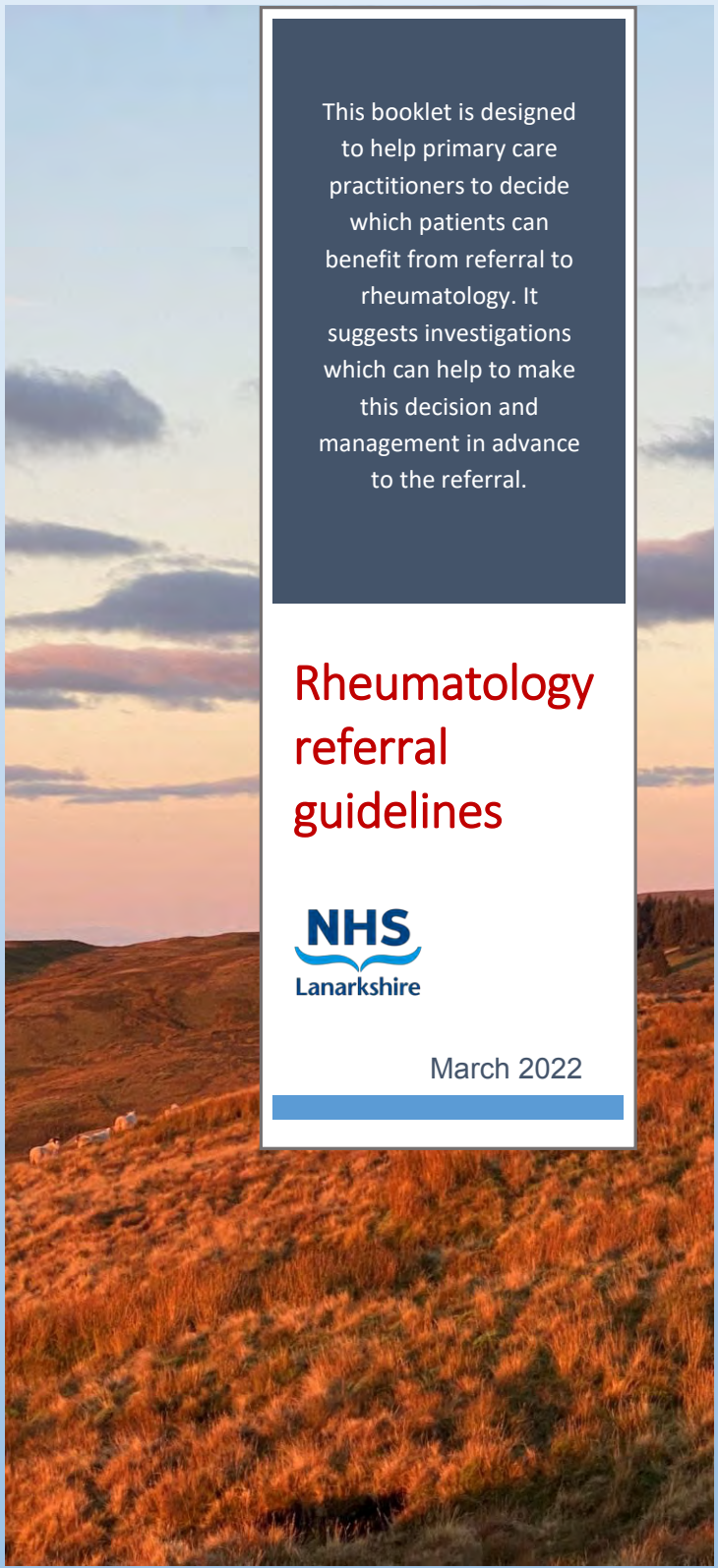

This booklet is designed to help primary care practitioners to decide which patients can benefit from referral to rheumatology. It suggests investigations which can help to make this decision and management in advance to the referral.

Rheumatology referral guidelines

NHS Lanarkshire

March 2022

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AXIAL SPONDYLOARTHRITIS REFERRAL GUIDELINE

Axial SpA (axSpA) is a chronic inflammatory condition predominantly involving the spine and sacroiliac joints (SIJ), with or without extra-spinal manifestations including peripheral arthritis, enthesitis, iritis, psoriasis and inflammatory bowel disease (IBD).

HISTORY

A key feature of axial SpA is low back pain in patients below 45 years of age, waking at night, improving with movement and responding to non-steroidal anti-inflammatory drugs usually within 48 hours.

Risk factors include psoriasis, IBD, recent genitourinary infection and a family history of spondyloarthritis or psoriasis.

EXAMINATION

- Reduced range of spinal movements, hip and chest expansion.
- Check for: peripheral arthritis, enthesitis, dactylitis, psoriasis (including psoriatic nail symptoms), and uveitis.

