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GUIDELINES

BSR and BHPR guidelines for the management of polymyalgia rheumatica

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Executive summary

Scope and purpose

PMR is the most common inflammatory rheumatic disease in the elderly and is one of the biggest indications for long-term steroid therapy. There are difficulties in diagnosis, with heterogeneity in presentation, response to steroids and disease course.

The aim of these guidelines is a safe and specific diagnostic process for PMR, using continued assessment, and discouragement of hasty initial treatment. Their scope is to provide advice for the diagnosis of PMR, management and monitoring of disease activity, complications and relapse. The management of GCA is not covered and is published separately.

The full guideline is available at *Rheumatology* online.

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Guidelines

(1) We recommend that a safe, stepped diagnostic process be adopted for the evaluation of PMR (Strength of recommendation C).

Diagnosis of PMR should start with evaluation of core inclusion and exclusion criteria, followed by assessment of the response to a standardized dose of steroid [1]. Atypical features or response to steroid should prompt consideration of alternative pathology, and specialist referral.

Unlike with GCA, urgent institution of steroid therapy is not necessary and can be delayed to allow full assessment. However, if the patient does present with symptoms suspicious of GCA, then urgent institution of high-dose steroid therapy is needed (see Guidelines for Management of GCA).

(i) Core inclusion criteria:

- Age >50 years, duration >2 weeks
- Bilateral shoulder or pelvic girdle aching, or both
- Morning stiffness duration of >45 min
- Evidence of an acute-phase response

PMR can be diagnosed with normal inflammatory markers, if there is a classic clinical picture and response to steroids. These patients should be referred for specialist assessment.

(ii) Core exclusion criteria:

- Active infection
- Active cancer
- Active GCA (see part iii)

The presence of the following conditions decreases the probability of PMR, and they should also be excluded:

- Other inflammatory rheumatic diseases
- Drug-induced myalgia
- Chronic pain syndromes
- Endocrine disease
- Neurological conditions, e.g. Parkinsons disease

(iii) Patients should be assessed for evidence of GCA, as this requires urgent institution of high-dose steroid (see separate guidelines)

- Abrupt-onset headache (usually temporal) and temporal tenderness
- Visual disturbance, including diplopia
- Jaw or tongue claudication
- Prominence, beading or diminished pulse on examination of the temporal artery
- Upper cranial nerve palsies
- Limb claudication or other evidence of large-vessel involvement

(iv) Patients should be assessed for response to an initial standardized dose of prednisolone 15 mg daily orally [1, 2].

A patient-reported global improvement of $\geqslant$70% within a week of commencing steroids is consistent with PMR, with normalization of inflammatory markers in 4 weeks. A lesser response should prompt the search for an alternative condition.

(v) The diagnosis of PMR should be confirmed on further follow-up [2]. Follow-up visits should include vigilance for mimicking conditions.

(2) We recommend documentation in the patient's medical record of a minimum data set, which forms the basis for the diagnosis.

- The core clinical inclusion and any exclusion criteria
- Laboratory investigations before commencement of steroid therapy
  - Full blood count
  - ESR/plasma viscosity and/or CRP
  - Urea and electrolytes
  - Liver function tests
  - Bone profile
  - Protein electrophoresis (also consider urinary Bence Jones Protein)
  - Thyroid stimulating hormone
  - Creatine kinase
  - RF (ANA and anti-CCP antibodies may be considered)
  - Dipstick urinalysis
  - Chest X-ray may be required

(3) We recommend the following approach for the evaluation of proximal pain and stiffness [3] (Fig. 1).

(4) We recommend early specialist referral in the following circumstances (C).

Atypical features or features that increase likelihood of a non-PMR diagnosis:

- Age <60 years
- Chronic onset (>2 months)
- Lack of shoulder involvement
- Lack of inflammatory stiffness
- Prominent systemic features, weight loss, night pain, neurological signs
- Features of other rheumatic disease
- Normal or extremely high acute-phase response

Treatment dilemmas such as:

- Incomplete, poorly sustained or non-response to corticosteroids
- Inability to reduce corticosteroids
- Contraindications to corticosteroid therapy
- The need for prolonged corticosteroid therapy (>2 years)

However, patients with a typical clinical picture and complete sustained response to treatment, and no adverse events can be managed in primary care.

