

Standards of Care
for people with
**Connective
Tissue
Diseases**



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ARMA is the umbrella organisation for the UK musculoskeletal community. ARMA is a registered charity No 1108851. Our member organisations are:

- Arthritis Care
- Arthritis Research Campaign
- BackCare
- British Chiropractic Association
- British Coalition of Heritable Disorders of Connective Tissue
- British Health Professionals in Rheumatology
- British Institute of Musculoskeletal Medicine
- British Orthopaedic Association
- British Osteopathic Association
- British Pain Society
- British Sjögren's Syndrome Association
- British Society for Paediatric and Adolescent Rheumatology
- British Society for Rheumatology
- British Society of Rehabilitation Medicine
- Chartered Society of Physiotherapy
- Children's Chronic Arthritis Association
- CHOICES for Families of Children with Arthritis
- College of Occupational Therapy Specialist Section – Rheumatology
- Early Rheumatoid Arthritis Network
- Lupus UK
- Manipulation Association of Chartered Physiotherapists (UK)
- Marfan Association (UK)
- National Ankylosing Spondylitis Society
- National Association for the Relief of Paget's Disease
- National Osteoporosis Society
- National Rheumatoid Arthritis Society
- Podiatry Rheumatic Care Association
- Primary Care Rheumatology Society
- Psoriatic Arthropathy Alliance
- Raynaud's and Scleroderma Association
- Rheumatoid Arthritis Surgical Society
- Royal College of Nursing Rheumatology Forum
- Scleroderma Society
- Society for Back Pain Research

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The contents of this document and further resources including contact details for our member organisations, further information about our work and this project, including additional examples of good practice and resources to support implementation, are available on the ARMA website at www.arma.net.uk.

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Connective tissue diseases – what are they?

The connective tissue diseases are a group of conditions characterised by multi-organ inflammation and autoimmunity. They are generally considered to be uncommon, but are lifelong conditions with significant impact on a person's health and daily life. When acutely active, such conditions can be life threatening and may require immediate access to specialist services to prevent multi-system damage. Symptoms vary depending on the disease, but many share the common symptoms of joint aches and pains, fatigue, muscle pain and weakness, rashes, skin changes and inflammatory changes in different organ systems. All of these conditions can affect children. The term does not encompass inherited conditions such as Marfan's disease or the Ehlers-Danlos group of inherited conditions of connective tissue.

Inflammatory myopathies

Inflammatory myopathies, including dermatomyositis and polymyositis, are a heterogeneous group of acquired diseases of skeletal muscle characterised by weakness and inflammation. They are rare, involving 5-10 per million adults. Dermatomyositis, which includes skin involvement, is seen most often. It can involve internal organs like the heart and lungs. Polymyositis mainly affects only the muscles. Although these disorders can be life threatening, with modern treatment most patients achieve good long-term outcomes¹.

Inflammatory myositis impacts on many aspects of life including employment, social activities and family relationships. Mainly this is due to muscle weakness. But fatigue and general ill-health associated with persistent inflammation and the involvement of internal organs such as the lungs also have debilitating effects². Patients with myositis are seen by several specialists including rheumatologists, neurologists and dermatologists and this creates particular challenges in delivering high-quality care for people with a rare condition.

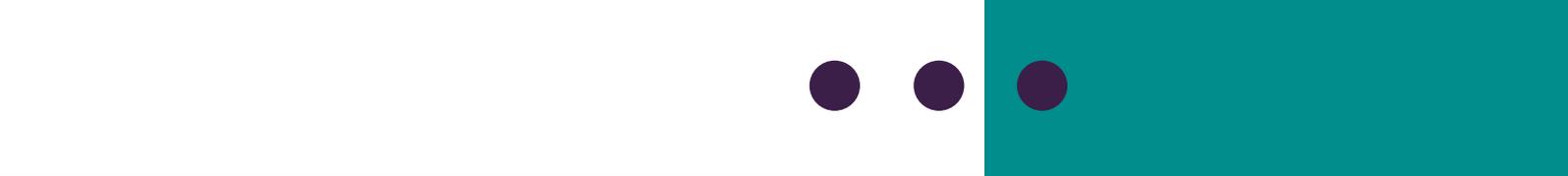
Scleroderma

Scleroderma (systemic sclerosis) is an uncommon connective tissue disease affecting around 1 in 10,000 of the population but has a major impact on those who develop the disease. It is a multi-system connective tissue disease that affects the musculoskeletal system and skin but also can involve internal organs such as the heart, lung and kidneys. It can be life-threatening and has the highest mortality per case of any of the connective tissue diseases.

Scleroderma impacts on almost all aspects of the life of someone with the condition, including employment, social interactions and family relationships by restricting function, causing fatigue and by affecting vital organs such as the heart, lungs, kidneys and gastrointestinal tract. The diversity of scleroderma presents a real challenge to developing recommendations for management but without these, care will be fragmented and inadequate, and there is a real possibility that important treatable aspects of the condition will be neglected.

Sjögren's syndrome

Sjögren's syndrome causes severe dryness of the eyes and mouth with an accompanying arthritis. It usually starts over the age of 50 and prevalence figures between 0.5% and 3% of the population have been claimed. Swelling of the lymph glands in the head and neck and of the parotid gland are well recognised complications, as are poor circulation, fatigue and



neurological complications. It is not usually a fatal disease but is associated with a greatly increased risk (up to 40 times) of a non-Hodgkin's B cell lymphoma.