HELPFUL INVESTIGATIONS

- ESR and CRP – usually elevated
- HLA-B27 – may be normal

PRIMARY CARE MANAGEMENT

- Treat symptoms with full dose NSAID and/or analgesics pending clinic review.
- Early involvement of the allied health care team (physiotherapy, OT, orthotics, podiatry, etc.) should be considered.

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WHO TO REFER

Refer patients with low back pain that started before the age of 45 years and has lasted for longer than 3 months, and if 4 or more of the following additional criteria are also present:

- low back pain that started before the age of 35 years (this further increases likelihood of axial spondyloarthritis)
- waking during second half of the night because of symptoms
- buttock pain
- improvement with movement
- improvement with taking non-steroidal anti-inflammatory drugs (often within 48 hours)
- a first-degree relative with spondyloarthritis
- current or past arthritis
- current or past enthesitis
- current or past psoriasis

If exactly 3 of the additional criteria are present, perform an HLA-B27 test. If the test is positive, refer the person to a rheumatologist for a spondyloarthritis assessment.

When screening criteria are not met but clinical suspicion remains, advice the person to seek repeat assessment if new signs, symptoms or risk factors develop. This may be particularly appropriate if the person has or had psoriasis, inflammatory bowel disease or uveitis.

WHO NOT TO REFER

- If there is no evidence of inflammatory back pain in the history (as described above). Consider NHSL Lumbar/Spinal Pathway for other cases.

For more information:

NICE guideline on Spondyloarthritis for 16s and over (2017)

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CONNECTIVE TISSUE DISEASE REFERRAL GUIDELINE

Connective tissue diseases (CTD) cover a wide range of autoimmune diseases. They are often associated with particular autoantibodies. The 'classic' CTDs include systemic lupus erythematosus (SLE), rheumatoid arthritis (RA), systemic sclerosis (or scleroderma), polymyositis and dermatomyositis.
Patients may present with a variety of symptoms and these can vary greatly between individuals.

HISTORY

- Photosensitive skin rash
- Polyarthralgia or polyarthritis
- Muscle pain and weakness
- Mouth ulcers
- Dry eyes and/or dry mouth
- Raynaud's phenomenon
- Hair loss
- Pleurisy
- Dyspnoea
- Recurrent miscarriages

EXAMINATION

- Skin rash
- Synovitis
- Mouth ulcers
- Sclerodactyly / scleroderma
- Scarring alopecia
- Raynaud's phenomenon / Digital ulceration
- Telangiectasia
- Pleural or pericardial effusion

HELPFUL INVESTIGATIONS

- FBC with anaemia, leucopaenia or thrombocytopaenia
- Raised ESR
- Strongly positive ANA and positive anti-dsDNA

WHEN TO CHECK ANA

ANA testing has a high degree of sensitivity (>95%) for SLE and other connective tissue diseases; however the utility of the test is limited by very low specificity (<60%) which severely limits its value in the investigation of patients who have only non-specific symptoms. Consequently, the ANA test is not a good screening test for SLE or other autoimmune diseases.
The clinical value of an ANA test is tremendously enhanced by testing when there is a reasonable pre-test probability (i.e. clinical suspicion) of a connective tissue disease.

One reason that the specificity of the ANA test is low is that ANA can also be found in non-rheumatic inflammatory diseases such as autoimmune hepatitis, primary biliary cirrhosis, Crohn's disease, chronic infectious diseases (TB, SBE, infectious mononucleosis) and lymphoproliferative disorders.

ANA can also be induced by many drugs.

Drugs associated with ANA production and lupus-like disease

Procainamide	D-penicillamine	Terbinafine	Hydralazine
Isoniazid	Minocycline	Quinidine	Methyldopa
Phenytoin	Chlorpromazine	Anti-TNF agents	

Clinically significant titres of ANA are usually >1:160
ANA 1:40 occur in 20 – 30% of normal healthy individuals
ANA 1:80 occur in 10 – 15% of normal healthy individuals
ANA 1:160 occur in 5% of normal healthy individuals
ANA 1:320 occur in 3% of normal healthy individuals
In the elderly, over 70 years, up to 70% have a positive ANA of 1:40

Anti-dsDNA antibody testing is performed as a reflex by the laboratory where the ANA screening test is found to be significantly positive. Anti-dsDNA antibodies are principally associated with lupus.

PRIMARY CARE MANAGEMENT

Manage symptoms pending clinic review

WHO TO REFER

- Patients with several CTD symptoms or signs (typically 4 or more) who test ANA positive **AND** anti-dsDNA positive.

'False positive' ANA results up to 1:80/1:160 (and sometimes beyond) are relatively non-specific and, in themselves, not highly indicative of a connective tissue disorder.

Anti-dsDNA levels up to 75 IU/ml are considered as negative and not suggestive of lupus, particularly where relevant clinical indicators are absent. A second test (crithidia), which also detects dsDNA autoantibodies, is routinely undertaken as a safety net by the laboratory on samples where the quantitative anti-dsDNA result is >30 IU/ml to identify borderline positive samples.