(5) We recommend initiation of low-dose steroid therapy with gradually tailored tapering in straightforward PMR (B).

In the absence of GCA, urgent steroid therapy is not indicated before the clinical evaluation is complete.

The suggested regimen is:

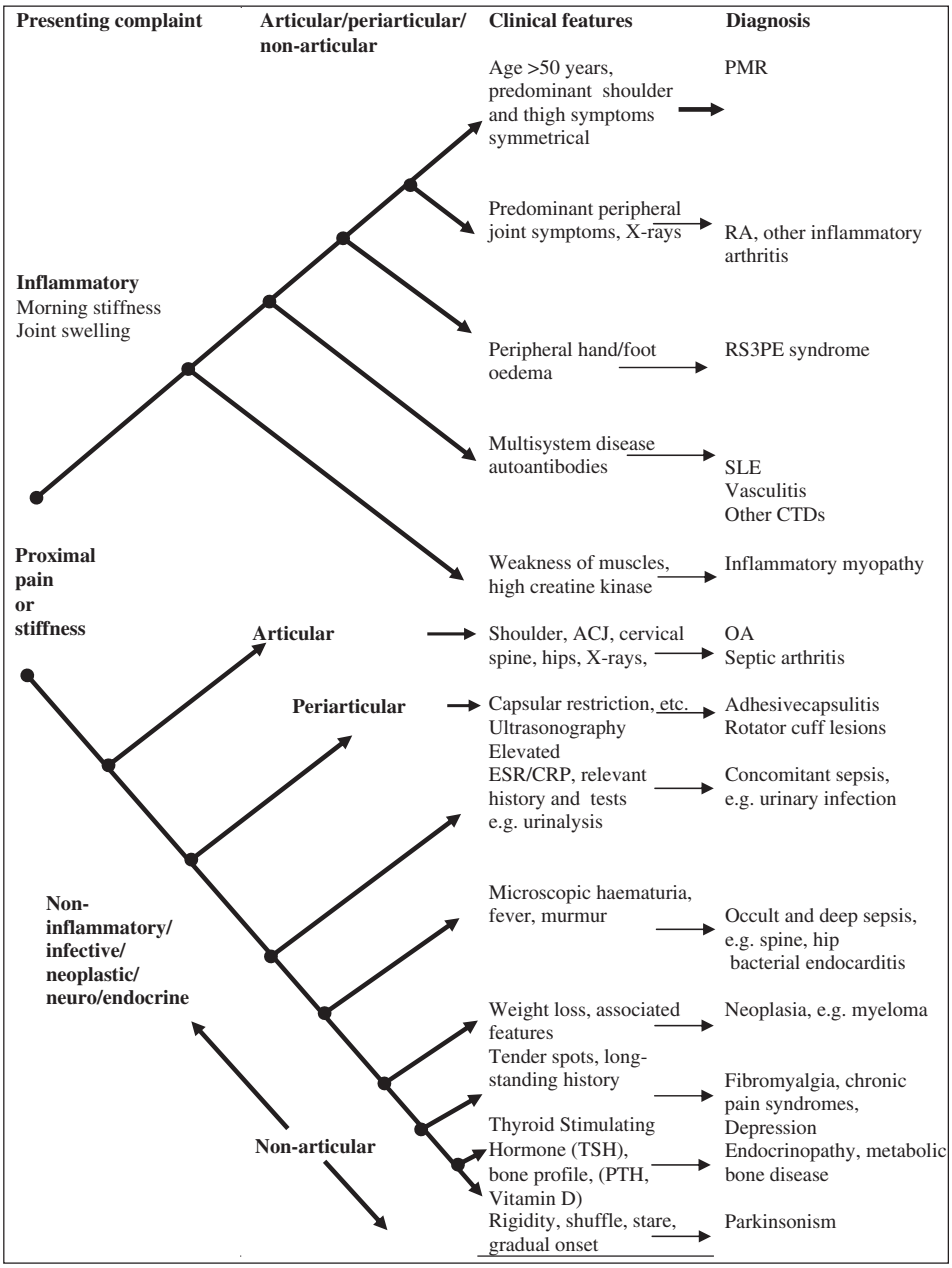
- Daily prednisolone 15 mg for 3 weeks
- Then 12.5 mg for 3 weeks
- Then 10 mg for 4–6 weeks
- Then reduction by 1 mg every 4–8 weeks or alternate day reductions (e.g. 10/7.5 mg alternate days, etc.)