The treatment of Sjögren's syndrome is principally symptomatic using replacement eye drops and saliva solutions to keep the eyes and mouth as moist as possible. Our increased understanding of the causes of this disease are also leading to the development of some newer therapies for it.

System lupus erythematosus

Systemic lupus erythematosus (SLE or sometimes known just as lupus) is a connective tissue disease which is mostly found in women during the childbearing years. It affects between 40 and 200 people per 100,000, being commonest in the black population. Although the skin and joints are the most commonly affected organs, lupus is a disease which can affect any organ or system.

Though the mortality and morbidity rates for lupus have improved significantly, it retains a life threatening capacity and has major effects on the quality of life of people living with the condition. It causes a huge range of problems from severe fatigue to renal failure requiring renal dialysis and/or transplantation. It is a great mimic of other conditions and one of the ongoing challenges is to recognise the condition as soon as possible and thus to treat it appropriately. Important advances in our understanding of the cause of this disease are leading to some exciting new developments in its treatment.

Vasculitis

The systemic vasculitides (Wegener's granulomatosis, microscopic polyangiitis, Churg Strauss syndrome, Polyarteritis nodosa) are a group of uncommon conditions characterised by inflammation of blood vessels which can lead to organ failure and death. The conditions affect 20 per 1,000,000 persons per year, and 20 per 100,000 overall. They are commoner in the white Caucasian population. The most common organs to be involved are the skin, kidneys and nerves. Treatment involves intensive suppression of the immune system in many cases.

What is the impact of connective tissue diseases?

Connective tissue diseases impact on many aspects of life including employment, social activities and family relationships. Mainly this is due to fatigue, joint pain, and muscle weakness. But fatigue and general ill-health associated with persistent inflammation and the involvement of internal organs such the kidneys and lungs also have debilitating effects. Patients with connective tissue diseases are seen by several specialists including rheumatologists, nephrologists, neurologists and dermatologists and this creates particular challenges in delivering high quality care for people with uncommon conditions.

Why we need Standards of Care

Evidence shows that people's experiences and the quality of care they receive varies a great deal across the UK depending on the approach and configuration of their local services. For people with connective tissue diseases these variations can be a matter of life or death, and will almost certainly have a profound impact on the management of their condition, and

consequently their daily life. The care of people with connective tissue diseases requires the input of a range of health professionals and others in voluntary and public sector support roles, such as advice and employment services as well as self-management.

Whilst there is some good evidence for the effectiveness of many interventions and treatments, for many connective tissue diseases however, the evidence base is limited. Despite this, many services are delivering the innovative and successful management of connective tissue diseases, but to date there has been no agreed common standard of how this should be best achieved.

For many years musculoskeletal conditions have not featured significantly in major health policies across the UK: The National Service Framework for long-term conditions in England mainly focuses on neurological conditions, and no musculoskeletal conditions feature in the Quality and Outcomes Framework of the current UK General Medical Services Contract. This has led to a lack of profile and priority for musculoskeletal conditions in the delivery of services.

The public policy agenda is better now for musculoskeletal conditions than when ARMA first published Standards of Care in 2004. In recent times there have been a number of policies published which have implications for musculoskeletal services. The publication of the *Our Health, Our Care, Our Say* White Paper in January 2006 outlined some important aims: better prevention services with earlier interventions; more choice and a louder voice; tackling inequalities and improving access to community services; and more support for people with long-term needs. These are also fundamental principles within the Standards of Care.

In addition the development of the Musculoskeletal Services Framework for England, a Welsh Arthritis Strategy and the ICATs scheme in Northern Ireland are welcome policy initiatives that will help service providers and commissioners to achieve the ARMA Standards of Care. People with connective tissue diseases are also likely to be affected by other areas of policy, such as the recommendations of the Welfare Reform Bill, which places a heavy emphasis on supporting people back into work.

These Standards of Care aim to bring together existing evidence and good practice to create a framework for services which really meet the needs of the many people who experience connective tissue diseases. Implementation of these Standards should:

- Improve prevention and effective treatment of connective tissue diseases and so improve the quality of life of individuals who are affected by connective tissue diseases
- Identify for people with connective tissue diseases the care and treatment they can expect
- Enable the NHS to make more effective use of resources by helping to prevent avoidable disability and by reducing the number of return GP consultations and hospital appointments due to connective tissue diseases
- Promote consistent advice and treatment
- Reduce levels of disability due to connective tissue diseases
- Improve productivity and reduce the benefits bill by enabling people to stay in work.

1 Dalakas MC, Hohlfeld R, Polymyositis and dermatomyositis. *Lancet*, 2003, 362, pp. 971-982.

2 *ibid*

ARMA's Standards of Care for people with connective tissue diseases are intended to support people of all ages with connective tissue diseases to lead independent lives and reach their full health potential through:

- access to information, support and knowledge that optimise musculoskeletal health for everyone and enable self-management
- access to the right services that enable early diagnosis and treatment
- access to ongoing and responsive treatment and support.

The Standards define what services are appropriate under these three themes and suggest ways of providing them effectively and in a measurable way in the form of key interventions. A detailed rationale for the Standards draws on available evidence and examples of good practice drawn from ARMA's ongoing call for good practice: a database giving details of these and other examples is available at www.arma.uk.net.

The Standards are not guidelines, or algorithms of care, though they refer to these where available.