WHO NOT TO REFER

- Patients with arthralgia who have a positive ANA and negative anti-dsDNA, with no other symptoms and signs.
- Patients with Raynaud's who have a positive ANA and negative anti-dsDNA, with no other symptoms or signs.
- Patients with generalized pain who have a positive ANA and negative anti-dsDNA, with no other symptoms or signs.

FIBROMYALGIA REFERRAL GUIDELINE

Fibromyalgia (FMS) is a syndrome of chronic widespread pain and tenderness associated with fatigue, unrefreshing sleep and perception of cognitive dysfunction. There is no organic muscle or joint pathology.

Diagnosis of the condition should be primarily conducted in primary care however a secondary care referral should occur when there is diagnostic uncertainty.

HISTORY

- Chronic widespread pain (3 or more months of generalised pain, affecting both sides of the body and above and below the waist, with no other explanation)
- Widespread tenderness
- Fatigue
- Non-refreshed sleep
- Mood disturbance
- Cognitive impairment

EXAMINATION

Widespread tenderness.

Previous diagnostic criteria for fibromyalgia have included specific tender points on examination but these have proved hard to standardise and are not required for diagnosis.

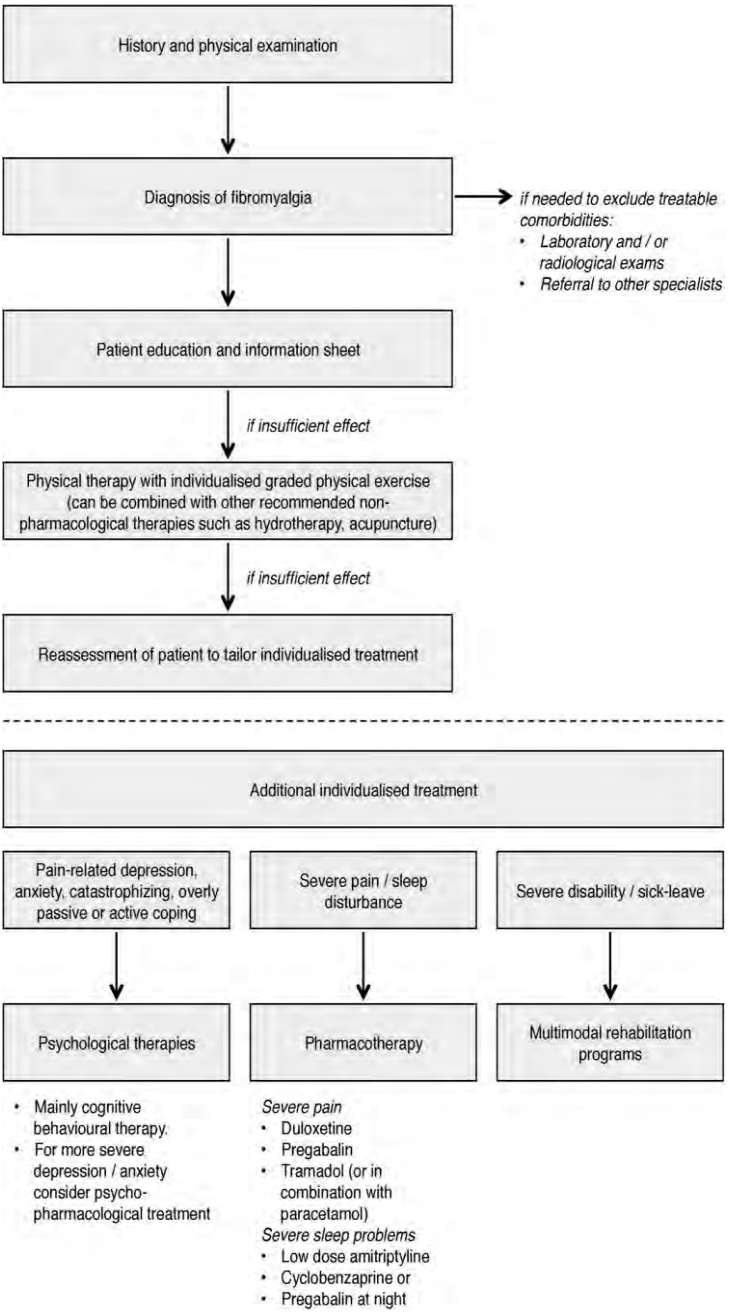
HELPFUL INVESTIGATIONS

Fibromyalgia is a clinical diagnosis. There are no specific diagnostic tests. Simple blood tests are normal although FMS can coexist with other diseases.