However, there is no consistent evidence for an ideal steroid regimen suitable for all patients. Therefore, the approach to treatment must be flexible and tailored to the individual as there is heterogeneity in disease course. Some benefit from a more gradual steroid taper. Dose adjustment may be required for disease severity, comorbidity, side effects and patient wishes.

Intramuscular methylprednisolone (i.m. depomedrone) may be used in milder cases and may reduce the risk of steroid-related complications. Initial dose is 120 mg every 3–4 weeks, reducing by 20 mg every 2–3 months [4].

Usually 1–2 years of treatment is needed [5]. The need for ongoing therapy after 2 years of treatment should

Fig. 1 Approach to the evaluation of proximal pain and stiffness. ACJ: acromio-clavicular joint.

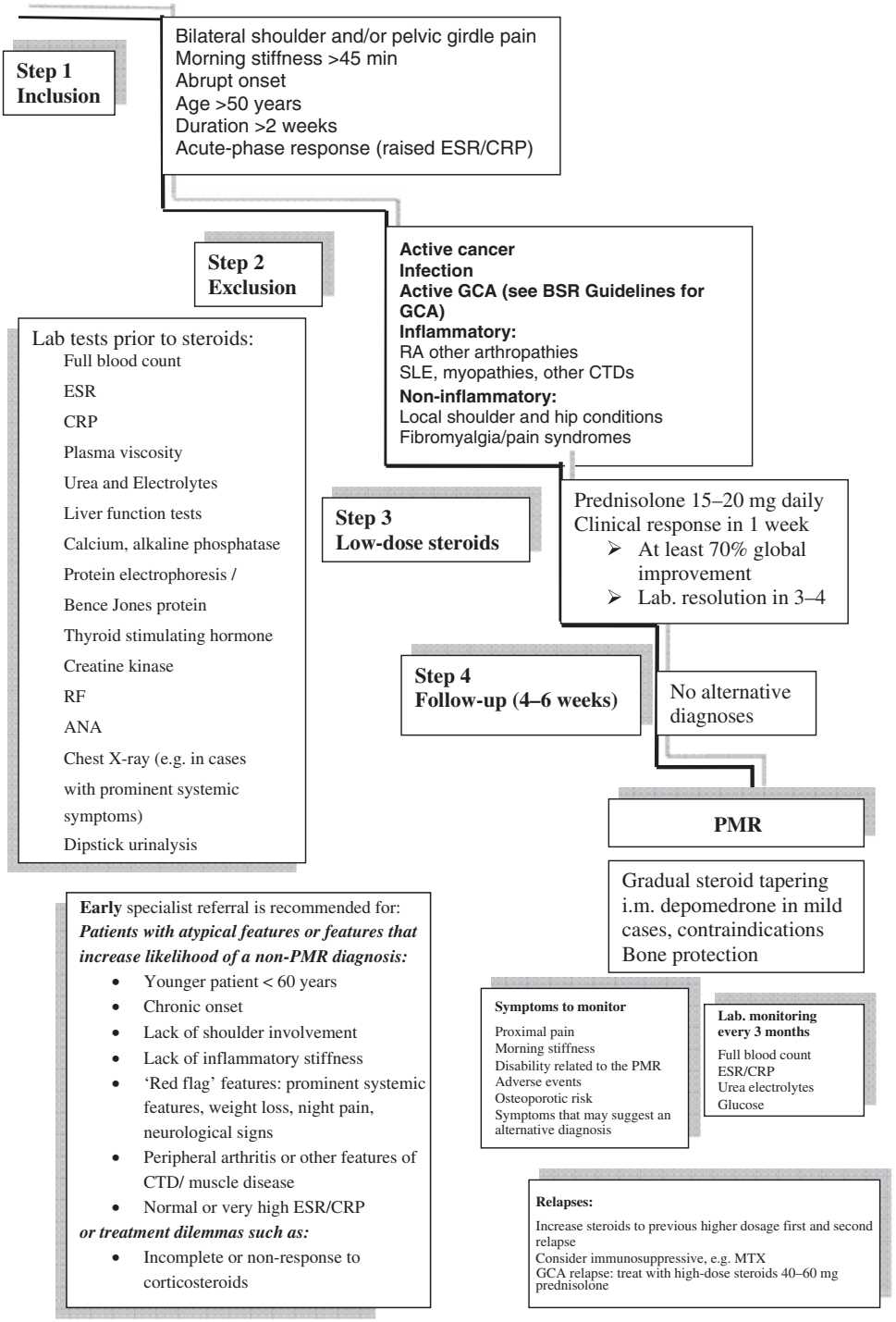


prompt the consideration of an alternative diagnosis, and referral for specialist evaluation.

(6) We recommend the use of bone protection when initiating steroids for PMR to prevent the complications of osteoporosis (A−).

- Individuals with high fracture risk, e.g. aged ⩾65 years or prior fragility fracture
  - Bisphosphonate with calcium and vitamin D supplementation
  - DEXA not required
- Other individuals
  - Calcium and vitamin D supplementation when starting steroid therapy.
  - DEXA scan recommended
  - A bone-sparing agent may be indicated if *T*-score is −1.5 or lower.
- Individuals requiring higher initial steroid dose
  - Bisphosphonate with calcium and vitamin D supplementation (because higher cumulative steroid dose is likely)

FIG. 2 Approach to diagnosis and management of polymyalgia.



(7) We recommend vigilant monitoring of patients for response to treatment and disease activity (B).

Follow-up schedule:

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

Early follow-up is necessary as part of the diagnosis to evaluate response to initial therapy [2], and the first follow-up should occur at 1–3 weeks before commencement of steroids.

Clinical assessment:

At each visit, patients should be assessed for the following:

- Response to treatment: proximal pain, fatigue and morning stiffness

