# Polymyalgia Rheumatica:
## Diagnosis and Management

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Dr Rukhsana Hussain

PenninegGPLearning

# OBJECTIVES

- Present an Overview of Polymyalgia Rheumatica (PMR) as a condition
- Revise the diagnostic criteria for PMR
- Review the recommended investigations prior to making the diagnosis
- Summarise guidelines regarding management
- Outline specialist referral criteria

# OVERVIEW[5]

**Polymyalgia rheumatica** is a chronic, systemic rheumatic inflammatory disease characterized by aching and morning stiffness in the neck, shoulder, and pelvic girdle in people older than 50 years of age.

**The cause of polymyalgia rheumatica (PMR) is unknown,** although genetic and environmental factors are thought to contribute to disease susceptibility and severity.

**Polymyalgia rheumatica (PMR) is the most common inflammatory rheumatic disease in older people,** and one of the most common indications for long-term corticosteroid treatment in the UK, accounting for 22% of prescriptions.

# OVERVIEW- Risk factors[5]

**Older age** The highest incidence is in people older than 65 years of age, with a peak in the 70–80 year age group. It is seldom diagnosed in people younger than 50 years of age

**Female gender** Over 65% of people who have PMR are women, and the lifetime risk for developing PMR is estimated at 2.4% for women and 1.7% for men

**Northern European ancestry** PMR is most common in people of Northern European ancestry, with an incidence of 41–113 cases per 100,000 persons and a prevalence of about 6 cases per 1000 persons in those older than 50 years. It is uncommon in people of Middle Eastern, Asian, African, and Hispanic descent

**Infection** Cyclic fluctuations and peaks in incidence have been observed in the winter and associated with epidemics of mycoplasma, chlamydia pneumonia, and parvovirus B19 infections

# OVERVIEW- Complications[5]

**Giant cell arteritis (GCA) and its complications** can occur, abruptly and without warning, early in the course of polymyalgia rheumatica (PMR)

About 15–20% of people with PMR develop GCA, and 40–50% of people with GCA have symptoms of PMR

**Complications of long-term corticosteroid treatment** are common and may occur in up to 50% of people

# OVERVIEW- Prognosis[5]

**The overall prognosis of polymyalgia rheumatica (PMR) is good.**

Response to systemic corticosteroids is rapid and dramatic, with many symptoms resolving within 24–72 hours.

However:

- About 29–45% of people with PMR do not adequately respond to systemic corticosteroids within 3–4 weeks
- Treatment for 1–2 years is often required
- Relapse is common but responds to restarting or increasing the dose of systemic corticosteroids. Less commonly, there is a chronic relapsing course that may require a longer course of treatment

# OVERVIEW- Prognosis[5]

PMR is not associated with increased mortality but morbidity and mortality may occur as a result of corticosteroid adverse effects.

prognosis is better in men than in women[3]

higher inflammatory markers are associated with longer duration of therapy and more relapse[3]

# CHALLENGES IN THE DIAGNOSIS OF POLYMYALGIA RHEUMATICA[1]

The cardinal features of PMR – proximal pain and stiffness with a raised acute phase response – are familiar to most physicians.

However, the diagnostic presentation is varied and this may lead to diagnostic error:

- proximal pain and stiffness can occur in many other illnesses
- a third of patients have systemic symptoms such as fever, anorexia and weight loss
- a considerable number (15–30%) may have distal musculoskeletal manifestations such as peripheral arthritis, distal swelling with pitting oedema and carpal tunnel syndrome.

# CHALLENGES IN THE DIAGNOSIS OF POLYMYALGIA RHEUMATICA[1]

PMR is also associated with giant cell arteritis (GCA) in 10% of the cases and up to 50% of cases of GCA may have polymyalgic symptoms at presentation.

Moreover, an acute phase response can occur in other settings such as other rheumatological conditions, neoplasia and infection.

# CHALLENGES IN THE DIAGNOSIS OF POLYMYALGIA RHEUMATICA[1]

Many clinicians and two empirically developed sets of diagnostic criteria use a response to corticosteroids as the main defining feature of this condition.

This, too, may encourage diagnostic error, since corticosteroids are potent anti-inflammatory agents that can mask symptoms from a host of serious conditions ranging from osteoarthritis, rotator cuff problems, rheumatoid arthritis, cancer and infection, especially if used in high doses and for protracted lengths of time.

# DIAGNOSIS[1]

Recommended stepwise approach:

1. Core inclusion criteria
2. Core exclusion criteria
3. Exclude mimicking conditions
4. Assessment of response to steroids (15mg prednisolone)
5. Confirmation of diagnosis at early follow up

# CORE INCLUSION CRITERIA[1]

- Age > 50 years
- Bilateral shoulder or pelvic girdle aching or both
- Morning stiffness >45mins
- Duration of symptoms > 2 weeks
- Abrupt onset
- Evidence of acute phase response (increased ESR/CRP)

# CORE INCLUSION CRITERIA- Notes[5]

**Bilateral shoulder and/or pelvic girdle pain.** Initially this may be unilateral but quickly becomes bilateral, is worse with movement, and interferes with sleep. Shoulder pain may radiate to the elbows and is the presenting feature in 70–95% of people.