- U&E, LFT, TFT, bone profile
- ESR and CRP

PRIMARY CARE MANAGEMENT

- Information such as that provided by Versus Arthritis https://www.versusarthritis.org/about-arthritis/conditions/fibromyalgia/ and Michigan University https://www.uofmhealth.org/health-library/hw196365#hw196368
- Regular aerobic exercise (20 minutes 2-3 times per week)
- A pain management approach including elements of education, exercise and psychological support such as CBT (see the diagram below) https://www.uofmhealth.org/health-library/hw196365#hw196365-HealthTools
- Many reviews have indicated that antidepressants are useful in the treatment of fibromyalgia; the best evidence for improvement in pain and in quality of life is for duloxetine and for pregabalin (neither is licensed for the treatment of fibromyalgia in the UK).



EULAR revised recommendations for the management of fibromyalgia https://ard.bmj.com/content/76/2/318.full

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WHO TO REFER

- Patients with joint swelling and tenderness
- Patients with joint pain and elevated inflammatory markers (ESR and CRP)
- Patients in whom there is clinical suspicion of underlying connective tissue disease – patients with a photosensitive rash, Raynaud's, recurrent oral ulceration or sicca symptoms and a positive ANA. A weakly positive (1:40 or 1:160) antinuclear antibody (ANA) is a common laboratory finding (seen in up to 15% of healthy people) and unless there are clinical features to suggest a connective tissue disorder, these patients will not require Rheumatology review.

WHO NOT TO REFER

- Patients with generalized pain and/or fatigue with no other symptoms or signs and with normal simple test results.
- Patients who have previously been discharged from Rheumatology with a diagnosis of fibromyalgia/widespread pain are very unlikely to benefit from further review and the process may simply delay referral to the Pain service.

For more information:
Fibromyalgia Syndrome (FMS) Parliamentary Briefing

GCA REFERRAL GUIDELINE

Diagram version and GCA probability score

Giant Cell Arteritis (GCA) or Temporal Arteritis (TA) is a form of Large Vessel Vasculitis (inflammation of the blood vessels) which affects mainly elderly population (usually above 65 years of age). The complications include permanent loss of vision, stroke and other complications related to arterial occlusion.

Treatment with high doses of steroids usually leads to resolution of symptoms within 24 hours (this is not specific effect and does not confirm the diagnosis). In uncertain cases the use of steroids before confirmation of GCA delays diagnosis and poses risk of prolonged, unnecessary treatment.

Patients with suspected GCA should be therefore seen in the specialist clinic as soon as possible, best within 24 - 48 hours from first presentation. Please call the rheumatology on 01698 366088 to make sure that the appointment will be arranged on time.

HISTORY

- Sub-acute/acute onset
- New type of headache
- Scalp tenderness – on brushing/combing hair, resting head on the pillow
- Jaw or tongue claudication – pain occurring while chewing
- Unilateral blurring, double vision, visual loss
- Limb claudication
- Systemic upset - weight loss, night sweats, fever
- Shoulder +/- pelvic girdle muscles pain and stiffness – difficulty turning in bed and rising arms

EXAMINATION

- Temporal artery – tenderness, thickening, beading, reduced or absent pulsation
- Scalp tenderness
- Peripheral pulses upper extremities – diminished or lack
- Auscultation of subclavian, axillary, brachial arteries – bruits
- Blood pressure on both arms – discrepancy of more than 20mmHg in systolic pressure
- Visual acuity, eye movements, visual fields

HELPFUL INVESTIGATIONS

- ESR and CRP – raised
- Full blood count – low Hb, raised PLT
- U&E, LFT, ALP
- Fundoscopy – pale, swollen optic disc with haemorrhages (anterior ischaemic optic neuritis)

PRIMARY CARE MANAGEMENT

- If high risk of GCA (or GCA probability score above 12) – prednisolone 40mg daily plus omeprazole
- If tongue claudication and/or new visual symptoms (partial/complete visual loss, amaurosis fugax, diplopia) – prednisolone 60mg daily plus omeprazole
- Refer to rheumatology urgently – call 01698 366088
- Refer to ophthalmology urgently (all with new visual symptoms) – call 01355 585387

For more information:
British Society for Rheumatology guideline on diagnosis and treatment of giant cell arteritis, Jan 2020

WHO TO REFER

- Patients of 50 years of age and older
- Acute/sub-acute onset of headache and scalp tenderness
- Other symptoms of GCA (as above)
- Raised ESR/CRP (if typical presentation do not delay the referral by waiting for test results)
- or GCA probability score ≥ 10

WHO NOT TO REFER*

- Patients under 50 year of age
- Chronic headache/neck/shoulder pain
- Normal ESR and CRP
- Alternative diagnosis more likely, e.g.
    - Migraine
    - Infection including sinusitis
    - Herpes zoster
    - Temporo-mandibular joint disease
    - Ear disease
    - Trigeminal neuralgia
    - Cervical spine disease
    - TIA
    - Intracranial pathology
    - Malignancy
- Pain on mouth opening but not aggravated by chewing
- Bilateral blurred vision of known cause (glaucoma, cataract, uncontrolled diabetes) and corrected with glasses
- Or GCA probability score < 10
- GCA is rare in non- Caucasians