The Standards of Care for people with connective tissue diseases form part of a suite of Standards; other Standards published to date are for back pain, inflammatory arthritis, metabolic bone disease, osteoarthritis and regional musculoskeletal pain.

The Standards acknowledge the fact that those planning and delivering services around the UK face differing demographic, geographic and economic factors, which will affect how the Standards are implemented in each locality. We hope the Standards will act as a tool for all stakeholders - service users, providers, commissioners and policy-makers to work together to review and improve their local musculoskeletal services.

Key principles – the user-centred approach

The project has been driven by the needs of people living with musculoskeletal conditions. It began with the establishment of a set of key principles for care, developed by a group of people living with musculoskeletal conditions and consulted upon widely. These principles have underpinned the development of each set of condition-specific Standards. The key principles, which can be found on ARMA's website www.arma.uk.net, affirm that 'patients' are individuals who need different types of advice and support at different times; and who need integrated services providing advice and support that cover all aspects of managing and living with the condition – clinical, personal, social and employment/education. In particular, the Standards recognise that health services play a key role in supporting people to maintain or return to employment or education.

Nevertheless, while these standards focus on health services, it must be recognised that people with connective tissue diseases and other musculoskeletal conditions have wide-ranging needs. Social care often plays a key role in ensuring people can remain as active and independent as possible. Factors such as access to transport and the built environment may have a major impact on quality of life. More work is needed to understand and meet these needs.

Musculoskeletal conditions affect families and carers as well as individuals. Indeed, many people with these conditions may be carers themselves. The Standards do not make specific recommendations on issues relating to carers: this also needs to be the subject of further work to ensure that carers' needs are understood and addressed.



How the Standards were developed

The Standards of Care for people with connective tissue diseases were developed by an expert working group, facilitated by ARMA. The group included people with connective tissue diseases, representatives from user organisations, experienced service providers and experts from many professions, from around the UK. Starting with a review of the needs of people with connective tissue diseases, the group met four times between December 2005 and June 2006 to determine evidence-based Standards to meet those needs, consulting widely and publicly on the drafts. The Acknowledgements on page 21 give details of the working group membership.

Clinical experts have identified the evidence base, including relevant guidelines for the management of connective tissue diseases. References are shown as footnotes in the Standards. Evidence has not been graded for the purposes of this document. For further details on the evidence base, please refer to the references quoted in the document.

The resulting Standards are therefore based firmly on the experiences and preferences of people with connective tissue diseases, and on evidence and good practice where this is available. ARMA plans to review these Standards in 2009, or sooner if there are significant developments in care for people with connective tissue diseases.

Next steps

The publication of these Standards is the beginning of an ongoing programme involving the whole community to improve musculoskeletal services. We are circulating the Standards widely to people with musculoskeletal conditions, doctors, allied health practitioners, providers and commissioners of health services, voluntary organisations and policy makers. We will publish audit tools to support the Standards' implementation. We are also collecting and sharing examples of good practice, which are accessible to everyone through our online database.

We invite all stakeholders to make a commitment to implementing the Standards. First steps might be to:

- audit existing services
- identify champions for change in musculoskeletal services, and set up a working group to develop your local strategy and priorities
- work in partnership with all stakeholders, including national and local voluntary organisations, to involve service users in designing and developing services.

Above all share your success! Tell us about your initiatives; send us examples of good practice; help to build a national resource for high-quality musculoskeletal services.

Promoting awareness

Standard 1

Health and community services, the voluntary sector and other agencies should provide information about connective tissue diseases through a range of media, which should include advice and support. They should also be made aware of local and national service user support organisations.

Information on services, treatments and providers

Standard 2

People with connective tissue diseases should have access to information and guidance which enables them to make informed choices on a range of service providers including

specialist services with an expertise in their condition, the treatments they offer and the facilities in which they are delivered.

Promoting health

Standard 3

Connective tissue diseases are complex and unpredictable conditions. Health, social and education services and voluntary organisations should make information available to the public (in a range of languages and formats) on risk factors that could impact on their health. This should be standardised and include information on physical activity and exercise and nutrition. It should be available in many settings including pharmacies, GP surgeries, schools and colleges.

The rationale

- Connective tissue diseases are complex and unpredictable conditions. Access to information about them and about available support groups can enable those with the condition to play a greater role in the management of their condition.
- All people with connective tissue diseases should have the skills to influence positively their own health and lifestyle, through knowledge, informed consent, and access to appropriate information.
- Smoking and obesity are risk factors for complications in connective tissue diseases, for example kidney disease and premature heart attacks or strokes in lupus³.
- Connective tissue diseases affect a wide range of people, for example there is an increased incidence of lupus in some minority ethnic groups^{4,5}. It is therefore vital that information is available in a range of formats and languages to meet the needs of all people with the conditions.
- Raising awareness of connective tissue diseases amongst the public and wider health community can help in the recognition of symptoms, and the referral to an appropriate unit or specialist centre. This early identification will improve outcomes for patients and could be life-saving.
- Raising the public profile of connective tissue diseases should also increase employers' understanding of the diseases and enable them to support staff members with the condition.
- In order for people with connective tissue disease to become active and equal partners in their own care they need to be well-informed about their condition, empowered to take

responsibility for their musculoskeletal health, and able to make informed choices about treatments, providers and settings for care.

