Newer Therapies & The Horizon

The treatment landscape is broadening. Several newer agents have shown excellent activity in POEMS — expanding what's possible for relapsed patients and for those who can't undergo transplant.

Daratumumab-Based Regimens

Daratumumab is an anti-CD38 monoclonal antibody originally approved for multiple myeloma. Mayo Clinic has published the largest experience to date using **daratumumab, carfilzomib, and pomalidomide** combinations in POEMS — reporting deep and durable responses in both newly diagnosed and relapsed patients. **Daratumumab with lenalidomide and dexamethasone (DRd)** is emerging as a highly effective frontline option, especially for patients not immediately transplant-eligible. These regimens are increasingly used as induction before ASCT to reduce VEGF rapidly.

Carfilzomib

A second-generation proteasome inhibitor. Unlike bortezomib, carfilzomib does **not** cause peripheral neuropathy — a significant advantage in a disease that already has neuropathy as a defining feature. It has become an important option in relapsed disease.

Pomalidomide

A third-generation immunomodulatory drug (IMiD), effective in relapsed POEMS when lenalidomide has failed. Generally well tolerated. Often used in combination with daratumumab and dexamethasone.

BCMA Bispecific Antibodies

The BCMA/CD3 bispecific antibody **CM336** has entered clinical trials specifically in POEMS syndrome — one of the first targeted immunotherapy trials in this disease. BCMA-targeted bispecifics and CAR-T cell therapies have transformed relapsed myeloma care, and early signals in POEMS are encouraging. Enrollment is ongoing at select centers.

Supportive Therapies Aimed at VEGF Downstream Effects

Agents targeting VEGF downstream effects — including selective vascular stabilization strategies — are being studied. As of this writing, none are in routine clinical use.

SECTION 11

What Recovery Looks Like

Recovery from POEMS is slow, non-linear, and deeply personal — but the data on what's possible is genuinely extraordinary.

> *"In a Mayo Clinic series of 60 POEMS patients who underwent ASCT, 45% required a wheelchair before transplant. At follow-up, that number was zero."*
>
> Mauermann et al., *Neurology* 2015

Chiba University reported similar outcomes — chairbound patients walking again, often years after transplant. The Kuwabara group documented measurable nerve conduction velocity improvement in most patients, with continued gains over 24 months.

Realistic Recovery Timeline

- **0–3 months** VEGF drops rapidly. Edema, ascites, and pleural effusions resolve. Skin changes begin to fade. Papilledema improves. Fatigue may still be severe.
- **3–6 months** Pain often improves. Endocrine dysfunction begins to recover — testosterone can rise, thyroid function normalize. Some motor improvement begins. Most patients feel meaningfully better.
- **6–12 months** Peripheral nerves begin to remyelinate. Motor strength and sensation improve in a bottom-up pattern. Many patients regain the ability to walk without assistance.
- **12–24 months** Continued nerve recovery. Pulmonary hypertension often reverses. Kidney function can improve. Many return to work.
- **2–5 years** Slow ongoing gains. Distal sensation (especially in the feet) is often the last to return and may remain incomplete.

WHAT TO EXPECT

Recovery is not linear and is often punctuated by plateaus. Physical therapy matters enormously — maintained range of motion, graded strengthening, and aerobic conditioning all support nerve recovery. Mental health support is essential: the psychological weight of delayed diagnosis, lost function, and slow recovery is real.

SECTION 12

Supportive Care & Monitoring

Treating the clone is half of POEMS care. The other half is managing everything it broke.

Thromboprophylaxis

Individualized, but most expert centers use prophylactic anticoagulation during active disease and peri-transplant. Aspirin alone is generally considered insufficient given the arterial event rate.

Neuropathy — Symptomatic Care

- **Physical therapy** from the time of diagnosis onward — stretching, strengthening, balance training
- **Orthotics** (ankle-foot orthoses, for example) to prevent foot drop and support gait
- **Gabapentin, pregabalin, duloxetine, or tricyclic antidepressants** for neuropathic pain
- **Occupational therapy** for upper limb function and activities of daily living

Pulmonary & Sleep

Patients with pulmonary hypertension or sleep-disordered breathing benefit from **CPAP or BiPAP** where indicated. Pulmonology follow-up is important, especially in the first 12 months after treatment, as PH often reverses.

Endocrine

Replacement therapy as needed — testosterone for men with persistent hypogonadism, thyroid hormone for hypothyroidism, cortisol for adrenal insufficiency, insulin or oral agents for diabetes. Bone density scans are appropriate given the risk of secondary osteoporosis.

Monitoring Schedule

Interval	Tests
Every 3 months (first 2 years)	CBC, CMP, SPEP + immunofixation, free light chains, serum VEGF, physical exam
Every 6 months	Nerve conduction studies, echocardiogram (if prior PH or risk factors)
Annually	PET/CT or skeletal survey, ophthalmology exam, endocrine panel, pulmonary function testing
As indicated	Bone density scan, sleep study, renal referral if eGFR declining

After 2 years of sustained remission, the interval can often be extended to every 6 months, then annually.

SECTION 13

Expert Centers

POEMS is rare enough that outcomes depend heavily on clinician familiarity. Every patient should be seen, at least once, at a center with a dedicated POEMS program.

North America

Mayo Clinic (Rochester, MN) — The global reference center for POEMS care and research. Dr. Angela Dispenzieri's group has published the modern diagnostic criteria and most of the large cohort data on treatment outcomes, thrombosis, and newer therapies.

Cleveland Clinic — Active consultative hematology program with published POEMS experience.