***Call rheumatology on-call if unsure**

Fast Track Giant Cell Arteritis Referral Pathway

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1 **Age >50**
Most >65
Rare in non-Caucasions

2 **Symptoms of GCA***

3 **Subacute/ acute onset**

4 **Raised CRP and/or ESR**

****All 1-4 present? Refer to GCA Fast Track Pathway**

New **visual symptoms**, especially partial/complete vision loss, amaurosis fugax, diplopia

60mg Prednisolone + PPI
Urgent referral phone
Ophthalmology 01355 585387

Everyone else

40mg Prednisolone + PPI
(60mg if jaw claudication or possible visual symptoms)

email:
GCAfastrak@lanarkshire.scot.nhs.uk
or call **Rheumatology 01698 366088**

In completing SCI referral state clearly for 'GCA Fast Track Pathway'

Include patient's current phone no.

***Key symptoms of GCA:**

- New type of headache (head pain)
- Scalp tenderness
- Jaw or tongue claudication
- Visual (amaurosis fugax, double vision)
- Limb claudication
- Abnormal temporal artery

Often in conjunction with:

- PMR
- Systemic upset

**If 1-4 not present, patients will be triaged outside the fast track pathway. Use probability score as a guide (on reverse); If you need advice contact on-call Rheumatologist via switch board.

Aims of the GCA Fast Track Service

- Provide rapid access to specialist clinical assessment, temporal artery ultrasound, biopsy and other imaging for those with possible GCA
- Provide a secure diagnosis in as many patients as possible
- Reduce rate of sight loss and stroke in GCA
- Minimise the impact of Prednisolone in those who don't have GCA

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GCA probability score

Weightage	-3	0	1	2	3	
Age		≤49	50-60	61-65	>65	
Sex			M	F		
Onset		>24 weeks	12-24 weeks	6-11 weeks	<6 weeks	
CRP		0-5	6-10	11-24	≥25	

Symptoms	-3	0	1	2	3	
Cranial: Head and scalp pain		N	Y			
Constitutional (weight loss, night sweats, pyrexia)		N	Single		Combination	
Polymyalgia		N		Y		
Ischaemic (uniocular blurring, diplopia, amaurosis, jaw/tongue pain)		N			Y	

Signs	-3	0	1	2	3	
Visual (AION, CRAO, Field loss, RAPD)		N			Y	
Temporal artery abnormality		N	tenderness	thickening	loss of pulse	
Extra-cranial artery abnormality		N	thickening	bruit	loss of pulse	
Cranial N palsy (3.4.6)		N			Y	

Alternative diagnosis	-3	0	1	2	3	Score
Active infection	Y	N				
Active cancer	Y	N				
Systemic rheumatic diseases	Y	N				
Head and neck pathology	Y	N				
Other	Y	N				

Total score	

Total score ≥10 patients at risk of GCA

Total score <10 likely not GCA, do not start prednisolone

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GOUT REFERRAL GUIDELINE

Most patients with gout are managed in primary care. Where there is suspicion of septic arthritis urgent referral is indicated. Consider routine referral if there is diagnostic uncertainty or genuine treatment resistance (noting that compliance with therapy is notoriously poor).

HISTORY

- Severe pain usually in a single joint
- Acute onset (typically less than 24 hours, often overnight) with episodes lasting 1 to 2 weeks
- Frequently involves 1st MTPJ, foot or ankle, but may involve any joint especially if coexistent osteoarthritis

EXAMINATION

- Tender, hot, red, swollen joint(s)
- Tophi appear as chalky white deposits and are markers of severe disease. Typically form in digits or over elbows

HELPFUL INVESTIGATIONS

- CRP – often elevated,
- FBC, U&E, LFTs
- Urate –baseline often normal, 4-6 weeks post attack often elevated
- X-ray of symptomatic joints (characteristic erosions in established disease)
- Good practice recommends screening patients for cardiovascular risk factors
- Synovial aspirate usually not performed in primary care

ACUTE GOUT

- Exclude septic arthritis
- Continue allopurinol if on this already
- Treat as early as possible
- NSAIDs at maximum doses are the treatment of choice if no contra-indications
- Colchicine: 0.5mg bd - qds is an alternative to NSAIDs (Can be used safely if eGFR <30ml/min at reduced doses)
- Corticosteroids (intra-articular or oral Prednisolone 20mg tapered over 1-2 weeks) are an alternative in those where NSAIDs & colchicine are not tolerated or are contraindicated
- Adjunctive non-pharmacological treatment (eg topical, ice, rest)
- Review at 4-6 weeks
    - Assess lifestyle factors (diet, exercise, alcohol, sugary drinks)
    - Assess cardiovascular risk factors (obesity, hypertension, lipids, diabetes)
    - Review prescribed medication (diuretics)
    - Perform SUA, renal function

ALLOPURINOL

Indications

- Two or more attacks of uncomplicated gout within one year
- One attack of gout with gouty tophi; or renal insufficiency; or uric acid stones; or need to continue treatment with diuretics

Dose titration

- Start 1-2 weeks after acute attack settled

- Increase dose every 4 weeks aiming for target uric acid <300micromol/l
- Start at a dose of 50-100mg titrating the dose by 50-100mg every 4 weeks to a target SUA of < 300 micromol/l and maximum dose of 900mg.
- Lower doses are usually required to achieve target SUA in those with renal impairment.