Hip and neck pain is the presenting feature in 50–70% of people. Hip pain may radiate to the knees.

**Stiffness lasting for at least 45 minutes** after waking or periods of rest that may cause the person to have difficulty turning over in bed, rising from a bed or a chair, or raising their arms above shoulder height.

# CORE INCLUSION CRITERIA- Notes[5]

**Additional features that may accompany these core symptoms include:** Low-grade fever, fatigue, anorexia, weight loss, and depression - systemic symptoms occur in 40–50% of people with PMR.

Bilateral upper arm tenderness - sometimes present.

Peripheral musculoskeletal signs - seen in approximately 50% of people and include:

- Carpal tunnel syndrome.
- Peripheral arthritis (predominantly affecting the knees and wrists), which is asymmetric and self-limiting.
- Swelling with pitting oedema of hands, wrists, feet, and ankles.

# CORE INCLUSION CRITERIA- Notes[5]

**Muscle strength is not usually impaired**, but muscle pain may make testing difficult. If symptoms are protracted, disuse atrophy of muscle can occur, leading to muscle weakness.

# CORE EXCLUSION CRITERIA AND MIMICKING CONDITIONS[1]

- **Giant Cell/Temporal Arteritis**
- **Active Cancer**
- **Infection** e.g. TB, Brucellosis
- *Other inflammatory conditions*:

  Rheumatoid Arthritis, Other Arthropathies, SLE, myopathies and other connective tissue disease

# CORE EXCLUSION CRITERIA AND MIMICKING CONDITIONS[1]

- *Non-inflammatory conditions*: Local shoulder and hip conditions, Fibromyalgia/pain syndromes, Osteoarthritis
- *Endocrine conditions* e.g. Hypothyroidism
- *Drug induced* e.g. Statins – statin related myalgia/myopathy
- *Bone disease* e.g. Osteomalacia, Paget's disease
- Parkinsonism

# TEMPORAL ARTERITIS[1]

- Needs urgent high dose steroids (40-60mg prednisolone)
- Abrupt onset headache (usually temporal) + temporal tenderness
- Visual disturbance (inc diplopia)
- Jaw/tongue claudication
- Upper cranial nerve palsies
- Prominence/beading/diminished pulsation temporal artery

# RECOMMENDED INVESTIGATIONS PRIOR TO TREATMENT WITH STEROIDS[1]

- FBC
- ESR/CRP/Plasma Viscosity – usually elevated
- U&Es
- LFTs
- Bone profile/Vitamin D
- Protein electrophoresis (consider urine Bence-Jones protein also)
- TFTs
- CK
- Rheumatoid factor (should be negative)
- ANA and anti-CCP may be considered
- Dipstick urinalysis
- CXR (e.g. Prominent systemic symptoms)

# ASSESSMENT OF RESPONSE TO STEROIDS[5]

**If PMR is the most likely diagnosis:**

Prescribe a trial of oral prednisolone 15 mg daily, and arrange follow up after 1 week to assess clinical response.

**Make a working diagnosis of PMR if there is a patient-reported global improvement of 70% or more within a week, and normalization of inflammatory markers within 4 weeks.**

If there is a lesser response, consider increasing the dose to 20 mg prednisolone and reassess response.

If, despite increasing the dose of prednisolone, response is still less than 70%, reconsider the diagnosis and refer to an appropriate specialist.

# MANAGEMENT[5]

**For people in whom a working diagnosis of Polymyalgia Rheumatica (PMR) has been made:**

- **Reduce the dose of prednisolone slowly** when symptoms are fully controlled.
- **Ensure the person is provided with a blue steroid card**, and discuss potential adverse effects of corticosteroids. In particular, advise them:
  - Not to stop taking prednisolone abruptly and to seek medical advice if they are experiencing problems taking it.
  - To avoid close contact with people who have chickenpox, shingles, or measles if they do not have immunity to chickenpox or measles and to seek medical advice if they are exposed.

# MANAGEMENT[5]

- **Provide written information** on PMR and regional patient support groups. Polymyalgia Rheumatica and Giant Cell Arteritis (PMRGCA) UK (www.pmrgcauk.com) provide information packs, a helpline, newsletters, support groups, and a web forum for people with PMR and GCA.

  Versus Arthritis (www.versusarthritis.org), formed in 2018 following a merger of Arthritis Care and Arthritis Research UK, have information booklets on Polymyalgia rheumatica and Giant cell arteritis.