Putting the Standards into practice: **key interventions**

- i Health and community services should make information available to the public on signs and symptoms of connective tissue diseases and where and when to seek professional advice. This information should be available in a range of formats and languages.
- ii Health promotion campaigns should include information about the increased risk of cardiovascular disease, and highlight that stopping smoking and controlling blood pressure, cholesterol and weight reduce these risks⁶.

Good Practice Example - A Lupus and Scleroderma Education and Self-management Programmes

A team including a rheumatologist, nurse specialist, occupational therapist, physiotherapist and patient educators have developed an education and self-management programme for people with lupus and scleroderma. The programme was developed with input from people with connective tissue diseases and the patient educators lead some of the sessions.

This initiative offers people with connective tissue disease both education and advice about their conditions, as well as the opportunity to learn about self-management and find effective ways to live with chronic conditions. The programme consists of four three-hour sessions plus a three-hour follow up at 6 months to review changes resulting from the programme.

A
good practice

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3. Bruce I. Not only... but also: factors that contribute to accelerated atherosclerosis and premature coronary heart disease in systemic lupus erythematosus. *Rheumatology*, 2005, 44, pp. 1492-1501.
 4. Ehrenstein MR, Isenberg DA. Systemic Lupus Erythematosus in adults - clinical features and aetiopathogenesis. In Isenberg DA, Maddison PJ, Woo P, Glass D, Breedveld FC, *Oxford Textbook of Rheumatology*, 3rd Edition. Oxford: Oxford University Press, 2004.
 5. Johnson AE, Gordon C, Palmer RG, and Bacon PA. The prevalence and incidence of systemic lupus erythematosus in Birmingham, England, Relationship to ethnicity and country of birth. *Arthritis and Rheumatism*, (1995, 38(4), pp. 551-558.
 6. Symmons D, Bruce I. Management of cardiovascular risk in RA and SLE - *arc Report on Rheumatic Diseases*, Series 5, 2006.

Access to diagnosis

Standard 4

Health professionals in primary care should be well-trained and informed about the full range of connective tissue diseases. They should recognise and be able to screen for the clinical features which may indicate connective tissue disease and know how to access appropriate secondary care services. Their services should be accessible at times and locations which are appropriate to the needs of service users.

Health professionals in primary care should be able to identify people who require immediate/emergency access to secondary or specialised care.

Integrated care

Standard 5

People with connective tissue diseases should experience a seamless service in the management of their condition. Primary and secondary care services, including the full multidisciplinary team, should be

well-integrated with specialist centres, from whom they should receive information and advice. Clinicians in both secondary care and specialist centres should be well-integrated into national and international practice and research networks.

Assessment of needs

Standard 6

Upon diagnosis, people should be offered a full assessment of their condition, general health, psychosocial and pain management needs. These assessments should consider referrals or communication with other services. For children and young people, specific attention should be paid to their educational, emotional and social development needs. Comprehensive regular specialist reviews should be continued, involving all relevant specialists. This should be a long-term process. Children and young people should be managed in an age-appropriate environment. Adolescents should have a choice about whether they are managed in a paediatric or adult service⁷.

The rationale

- Early diagnosis and access to specialist services can improve outcomes and reduce life-threatening events related to connective tissue diseases. Therefore a delay in diagnosis may be dangerous. Many connective tissue diseases can present with multiple and varied symptoms⁸ and people with the conditions often attend numerous physicians in an attempt to gain a diagnosis.

GPs should have access to appropriate specialist units where information can be gained about appropriate screening assessment in primary care, prior to referral to secondary care services. This should improve early diagnosis rates in primary care.

- The quality of care is improved with integrated care across primary, secondary and tertiary services. This leads to improved outcomes for people with connective tissue diseases.

Developing specialist centres which provide a “hub” of support and information should enable the “spoke” of primary and/or secondary care to manage those with connective tissue diseases more appropriately in the community.

Provision of information

Standard 7

Those diagnosed should be given an appropriate, timely explanation of their condition and its treatment, including medication, and should be given further written information for them to read after the consultation. They should also have access to someone of whom they can ask general questions about their condition. They should be given information about education, advice and support services and about patient organisations that offer advice and support services. They should also be offered and have prompt access to counselling services (see also Standard 19).

Individualised care plans

Standard 8

A plan for ongoing care should be offered and agreed between the patient and the multidisciplinary team. This should give constructive messages about the patient's condition and the roles for the patient and the multidisciplinary team and other relevant parties, e.g. family or employers, in the management of their condition. This care plan

should draw on good clinical practice, and consider any alternative or complementary therapies that the patient may be taking. It should be based on current guidelines and evidence where available.

Support to remain in, or return to, education or work

Standard 9

People with connective tissue diseases should be encouraged to remain active and mobile and be supported to remain in or return to education or work – either paid or unpaid. They should have access to information and services such as occupational therapy and support and rehabilitation services and access to a social worker to advise on accessing benefits and other services.

Rehabilitation services should work with people who have connective tissue diseases and their employers or educators to support them to stay in work or education. This might include advice and support and retraining. In particular, employers should be made aware of the fluctuating nature of connective tissue diseases.