It is important to distinguish between symptoms due to inflammation and those due to co-existing degenerative problems.
- Complications of disease including symptoms of GCA, e.g. headaches, jaw claudication and large-vessel disease
- Steroid-related adverse events
- Atypical features or those suggesting an alternative diagnosis

Laboratory monitoring:

- Full blood count, ESR/CRP, urea and electrolytes, glucose

Duration of treatment and follow-up:

- Usually 1–3 years of treatment, although some will require small doses of steroids beyond this. Flexibility in approach is necessary given the heterogeneous nature of disease. Steroids may be stopped when the patient is asymptomatic from their inflammatory symptoms.
- Isolated raised ESR or CRP is not an indication for continuing steroid therapy but may require investigation and referral.
- Persistent pain may arise from co-existing OA and rotator cuff tears.

(8) We recommend the following approach to relapse of disease.

Relapse is the recurrence of symptoms of PMR or onset of GCA, and not just unexplained raised ESR or CRP [6].

Treatment of relapse:

- Clinical features of GCA: treat as GCA (usually oral prednisolone 40–60 mg daily) (see GCA guideline)
- Clinical features of PMR: increase prednisolone to previous higher dose.
- Single i.m. injection of methylprednisolone (depomedrone) 120 mg can also be used.
- Further relapses: consider introducing DMARD therapy after two relapses

The approach to diagnosis and management of PMR is summarized in Fig. 2.

Patient education

An ARC patient information booklet is available. Further support is available from local patient groups under the auspices of PMRGCA-UK.

Recommendations for audit

Audit standards should include the minimum data set recorded prior to steroid therapy, initial steroid dose and taper, monitoring schedule, use of bone protection and provision of patient information.

Outcomes measures include disease relapse, persistent disease activity, cumulative steroid dosage, adverse events and complications of therapy and quality of life.

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