ACUTE GOUT PROPHYLAXIS

- Flares of gout may continue for 12 months after urate control, and are paradoxically more common with aggressive urate lowering.
- To reduce flares prophylaxis should be offered with colchicine 500mcg b.d. for up to 6 months. Consider low dose NSAID in patients intolerant of colchicine

ALTERNATIVES TO ALLOPURINOL

- If allopurinol contraindicated or intolerance use Febuxostat 80 -120mg

WHO TO REFER

- Tophaceous gout, when progressive despite adequate treatment
- Refractory gout – after 3 attacks whilst on adequate treatment
- If relative contraindication to febuxostat and allopurinol contraindicated or ineffective
- Those where the diagnosis is unclear with persistent symptoms

WHO NOT TO REFER

- Those who have a clear diagnosis of gout who are responding to treatment
- Tophaceous gout that is improving with treatment
- Check compliance with medication and patient understanding of treatment rationale in those who do not appear to be responding to treatment before referring

For more information: The British Society for Rheumatology Guideline for the Management of Gout

INFLAMMATORY ARTHRITIS REFERRAL GUIDELINE

The inflammatory arthritides are a heterogeneous group of autoimmune conditions that are characterised by inflammation within the synovial lining of joints or tendons. Each of the inflammatory arthritidies has a characteristic pattern of joint involvement, associated features and risk of complications.

A diagnosis of an inflammatory arthritis can only be made once inflammation within the affected joint (synovitis or joint effusion), tendon (tenosynovitis) or digit (dactylitis) has been confirmed by examination or, in some cases, imaging.

Prompt, early diagnosis of inflammatory arthritis and initiation of disease modifying anti-rheumatic (DMARD) therapy is vital to reduce the long term risk of permanent joint damage and disability; however, recognition of the early stages can be difficult.

HISTORY

Symptoms that are suggestive of an inflammatory arthritis include:

- Joint pain – usually localised to the joint; feels worse after inactivity and may improve with repeated movement or activity
- Joint swelling – localised to the joint or tendon, usually persistent
- Stiffness – inactivity gelling and prolonged morning stiffness
- Loss of function
- Constitutional – fatigue, weight loss, fevers / night sweats, loss of appetite, low mood

Associated features – are useful to confirm specific diagnosis once joint inflammation has been confirmed

- Seronegative Arthropathies – inflammatory back pain; preceding infection (especially urinary, GI or GU) and past or first degree relative history of Psoriasis, Inflammatory Bowel Disease or Uveitis
- Connective Tissue Symptoms – mouth ulcers, sicca symptoms, alopecia, rash and photosensitivity, Raynaud's phenomenon, serositis
- Sarcoidosis – dry cough, increased breathlessness, rash, erythema nodosum, painful red eyes

EXAMINATION

Focuses on identification of inflammatory change in, or around, the symptomatic joint; determining the distribution of affected joints and identifying evidence of joint deformity and dysfunction

- Synovitis – soft tissue swelling / thickening; may be tender, warm and erythematous with restricted movement; needs to be differentiate from hard, smooth, nodal swelling of osteoarthritis
- Tenosynovitis – linear or fusiform swelling and tenderness of the affected tendon,
- Dactylitis – inflammation of a whole digit; suggestive of seronegative arthritidies (especially psoriatic arthritis) but can be a feature of gout and sarcoidosis

HELPFUL INVESTIGATIONS

Most inflammatory arthritidies remain a clinical diagnosis where investigations support the clinical diagnosis. Negative investigations should not delay referral in patients with clinically evident joint swelling

- UE, LFT, FBC – assess suitability for DMARD therapy
- CRP, ESR – normal in 10-15%

- Rheumatoid Factor – positive in 65-70% of cases of rheumatoid arthritis
- ANA – if there are features suggestive of a connective tissue disease
- Xrays of Hands / Feet - often normal in the early stages of an inflammatory arthritis.

PRIMARY CARE MANAGEMENT

Patients with clinically suspected inflammatory arthritis should be referred urgently to the rheumatology department, especially if there is clinically evident synovitis or dactylitis.