- Living with a chronic condition can present many problems for the individual. Connective tissue diseases are complex and it can be difficult to recognise the symptoms that require immediate medical help. Access to nurse specialists and other members of the multidisciplinary team in specialist units should encourage knowledge and self-management skills⁹. Information about relevant patient organisations can provide the individual with access to clear support¹⁰.
- Patients should be given an explanation of their disease, its treatment, its impact on general health and potential effects on their social functioning. It is essential in long-term management of all patients to have a plan of care which may need to be modified in response to requirements, depending on the nature of the disease in question and variation in disease activity or severity as a result of treatment. Individual fully informed consent to any interventions, especially potentially toxic treatments such as Cyclophosphamide, must always be sought, in line with local policy.
- Access to a multidisciplinary team that is knowledgeable in connective tissue diseases enables a plan to be developed that is shared between the individual and the team. When a collaborative approach is applied and the person with the connective tissue disease is well-



informed about their condition and their role in managing it, better health outcomes are achieved.

- Those with chronic conditions require access to high quality support services that can work with individuals and their families to enable them to fulfil employment, family and social responsibilities according to individual needs.

Education and work can be severely compromised through illness and all people with connective tissue diseases should be enabled to remain in or return to the work of their choice where possible. This will require significant input from occupational therapists, rehabilitation teams and social workers.

Some people with connective tissue diseases may need to adapt to a significantly changed lifestyle and may require access to support and counselling to help them manage these changes.

Putting the Standards into practice: **key interventions**

- iii Health professionals in primary and secondary care, as well as service commissioners, should receive education and training to improve their knowledge and understanding of connective tissue diseases. Undergraduate medical education programmes should include education about recognising connective tissue diseases.

Primary and secondary care providers, including the emergency services, should be aware of the diagnostic features of the commonly seen connective tissue diseases and recognise 'red flags' that require immediate specialist support and management. This should include an early warning system for both newly-diagnosed patients and those with long-standing disease.

A specialist service for connective tissue diseases should be considered essential for most district general hospitals. In addition, outreach clinics and shared care should enable people with connective tissue diseases to be seen as locally as possible so that they do not have to travel great distances for their health care. As part of this, regular meetings between primary, secondary and tertiary care health professionals should be encouraged.

Red Flags

Any sudden change or worsening of the symptoms described below, particularly if involving more than one part of the body would be an indication for urgent assessment:

- Acute abdominal or chest pain
- Blood or protein in urine
- Breathlessness
- Unexplained fever
- Change in mental function
- Weight loss
- Severe generalised or sudden localised weakness
- Severe hypertension, particularly in scleroderma
- Sudden loss of hearing or vision
- Painful, cold, blue/black fingers and/or toes

- iv Early referral to appropriate units is essential for effective management of complex connective tissue diseases. Those with inactive quiescent disease also need to be managed in primary care, where physicians should be able to access secondary and tertiary specialist centres as necessary. By utilising a hub and spoke model of care, the expertise of specialist units should be transparent and supportive to the primary care team. Networks such as these should be

encouraged. Patients with apparently quiescent disease should have at least an annual assessment in a specialist centre in order to detect and address developing problems which may be clinically silent.

- v People being assessed for connective tissue diseases should have core assessment of weight, blood pressure, and urinalysis at every clinic visit. Referral to other members of the multidisciplinary team should be strongly considered in the management of the physical and psychosocial effects of living with an unpredictable chronic illness¹¹.
- vi Specialist connective tissue disease healthcare practitioners (consultant, nurse specialist) should be available to assess all those referred with connective tissue diseases. The practitioners should also provide an advice service to those in primary care (liaison nurses would be an excellent way of achieving this). Self-help groups should be integrated into secondary care settings, offering patient advice and support to those who require it. Health professionals should provide written advice, including user groups' information leaflets¹².
- vii All people diagnosed with connective tissue diseases should have a clear care plan that will be developed with their input. This should include current management plan and reasoning, setback plans and management of acute episodes that may require immediate medical attention. They should receive a copy of their plan which should be regularly reviewed. They should also be informed of who they should contact and when to contact them (including out of hours and at weekends) if they have any concerns about their condition.
- viii Rehabilitation services should be well-developed and well-informed about connective tissue diseases.

Employers and education providers should be supported in understanding the fluctuating

Good Practice Example - B Scleroderma Outreach Clinics

Specialised tertiary care services provide an opportunity for people with scleroderma to be seen by a specialist in a local setting. Outreach clinics staffed by scleroderma specialists are held every two weeks in rheumatology departments in secondary care centres, encouraging dialogue and expertise sharing between clinicians in secondary and tertiary care. This service provides an example of a working hub and spoke model which also helps build good relations within the musculoskeletal community.

B
good practice

Good Practice Example - C Drop-in Clinics

Three departments around the country have set up monthly drop-in clinics for people with lupus. The clinics are organised by people with lupus and include both medical and nursing support. This forum provides support for patients with lupus to discuss their problems together and to have trained medical staff on hand to help with any complicated problems. It is not designed to give an "extra" clinic appointment but rather, helps provide education and understanding of the problems faced by other people with the same condition. The drop-in clinics have been met with great enthusiasm by the patients and are always well attended.

C
good practice

nature of connective tissue diseases and in giving people with them a degree of flexibility when they are experiencing an acute episode of their disease.

- ix Service providers need to be fully briefed on the nature of connective tissue diseases so that the appropriate business cases can be made to develop and support services that meet the needs of people with the conditions, for example, to gain sufficient funding for treatments and specialist services.

good practice D

Good Practice Example - D Combined Clinics

Several departments around the country have set up combined clinics for people with connective tissue diseases. Some examples include rheumatology and renal clinics for people with scleroderma, lupus and/or vasculitis; rheumatology and dermatology clinics for psoriasis, lupus, scleroderma or vasculitis; rheumatology and ophthalmology clinics for uveitis and/or vasculitis; and rheumatology and respiratory for Churg-Strauss (vasculitis) and TNF risk.