Dana-Farber Cancer Institute (Boston) — Plasma cell disorder program with POEMS expertise.

Europe

University College London Hospitals (UCLH) — Maintains the UCLH POEMS Registry (103+ patients), the largest single-center European cohort. Published the definitive thrombosis data in POEMS.

Assistance Publique – Hôpitaux de Paris — Strong French POEMS network, with particular expertise in neurologic recovery.

Asia

Peking Union Medical College Hospital (Beijing) — The largest Chinese POEMS center. Published the definitive data on pulmonary hypertension prevalence and cardiac function in POEMS.

Chiba University (Japan) — Leading center for nerve regeneration studies after ASCT and for novel antibody therapies.

National Cancer Center (Tokyo) — Strong plasma cell disease program with POEMS experience.

> **HOW TO GET REFERRED**
>
> Ask your hematologist to request a **remote consultation** (second opinion) from one of these centers. Mayo and UCLH both accept international remote consultations and can review outside imaging and records. This is often the single most impactful step a newly diagnosed patient can take.

SECTION 14

Your Action Checklist

Concrete, ordered steps — for wherever you are in the journey.

If You Think You Might Have POEMS

- Ask your doctor for SPEP *plus* immunofixation, *plus* a 24-hour urine protein study, *plus* serum free light chains, *plus* serum VEGF
- Ask about a skeletal survey or PET/CT looking specifically for sclerotic bone lesions
- Ask whether an echocardiogram and a formal endocrine panel are warranted
- If you are being treated for CIDP and have not responded to IVIG or steroids — request a POEMS workup

If You Have Just Been Diagnosed

- Request a remote consultation with an expert center (Mayo, UCLH, Peking Union, Chiba)
- Get a baseline echocardiogram, ophthalmology exam, nerve conduction study, and full endocrine panel before starting treatment
- Ask about thromboprophylaxis during active disease
- Begin physical therapy early — don't wait for treatment to start
- Connect with a patient community (Bare Your Rare, POEMS Syndrome International)

If You Are in Treatment or Recovery

- Maintain the monitoring schedule (Section 12) — relapse is detectable early if looked for
- Track serum VEGF, M-protein level, and nerve conduction over time
- Report new symptoms promptly — new edema, new neuropathy, new endocrine changes
- Address mental health openly — many patients experience post-treatment depression or anxiety
- Plan exercise, sleep, and nutrition deliberately; the body is rebuilding

For Family & Caregivers

- Understand the recovery timeline is measured in years, not weeks
- Learn to recognize signs of relapse — returning edema, new neuropathy, new endocrine symptoms
- Help with logistics: appointment coordination, records management, insurance
- Take care of yourself too — caregiver burnout is real and common in rare disease

SECTION 15

Glossary

Plain-language definitions for key terms used in POEMS care.

ASCT (Autologous Stem Cell Transplant) — High-dose chemotherapy followed by reinfusion of the patient's own stem cells. The standard of care for disseminated POEMS in fit patients.

Castleman Disease — A separate lymphoproliferative disorder that can overlap with POEMS. About 15–24% of POEMS patients also have Castleman disease; a Castleman variant of POEMS exists without a clonal plasma cell disorder.

CIDP — Chronic inflammatory demyelinating polyradiculoneuropathy. The condition most often confused with POEMS.

Cytokine — A signaling protein released by cells to communicate with other cells. In POEMS, VEGF, IL-1β, IL-6, and TNF-α are all elevated.

Demyelination — Damage to the myelin sheath that insulates nerves, slowing or blocking signal conduction.

Endocrinopathy — Any disease of the endocrine (hormone-producing) glands.

Immunofixation — A specialized blood or urine test that identifies the specific type of monoclonal protein. More sensitive than SPEP alone.

M-protein (Monoclonal Protein) — An abnormal antibody fragment produced by a clone of plasma cells.

MGCS (Monoclonal Gammopathy of Clinical Significance) — A class of diseases in which a small monoclonal gammopathy causes organ damage out of proportion to the clone's size. POEMS is now formally classified here.

Organomegaly — Enlargement of internal organs, most often liver, spleen, or lymph nodes.

Papilledema — Swelling of the optic disc at the back of the eye, visible on ophthalmoscopy.

Paraneoplastic syndrome — A disease caused not by a tumor directly invading tissue, but by what the tumor secretes.

Plasmacytoma — A localized tumor of plasma cells. In POEMS these are typically sclerotic (hardened) and found in bone.

Polyneuropathy — Damage to multiple peripheral nerves at once, typically symmetric.

Pulmonary Hypertension (PH) — Elevated blood pressure in the pulmonary arteries. Present in 27–48% of POEMS patients.

SPEP (Serum Protein Electrophoresis) — A standard blood test that separates proteins by size and charge. Detects large M-proteins but misses 30–46% of POEMS M-proteins.

VEGF (Vascular Endothelial Growth Factor) — A signaling protein that regulates blood vessel growth and permeability. The central driver of POEMS pathology.

SECTION 16

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All information in this guide is drawn from peer-reviewed medical literature. Verified as of April 2026.

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You Found This Place.

Bare Your Rare exists because patient experience is expertise. This guide was built to make what took a community of patients, clinicians, and researchers decades to learn, findable in one evening — for the person who needs it tonight.

Share it freely with your specialist, your family, or anyone else navigating this diagnosis.

`bareyourrare.org/stories/poems-syndrome`

Bare Your Rare — a patient-built rare disease platform.
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This guide is educational and does not replace medical advice from your own doctor.
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