Prior to rheumatology clinic review, useful measures to manage symptoms include:

- NSAIDs – taken regularly at maximal doses. E.g Ibuprofen (upto 600mg qds), Naproxen (500mg bd), Etodolac (600mg od), Meloxicam (15mg od), Etoricoxib (60mg od)
- Simple analgesics – as tolerated – E.g Paracetamol, Cocodamol 8-30/500, Codydramol 10-30/500

Empirical corticosteroid therapy should be avoided wherever possible unless the patient is experiencing significant levels of pain or functional impairment

WHO TO REFER

Any patient where there is a high degree of clinical suspicion for an inflammatory arthritis, especially if there is clinically evident synovitis or dactylitis

WHO NOT TO REFER

- Patients with elevated Rheumatoid Factor and/or ANA titres but no suggestive symptoms / signs of inflammatory arthritis or connective tissue disease
- Patients in whom an alternative cause for symptoms seems more likely, e.g.
    - Osteoarthritis
    - Fibromyalgia (pain, myalgia and fatigue but low risk features for inflammatory arthritis - no evidence of synovitis or dactylitis, or deformity, normal autoantibodies and normal inflammatory marker levels)

MYOSITIS REFERRAL GUIDELINE

Idiopathic inflammatory myopathies (myositis) are a varied group of rare autoimmune diseases, mainly characterised by inflammation of the skeletal muscles, but involvement of skin and internal organs such as lungs, heart, and oesophagus is common.

Patients presenting with acute/subacute proximal muscle weakness and more than 10 fold increase in creatinine kinase (not explained by alternative, more likely cause – see below) should be referred to rheumatology urgently.

HISTORY & EXAMINATION

- Proximal muscle weakness/tetraparesis which can develop acutely (several days) or subacutely (several weeks up to a few months) and can be associated with at least one of the following:
    - Typical skin lesions such as:
        - Gottron papules on the dorsal aspect of the hands and fingers,
        - periorbital oedema and erythema of the face (heliotrope rash),
        - Rash on the anterior upper chest (V-sign) or the posterior neck (shawl sign).
        - Periungal erythema and telangiectasia
        - thickened and cracked skin of the ventral and dorsal aspects of the fingers and hands (“mechanic’s hands”)
        - ! in an amyopathic dermatomyositis there are only skin manifestations but there is no weakness of the muscles and no increase of the CK
    - Dysphagia

- Interstitial lung disease
- Raynaud's syndrome
- Sicca syndrome
- Arthritis
- In overlap syndromes, can be associated with any manifestation of other connective tissue disorders (Systemic sclerosis, SLE, Sjögren syndrome)
- Malignancy (lung, breast, ovary, lymphoma)

HELPFUL INVESTIGATIONS

- CK 10–50 fold increase and elevated liver enzymes

! consider CK normal values differences (race, gender)

! CK can be elevated post excessive exercise (repeat after one week of rest)

- Raised CRP +/- ESR
- FBC, U&E, LFTs
- TFT
- ANA

PRIMARY CARE MANAGEMENT

Manage symptoms pending rheumatology clinic review

WHO TO REFER

All patients with acute or subacute onset of symmetrical proximal muscle weakness with elevated CK 10–50 fold and raised CRP/ESR require urgent referral.

WHO NOT TO REFER

- Patients with diffuse myalgia and normal blood tests including CK and inflammatory markers
- Patients with chronic pain syndromes with no new signs or symptoms and normal blood tests
- Patients with alternative diagnosis/cause of elevated CK more likely, e.g.
    - viral infections
    - endocrine disorders
    - liver, cardiac and renal diseases
    - metabolic diseases
    - coeliac disease
    - drug induced myopathy
    - pregnancy
    - surgery
    - trauma or recent physical exercise
    - motor neuron disease
    - myasthenia gravis
    - muscular dystrophy

PMR REFERRAL GUIDELINE

Patients with typical presentation, who have a complete, sustained response to low-dose corticosteroids, and who have no adverse events can be managed in primary care.

HISTORY

- 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 (e.g. raised ESR, CRP)

EXAMINATION

- Shoulder, neck and hip range of movements - reduced
- Muscle strength - normal
- Patients should be assessed for evidence of GCA
    - Temporal artery – tenderness, thickening, beading, reduced or absent pulsation
    - Scalp tenderness
    - Peripheral pulses upper extremities – diminished or lack
    - Auscultation of subclavian, axillary, brachial arteries – bruits
    - Blood pressure on both arms – discrepancy of more than 20mmHg in systolic pressure
    - If visual symptoms - vision acuity, eye movements, visual fields

HELPFUL INVESTIGATIONS

- FBC, U&E, LFT, bone profile, ESR, CRP, plasma viscosity
- Immunoglobulins and electrophoresis (consider Bence Jones protein)
- TFTs
- CK
- Rheumatoid factor (anti-nuclear antibody may also be considered)
- CXR (if prominent systemic symptoms)
- Dipstick urinalysis

PRIMARY CARE MANAGEMENT

- Corticosteroids should be initiated and tapered as follows
    - Daily prednisolone 15mg for 3 weeks
    - Then 12.5mg for 3 weeks
    - Then 10mg for 4–6 weeks
    - Followed by reduction by 1mg every 4–8 weeks
    - Usually 1–2 years of steroid treatment is needed
- Bone protection (weekly bisphosphonate and calcium or vitamin D supplementation)

Assess for corticosteroid-related adverse events: weight gain, diabetes, osteoporosis, hypertension and lipid dysregulation.