One such combined clinic enables a senior rheumatology trainee to spend one session per week in a pulmonary hypertension clinic. People attending the pulmonary hypertension clinic often travel long distances to attend and about 80% have pulmonary hypertension related to a connective tissue disease. The combined clinic brings a senior trainee with experience of diagnosing and managing the connective tissue diseases that can result in hypertension together with the experts in the hypertension clinic.

This helps enable accurate diagnosis of underlying connective tissue disease and management of any other rheumatological problems. It also reduces travel time and cost for people with connective tissue disease who attend the combined clinic. The service has reduced the number of outpatient appointments scheduled in both the rheumatology and cardiology departments and has improved relations between the two.

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12 Dziadzio M, Fligelstone K, Smith R, Ensz S, Denton C, Black C. *Internet as a tool for patient support groups to provide quality information*. Abstract presented at EULAR Congress, Social Leagues in Berlin, 2004.

Involvement of people with connective tissue diseases in service development

Standard 10

Healthcare organisations should involve people with connective tissue diseases in the planning and development of connective tissue disease services. People who wish to participate in this process should be well-supported and have access to self-help groups and other voluntary organisations.

Access to specialist services

Standard 11

There should be access to at least one nominated physician in each district general hospital with specialist knowledge of connective tissue diseases. They should have access to and regular communication with regional specialist centres and they should refer on to these centres where appropriate. For children and young people, each district general hospital should have a paediatrician who contributes to a paediatric specialist clinical network. Emergency services should always be available for specialised connective tissue disease advice on immediate patient care.

The multidisciplinary team

Standard 12

A holistic approach should be used to manage connective tissue diseases. People with the conditions should have ongoing and emergency access to the local multi-disciplinary team who have training in the management of connective tissue diseases. They should have access to a complete range of multi-organ specialists that can be provided locally and nationally, not forgetting such areas as cosmetics (camouflage make-up) and reproductive health. There should be adequate time available in all consultations to discuss all of the patient's symptoms.

Fertility advice and support

Standard 13

Women should receive good advice about contraception and pre-pregnancy counselling, specifically about the optimal age and conditions for pregnancy, the effect of pregnancy on their connective tissue disease and the effect of their connective tissue disease on their pregnancy. All professionals who care for men and women with connective tissue diseases should consider the effects of their treatments on reproduction. There should be connective tissue disease pregnancy clinics in tertiary specialist centres.

Developing treatments

Standard 14

Units treating connective tissue diseases should be encouraged to take part in clinical trials and contribute to national and international clinical studies.

Access to novel treatments

Standard 15

People with connective tissue diseases, including children and young people, who are not responding to standard treatments should have access to off-label and non-licensed drugs recommended by their specialists. A system for expert review of these treatments and appropriate funding mechanisms should be in place.

Benefits and risks

Standard 16

People with connective tissue diseases should be fully informed about the benefits and risks of taking and not taking both standard and off-label treatments. They should be made aware of possible side effects, and their informed consent should be gained before treatment.

Pain relief

Standard 17

People with a diagnosis of connective tissue disease should be offered a choice of pain management strategies and symptomatic pain relief, if relevant. These should be in accordance with the best available evidence and national/international guidance and guidelines.

Palliative care

Standard 18

Palliative care services, including hospices, should be available for people with connective tissue diseases, where necessary. They and their families or carers should be made aware of the options available to them, and should be supported by specialist nurses in the community.

Self-management

Standard 19

People with connective tissue diseases should have ongoing access to information, advice and support for self-management which helps them to recognise, manage and avoid any complications of their disease or its treatment. This could include:

- A local or national nurse-led advice line
- National and local voluntary organisations,

including access to other people with their condition

- Self-management training, e.g. the Expert Patient Programme

They should be supported to make any necessary life style changes, including diet, employment etc. and helped to manage fatigue, depression and other psychosocial problems that can arise. This information and support should also be available to their families and carers.

Rehabilitation and support

Standard 20

Timely access to community support services, including health care, social and rehabilitation services, and voluntary organisations should be available to help people with connective tissue diseases to maintain or regain independence as quickly as possible.

Key healthworker

Standard 21

Individuals with complex conditions should have a key health worker who can support them and coordinate their access to the full range of health and social services. This should be available to people with co-morbidities and complications arising from their condition or its treatment, and/or people whose condition has become disabling.

The rationale

- The main aims of ongoing treatment are to control inflammation and to maintain and restore function. Therefore people need ongoing follow-up by the multidisciplinary team in primary or secondary care, as appropriate. Access to a health professional trained in footwear assessment should be considered.
- Anyone with a chronic illness should be encouraged to develop the skills to manage their own disease appropriate to personal needs. Where this happens there are often improved clinical and patient outcomes.

Support and advice can be given in a number of ways. For example, support may be provided directly by a specialist advice line and/or by enrolment on self-management programmes that empower individuals to manage their illness.

- Involving service users and their organisations can lead to improved service provision¹³. They can be helpful in developing services that best meet the needs of people with the conditions.

People with the same disease can support each other in a different way from health professionals. Self-help and other service user groups provide an essential service to support individuals, families, carers and also support scientific and patient-based research.

