5.3. Connective Tissue Disease and Vasculitis

Background

This group of heterogeneous and rare diseases are often diagnosed late in the course of the disease. They are linked by autoimmunity and organ inflammation, organ dysfunction or organ fibrosis. Referral will be based on clinical suspicion. Emergency referral to A&E or ambulatory care may be indicated in cases where there is significant threat to life, limb, or vital organ. The incidence and diagnostic pointers are given below:

<table>
<thead>
<tr>
<th>Disease</th>
<th>Incidence / 100,000</th>
<th>Female : Male</th>
</tr>
</thead>
<tbody>
<tr>
<td>Raynaud’s Phenomenon (incl secondary causes)</td>
<td>250</td>
<td>1.6 : 1</td>
</tr>
<tr>
<td>Giant Cell Arteritis</td>
<td>13 - 30</td>
<td>1.8 : 1</td>
</tr>
<tr>
<td>Sjogren’s syndrome</td>
<td>3.9 - 5.3</td>
<td>9 : 1</td>
</tr>
<tr>
<td>Systemic Lupus</td>
<td>3.8 - 4.8</td>
<td>6 - 10 : 1</td>
</tr>
<tr>
<td>Scleroderma</td>
<td>0.45 - 1.87</td>
<td>5 - 14 : 1</td>
</tr>
<tr>
<td>Polymyositis</td>
<td>0.2 - 1.0</td>
<td>2.5 : 1</td>
</tr>
<tr>
<td>Polyarteritis nodosa</td>
<td>0.2 - 0.9</td>
<td>1 : 2</td>
</tr>
</tbody>
</table>

Vasculitis

- Heterogeneous group of multi-system disorders.
- Unwell individual with systemic symptoms and polyarthritis; asthma; vasculitic rash; recurrent pneumonia with sinusitis/ENT disease; haematuria, proteinuria; new peripheral neurological symptoms.

Raynaud’s phenomenon

- 10% of women have primary Raynaud’s phenomenon. Lower risk of progression if onset of Raynaud’s phenomenon or chilblains in teenage years.
- Refer if severe symptoms interfering with activities of daily living.
- Refer if positive ANA.
- Refer if new onset in middle age (especially in men).

Sjogren’s Disease

- Suspect in middle age people with dry eyes, dry mouth; arthralgia; fatigue; recurrent or persistent parotitis. Most will have a positive ANA and raised ESR.

Systemic Lupus Erythematosus, Scleroderma (Systemic sclerosis), Polymyositis

- Heterogeneous group of multi-system disorders.
- Suspect in severe Raynaud’s disease; arthralgia/arthritis; recurrent pleurisy; characteristic rash; proximal myopathy; cytopenias; haematuria, proteinuria; or systemic symptoms. Almost all will have positive ANA; ESR and CRP may be normal.

Investigations

- FBC; U+E; LFT; ANA, ESR; CRP.
- Consider ENA, dsDNA Abs, anti-cardiolipin Abs, complement for Connective Tissue Disorders.
- Consider ANCA for vasculitis.
- Dip urine; consider urgent renal review if haematuria/heavy proteinuria.
Referral Criteria: Connective Tissue Disease and Vasculitis

Referral to General Practitioner

(Referral from triage, community or hospital services specifically for primary care medical review)

Inclusions

- Assessment and management of multi-morbidity and psychiatric co-morbidity.
- Medication reviews and non-urgent prescriptions. Note, for disease-modifying drugs and immunosuppressants initiated in secondary care, there must be an shared care protocol that has been agreed and accepted by both the primary and secondary care clinician responsible for an individual patient.
- Advice regarding achieving and maintaining optimal weight, nutrition, physical activity and healthy lifestyle, including smoking cessation advice.
- Discussion about fitness for work and sickness certification.
- Management following discharge from community or secondary care where no further intervention planned.
- Patients referred back from community services with known or suspected serious underlying pathology where non-urgent (for re-evaluation and possible referral to secondary care).

Exclusions

- Cases where there is significant threat to life, limb, or vital organ. (Emergency referral to A&E or ambulatory care).
- Patients seen in community or secondary care settings who need emergency or urgent assessment e.g. suspicion of inflammatory joint disease, peripheral vascular disease or fracture.
Referral to Community Physiotherapy or First Contact Physiotherapy in Primary Care

**Inclusions**
- Assessment and treatment of functional impairment.
- Assessment of falls and provision of walking aids.
- Improve general fitness and participation in regular physical activity.
- Exercises for enhancing flexibility and muscle strength.

**Exclusions**
- Cases where there is significant threat to life, limb, or vital organ. (Emergency referral to A&E or ambulatory care).
- Severe acute pain or inflammation.
- Patient needs medical opinion.
Referral Criteria: Connective Tissue Disease and Vasculitis

Referral to Rheumatology Interface Service

(May not be available in every CCG)

**Inclusions**

If rheumatological physiotherapy, hand therapy, occupational therapy, podiatry and psychology available:

- Assessment and treatment of functional impairment.
- Assessment of gait and stability, provision of orthotics.
- Assessment and provision of splints and braces.
- Advice and assessment about aids and adaptations to assist activities of daily living and promote independence.
- Psychological interventions (for example, relaxation, stress management) and cognitive coping skills.

If rheumatology nurse or therapist with rheumatology training available:

- Symptom and medication monitoring under shared care arrangement with primary and secondary care.

If rheumatologist or GP with a special interest available:

- Diagnostic assessment and management advice. Patients particularly suited to community assessment are those with vague and undifferentiated symptoms where clinically there is a lower index of suspicion of inflammatory disease. (Patients clinically with a high index of suspicion should be referred to secondary care, unless a community service is specifically commissioned for this group of patients.)

**Exclusions**

- Cases where there is significant threat to life, limb, or vital organ. (Emergency referral to A&E or ambulatory care).
- Severe acute pain or inflammation, unless service commissioned for acute assessment.
- Patient needs medical opinion, unless a medical opinion commissioned.
Referral to Secondary Care Rheumatology

**Inclusions**

- Suspected connective tissue disease or vasculitis. (See history and supporting tests in ‘Background’ above). Negative or absent blood tests should not delay referrals made on clinical grounds.

- Flare-up of connective tissue disease or vasculitis.

- Any imaging performed in primary or community care should be available on IEP/receiving unit, and the reports forwarded with the referral. Any blood tests and pathology performed in primary or community care should be available on ICE/OpenNet and key results forwarded with the referral. Any previous relevant outpatient summary letters and imaging reports should be forwarded with the referral.

**Exclusions**

- Cases where there is significant threat to life, limb, or vital organ. (Emergency referral to A&E or ambulatory care).

- Patient unable to manage at home due to severe symptoms. Discuss with rheumatology service in working hours; out of hours consider intermediate care assessment or other admission avoidance scheme, emergency admission or ambulatory care if critically unwell.