For more information: BSR and BHPR guidelines for the management of polymyalgia rheumatica 2009.

WHO TO REFER

- Atypical features
    - Younger patient < 60 years
    - Chronic onset (>2 weeks)
    - Lack of shoulder involvement
    - Lack of inflammatory stiffness
    - Red flag features: prominent systemic features, weight loss, night pain, neurological signs
    - Peripheral arthritis or other features of CTD or muscle disease
    - Normal or very high ESR/CRP
- Treatment dilemmas such as:
    - Incomplete or non-response to corticosteroids
    - Ill-sustained response to corticosteroids
    - Unable to reduce corticosteroids
    - Contraindications to corticosteroid therapy
    - The need for prolonged corticosteroid therapy (>2 years)

WHO NOT TO REFER

- Patients who have no atypical features, who have a complete sustained response to low-dose corticosteroids, and who have no adverse events.
- Patients with alternative diagnosis more likely
    - Active infection and cancer
    - Drug-induced myalgia, e.g. statins
    - Pain syndromes, e.g. fibromyalgia
    - Endocrine, e.g. thyroid
    - Neurological, e.g. Parkinson's disease
    - OA, degenerative and other peri-articular conditions of the shoulder, neck and hips.

VASCULITIS REFERRAL GUIDELINE

The primary systemic vasculitides are a varied group of rare diseases that are characterised by inflammation of blood vessels. They are often difficult to diagnose because the clinical manifestations are varied and can mimic several infectious, neoplastic, and other autoimmune conditions. The type of vasculitis is often related to a patient's age, gender, and ethnic origin.

A definitive diagnosis of systemic vasculitis should be made by a person experienced in diagnosing vasculitis, in the presence of characteristic symptoms and signs of vasculitis, and at least one of the following: histologic evidence of vasculitis; positive serology for ANCA; or specific indirect evidence of vasculitis.

HISTORY & EXAMINATION

The clinical presentations are highly varied and will depend upon the distribution and extent of organ involvement and size of the involved vessels.

- Systemic: Fever, weight loss, fatigue, myalgia, arthralgia
- Cutaneous: purpura, ulceration, gangrene
- Ocular: red eye, visual loss, diplopia
- Mucocutaneous: oral & genital ulceration
- ENT: Hearing loss (conductive, sensorineural); sinus pain; nasal crusting, epistaxis
- Chest: wheeze, cough, haemoptysis, dyspnoea
- Cardiac: pericarditis, cardiomyopathy, ischaemic chest pain
- Abdomen: Ischaemic abdominal pain, bloody diarrhoea
- CNS: Peripheral neuropathy, mononeuritis multiplex, CN palsy, cord lesions, stroke syndromes

- Renal: blood and protein on urine dipstick, deteriorating renal function

HELPFUL INVESTIGATIONS

- ESR, CRP – usually elevated
- FBC, U&E, LFTs
- If high clinical index of suspicion check ANCA (MPO/ PR3) – this can be negative
- Urine dipstick – if positive for blood/protein refer to renal
- Further investigations are guided by system involved – in many cases additional imaging or biopsies of the affected organ(s) is required to confirm the diagnosis of vasculitis

Most vasculitidies do not have an associated blood marker. Elevated ANCA levels are associated with the ANCA-associated, small vessel vasculitidies (e.g. Granulomatosis with Polyangiitis, Eosinophillic Granulomatosis with Polyangiitis, Microscopic Polyangiitis, Pauci-Immune Glomerulonephritis). However, in isolation, without other relevant clinical features, elevated ANCA levels are not diagnostic and can be misleading. Elevated ANCA levels can also be related to inflammatory bowel disease, primary sclerosing cholangitis, rheumatoid arthritis, SLE, autoimmune liver disease and some infectious disease.

PRIMARY CARE MANAGEMENT

If vasculitis is suspected then the patient should be referred urgently to the relevant specialty

WHO TO REFER

- The majority of patients seen with vasculitis are referred by secondary care once other causes have been ruled out
- Consider referring patients with unexplained systemic symptoms and raised ESR/CRP and/or positive ANCA (MPO/PR3) where there is no evidence of underlying infection or malignancy.
- If vasculitis affects one system only consider direct referral to the relevant specialty
    - Isolated cutaneous vasculitis – dermatology
    - Evidence of renal involvement – nephrology
    - Cerebral vasculitis alone – neurology

WHO NOT TO REFER

- Those who have known sepsis or malignancy with weakly positive ANCA (MPO/PR3)
- Diagnosis of vasculitis requires confirmation of vasculitic involvement within the symptomatic or dysfunctioning organ(s). Patients who are non-specifically unwell, who do not have localising symptoms or evidence of end-organ dysfunction are unlikely to have a vasculitis


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