- Connective tissue diseases are highly complex, multi-system diseases that can be life-threatening. Specialists with expertise in these areas must be available to maintain the health of those with connective tissue diseases.
- People with connective tissue diseases require access to a wide variety of health professionals who work together to support and manage the complexity of effects on the health of the individual. This can include different medical specialities as well as the full range of health professionals. Improved coordination of services should lead to improved service quality and health outcomes.
- People with connective tissue diseases require regular review both in primary and secondary care. A review that assesses the disease activity and damage, as well as the social effects of the condition and its treatment, is essential in forming a management plan for treating and supporting people with connective tissue diseases. Appropriate and timely referral to other members of the multidisciplinary team, such as occupational therapy, podiatry, physiotherapy and psychology is essential in order to maintain good health.
- People with connective tissue diseases present with many health problems that may or may not be related to their condition. The multidisciplinary team should work with them to identify these problems, discriminate between them and address them.
- People with connective tissue diseases often need access to more than one specialist. It is therefore important that they have an advocate that can negotiate on their behalf to access the appropriate parts of the health service. Coordination of their care in this manner can reduce the risk of complications and provide people with greater continuity.
- Due to the relative rarity of connective tissue diseases, the evidence base for treatment is limited and slow to accumulate. There are consequently few instances where an ideal treatment exists for many connective tissue diseases. In addition, many treatments are unlikely to be appraised by the National Institute for Health and Clinical Excellence (NICE). It is therefore essential to continue efforts to understand disease mechanisms and to develop and test new therapies and management strategies. Tertiary treatment centres and secondary care units, where appropriate, should be able to initiate such novel therapies which may be life-saving.
- Informing and educating patients about their treatments improves concordance^{14,15}. It is therefore vital that people with connective tissue diseases have access to information that enables them to understand the benefits and risks of both off-label and conventional drugs.
- Having access to a range of pain management strategies can lead to improved quality of life for people with connective tissue diseases.

- Support for patients with chronic damage as a result of connective tissue diseases is vital. For example, patients that reach end-stage renal failure require a comprehensive renal replacement programme. Similarly, the long-term effects of neuropathy may require long-term intervention from physiotherapists.
- Various complications of connective tissue diseases can lead to a significant impact on an individual's quality of life. People with the conditions need to be able to plan their own independent life as much as possible, with a variety of agencies providing support.
- In terminal states, those with connective tissue diseases should have access to the palliative care services that provide support and comfort to those with other end-of-life diseases¹⁶.

Putting the Standards into practice: **key interventions**

- x Self-help and service user organisations should be supported in their work and well-connected to health service providers.
- xi All district general hospitals should have a specialist service for connective tissue diseases as well as close links to specialist centres with expertise in connective tissue disease management.
- xii People with connective tissue diseases should have access to cross-specialty clinics that involve the full multidisciplinary team and are able to undertake a comprehensive investigation of their disease progression. Appointment times should be long enough to discuss the full range of symptoms and address all of the patient's needs. An absolute minimum appointment time for new patients should be 40 minutes, and 20 minutes for follow-up patients.

People with complex connective tissue diseases should have access to a specialist through their local physician or GP in order to deliver appropriate management plans when in crisis. Specialist connective tissue disease services should promote awareness of their expertise and availability to acute service providers.

- xiii An expert practitioner in connective tissue diseases should be available to act as coordinator for the individual with the condition, and help manage their progress through the health service.
- xiv All physicians should discuss the potential effects of the connective tissue disease and its treatment on the ability to conceive and the risk of drug-induced congenital abnormalities and any problems related to breastfeeding before starting treatment¹⁷. Pregnancy tests should be offered to female patients before starting drugs that are not safe in pregnancy.

Advice on contraception should be available from their specialist and/or primary care, to patients with connective tissue diseases to avoid unplanned pregnancies at inappropriate times (related to disease activity and/or drug therapy). Physicians need to be aware of the risks of the diseases and their treatment on fertility, pregnancy and breastfeeding and should refer female patients to an obstetrician interested in foetal and maternal medicine for specialist advice¹⁸.

- xv Units treating connective tissue diseases should be encouraged and resourced to take part in clinical trials and contribute to national and international clinical studies.
- xvi Specialist units providing a lead in multi-centre research studies for connective tissue diseases

should be networked nationally and internationally to ensure the sharing of knowledge about therapies and their rationale as they develop.

Funding guidance should be available for expensive treatments through local and national processes, in addition to those who commission services.

Some of the possible treatment options in managing rare complications are through off-label and non-licensed drugs that people with connective tissue diseases should have timely access to. Commissioners should be informed about the complexities of such disease management and the impact on the individual. This should enable them to make necessary funding arrangements to support the use of these treatments.

- xvii Those with connective tissue diseases should be aware of the complexities of treating rare disease and therefore the need to use novel drug treatments. Materials should be developed to inform them about the benefits and risks of the different treatments available.
- xviii Health service providers should make a range of pain management strategies available to people with connective tissue diseases.
- xix End-of-life management strategies should be available to those with connective tissue diseases, where necessary, in order to enable people with them to have a respectful death. Links should be established between connective tissue disease teams and palliative care teams. Hospices should be encouraged to accept patients with connective tissue diseases¹⁹.
- xx Self-management strategies are essential to enable service users to regain independence and plan their own lives. The use of self-management education programmes such as the Expert Patient Programme should be available. Other disease specific programmes such as education or self management should be encouraged in specialist units nationwide²⁰.
- xxi Specialist service telephone advice lines should be established at a local and national level^{21,22}.
- xxii Connective tissue disease clinics should be established to enable the regular or annual monitoring and assessment of people with connective tissue diseases, where their condition can be efficiently evaluated using standardised assessment measures. Some treatments may not be available through secondary care services. Therefore access to a

Good Practice Example - E Community education and follow up of patients with inflammatory arthritis and connective tissue diseases

A multidisciplinary musculoskeletal unit has developed nurse-led education for people with connective tissue diseases held in their home, after patient questionnaires revealed that people had difficulties attending hospital-based programmes.