- **Assess and manage osteoporotic fracture risk.**

- **Co-prescribe stomach protection**[1]

# MANAGEMENT[5]

- **Arrange routine reviews one week after any change in dose and at least every 3 months** in the first year following diagnosis.

- **Advise the person to arrange a review at other times:**

  Urgently, if they develop symptoms of GCA.

  Routinely, if they develop symptoms of relapsing PMR, including proximal pain, fatigue, and morning stiffness.

# MANAGEMENT-Corticosteroid therapy[1]

***Suggested initial oral steroid and tapering regimen*:**

- Prednisolone 15 mg daily for three weeks
- then 12.5 mg daily for three weeks
- then 10 mg daily for four to six weeks
- then reduction by 1 mg every four to eight weeks OR alternate day reductions (eg 7.5 mg/10 mg alternate days).

# MANAGEMENT-Corticosteroid therapy[1]

However, the approach to treatment must be flexible and tailored to the individual, with regards to disease activity, steroid toxicity and patient wishes.

***Duration of steroid therapy:***

- this is usually one to three years
- some may require small doses beyond this
- steroids may be stopped if continuing lack of inflammatory symptoms
- raised ESR/CRP without clinical symptoms is not an indication to continue corticosteroids.

# MONITORING AND FOLLOW UP[1]

Early follow up is necessary as part of the diagnostic process, and should occur at one to three weeks post steroid commencement.

The suggested ***follow-up schedule*** is:

- weeks 0, 1–3, 6
- months 3, 6, 9, 12 in first year (with extra visits for relapses or adverse events).

# MONITORING AND FOLLOW UP[1]

The following should be assessed at each monitoring visit:

***Clinical:***

- disease activity and response to treatment
- complications of disease, including GCA and large vessel vasculitis
- steroid-related side effects – assess BP
- atypical features or alternative pathology
- persistent pain may arise from degenerative or soft tissue pathology (eg rotator cuff tears)

# MONITORING AND FOLLOW UP[1]

***Laboratory:***

- full blood count
- ESR/plasma viscosity or CRP (based on RCP guideline- NICE CKS guidelines however do not recommend routinely monitoring inflammatory markers once a diagnosis of PMR has been made as relapse is defined as recurrence of symptoms and isolated raised inflammatory markers are not a definite indicator of relapse)
- urea and electrolytes
- glucose or urine dipstick.

# RELAPSE[1]

Relapse is the recurrence of PMR symptoms or the onset of GCA, not just unexplained raised ESR/CRP.

***Treatment of relapse:***

- Clinical features of GCA – treat as GCA (usually 40–60 mg prednisolone daily).
- Clinical features of PMR – increase prednisolone to previous higher dose. For the management of relapsing symptoms of PMR, the EULAR/ACR guideline recommends that oral prednisolone should be increased to the pre-relapse dose and then decreased gradually (within 4–8 weeks) to the dose at which the relapse occurred[5]

# RELAPSE[1]

- Consider adjuvant immunosuppressive therapy after two relapses. (would need referral to specialist for this)

# SPECIALIST REFERRAL CRITERIA[1]

The following patients should be referred for specialist evaluation.

**Atypical clinical presentations:**

- younger patient < 60 years
- chronic onset
- lack of shoulder involvement
- lack of inflammatory stiffness
- 'red flag' features: prominent systemic features, weight loss, night pain, neurological signs

# SPECIALIST REFERRAL CRITERIA[1]

- peripheral arthritis or other features of autoimmune or muscle disease
- normal or very high inflammatory markers (ie erythrocyte sedimentation rate/C-reactive protein (ESR/CRP) 100)

# SPECIALIST REFERRAL CRITERIA[1]

**Treatment dilemmas:**

- incomplete or non-response to corticosteroids
- ill-sustained response
- unable to reduce corticosteroid dose
- need for prolonged corticosteroid therapy (>2 years)
- contra-indications to corticosteroid therapy

# SUMMARY

- Making a diagnosis of PMR can be challenging but using a stepwise approach as recommended by guidelines can make it easier
- It is important that patients are given information and support at first diagnosis given the long term treatment that they will be receiving with steroids
- Referral criteria are clear and GPs should regularly review whether their patients require referral to a specialist.
- The steroid regimes to manage PMR have historically been varied, often resulting in weaning down steroid doses too soon, causing relapse. The guidelines aim to provide GPs with a consistent approach they can follow and improving knowledge of them is of utmost importance.

# RESOURCES


1. https://www.rcplondon.ac.uk/guidelines-policy/diagnosis-and-management-polymyalgia-rheumatica (June 2010)
2. https://academic.oup.com/rheumatology/article/49/1/186/1789113
3. https://www.gpnotebook.co.uk/simplepage.cfm?ID=-1576665046
4. https://www.rheumatology.org/Portals/0/Files/2015%20PMR%20guidelines.pdf
5. https://cks.nice.org.uk/polymyalgia-rheumatica#!background (Updated January 2019)