The home-based education programme takes place over three visits and is tailored to each individual, using booklets and in-house publications. People participating in the programme can also access a helpline, and nurses can respond via telephone or additional home visits.

The nurses can also implement medication changes or intramuscular steroid injections authorised by the consultant while visiting the patient at home, and they can make referrals directly to any member of the multidisciplinary team or book urgent outpatient appointments.

specialist who is involved in multi-centre national and international collaborative working groups is essential.

- xxiii Where necessary, those with connective tissue diseases must be able to access support and rehabilitation services in their local area. (This might also include support for renal therapy and home oxygen where appropriate). Self-help groups and service user organisations can also provide useful personal support.

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Allied Health Professional (AHP)

a person who offers advice and clinical care who is not necessarily medically qualified. For example, this would include a nurse, pharmacist, physiotherapist, occupational therapist and podiatrist. These healthcare professionals are registered with, and regulated by the Health Professions Council. Other healthcare professionals, such as manual therapists (osteopaths and chiropractors) should also be qualified and registered with their own professional body.

chronic illness

a long-term illness for which there is no cure but where the activity of the disease can be controlled.

clinical feature

involvement of part of the body that can be detected by careful examination or investigation.

community services

services that are based in or around primary care.

co-morbidity

other diseases that one person might have.

complications

secondary conditions or negative reactions occurring during the course of the condition.

concordance

a process by which a person with a connective tissue disease and their health care professionals agree on the best use of medicines and treatments.

congenital abnormalities

conditions that develop at or before birth. They are often inherited.

cross-specialty (or combined) clinics

a clinic in which specialist from two or more disciplines are present at the same time. For example, a clinic with two or more specialists such as nephrology and rheumatology.

Expert Patient Programme

the name given to an initiative to help people with long-term conditions maintain their health and improve their quality of life. A key element of this initiative is lay-led self-management training whose primary aim is to facilitate the development of self-management skills rather than to provide medical information.

hospice

a healthcare institution specialising in palliative care for people with life threatening or life limiting conditions.

hypertension

high blood pressure.

integrated services

services which achieve seamless care across primary, secondary and tertiary care.

interventions

a general term covering treatments, advice, education and other care that a practitioner may give.

key health worker

health professional who co-ordinates the care of a person with a connective tissue disease.

multidisciplinary team

a healthcare team that includes professionals from different disciplines working together to provide a comprehensive service for people with connective tissue diseases. The team may include physicians, doctors in training (both hospital and GPs), GPs with a special interest in rheumatology (GPwSI), nurse specialist, physiotherapist, occupational therapist, dietician, podiatrist, orthotist, psychologist, pharmacist and social worker.

multi-system diseases

conditions involving more than one part of the body or organ, i.e. muscles, lungs, heart, kidneys, skin, etc.

neuropathy

a disease of the peripheral nerves (those outside the brain and spinal cord), usually causing weakness and numbness.

non-licensed drugs

a drug which has not been approved by the drug regulatory authorities (Medicines and Health Care products Regulatory Authority (MHRA) in the UK) for use in the particular connective tissue disease/condition.

nurse specialist

a nurse who has advanced knowledge and competence in a particular area of nursing practice, such as rheumatology.

off-label drugs

a drug which has licence but is being used outside the conditions for which it is licenced.

opioids

a group of medications that can be prescribed for strong pain control.

outreach clinics

a service arrangement in which a connective tissue disease specialist travels to a hospital to share expertise where there is no existing specialist clinic. Outreach also covers secondary to primary care, as with a hub and spoke arrangement where specialists travel from their base to other neighbouring health care facilities. This can be either from tertiary care to secondary care; or a secondary care physician visiting a community-based facility.

paediatric specialist clinical network

a network of linked clinicians via a hub and spoke arrangement.

primary care

care services available in the community, for example through a community pharmacist or the care provided by a GP. This is often a person's first point of contact for advice, information and treatment.

psychosocial support

professional care which addresses a person's psychological and social health needs. This may include support to reduce a person's distress, fear or ability to cope, support for social and family relationships, and support/advice about employment or benefits.

quiescent

a term describing a disease that is in an inactive or undetectable phase.

red flags

a group of symptoms or signs (clinical indicators), any one or more of which may suggest a possibility of serious disease.

secondary care

care available usually in a hospital setting. People generally need referral from a professional in primary care.

self-management

learning a range of techniques to help manage life with a condition more effectively, including exercise, eating healthily, etc.

specialist centre

a clinic/unit in a hospital specialising in specific connective tissue disease conditions.

symptomatic pain relief

relief of pain. This may be achieved by use of analgesics (pain killing drugs), injections or complimentary therapies such as acupuncture.

treatment

drugs or other interventions intended to alleviate the symptoms of a connective tissue disease.

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Standards of Care
for people with

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