

2021

Making the Case for Better Lupus and Rare
Auto-Immune Disease Provision for
Patients in Wales

FTWW

FAIR TREATMENT FOR
THE WOMEN OF WALES

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Foreword

Since I was diagnosed with a form of lupus in 2013, I have struggled to understand certain aspects of NHS Wales management of the condition. I have been denied a second opinion from a lupus expert many times and ended up paying privately. As a result, my diagnosis changed from Sub-Acute Cutaneous Lupus Erythematosus (SCLE), which primarily effects the skin and is usually only symptomatic in the summer months, to SLE or Systemic Lupus Erythematosus, a systemic chronic Connective Tissue Disease / Auto-immune Disease.

The more I tried to educate myself about my chronic condition and discover the reasoning behind the decisions made in respect of my NHS care in Wales, the more confused and frustrated I became. It is rarely possible to access services outside of your own health board, even if it does not offer specialist care. The low numbers accessing those services is then given as a reason for not investing further, leaving patients with nowhere to turn.

I quickly realised I was not the only one in this position when I started Pembrokeshire Lupus Support Group in September 2017 and began raising awareness of the situation. More and more people contacted me, telling me I was doing something very worthwhile. Lupus patients thanked me. I could not help but be moved by some of their experiences and this has spurred me on.

All the people with whom I have engaged on this journey agree on one thing: the current system for people living with lupus and other rare auto-immune diseases in Wales is not working. We do not feel that we are diagnosed, treated, or monitored correctly. Our condition is not taken seriously enough. We do not think that national Guidelines for the Management of Adults with Lupus are followed. We feel ignored. We feel neglected. We feel misunderstood. We feel alone. We all think we deserve better. My wish is to work with those involved in the provision of lupus care in Wales, to improve things for all of us. To join this campaign, please visit Appendix 1.

I would like to thank you for reading this report which contains an outline of current practice in Wales, patient testimonies, and a series of recommendations for improved care provision for lupus and, by extension, other rare auto-immune diseases.

Wendy Diment

Report Author

On behalf of Lupus UK, Wendy runs two support groups in Wales, in Pembrokeshire and Carmarthenshire. She also works closely with the charity's Cardiff-based Support Group Co-ordinator. In May 2020, Wendy became a Trustee for Lupus UK and, a year later, was appointed Vice-Chair. A long-standing member of Fair Treatment for the Women of Wales, Wendy became FTWW's Auto-Immune Conditions Adviser and Campaign Lead also in 2020. Wendy has featured in various media coverage of long-term illness and disability in Wales, including BBC News Online where she discussed how Welsh Centres of Excellence could 'extend lives' for lupus patients, and ITV's Wales This Week looking at the impact of the pandemic on numbers of people living with chronic conditions.

Part 1: Introduction and Aims of this Report

Where possible, this report makes clear that references to Lupus should also include other Rare Auto-Immune Rheumatic Diseases (RAIRDs), in particular all connective tissue disease, i.e. Undifferentiated Connective Tissue Disease (UCTD), Mixed Connective Tissue Disease (MCTD), Sjogren's, Vasculitis, Myositis, Scleroderma, and Raynaud's.

All of these rare auto-immune illnesses are represented by RAIRDA – the Rare Auto-Immune Rheumatic Disease Alliance, formed by the British Society for Rheumatology (BSR), Vasculitis UK, Scleroderma & Raynaud's UK, LUPUS UK, and the British Sjogren's Syndrome Association.

Lupus specifically is estimated to affect 25-96 people per 100,000. The higher end of the range comes from a study in 2012 by Dr Frances Rees (1). This would suggest approximately 1 in 1,000 of the UK population have lupus. Population demographics are an important consideration because the prevalence is approximately 3-4 times higher in people of Black and Asian heritage.

The mean age at diagnosis in the general population is 48.9 years but is lower in those of African ancestry in the UK and North America. Patients with African ancestry tend to present younger and are at considerable risk of developing more serious kidney involvement. As an estimate, the total number of lupus patients in Wales is 3,100.

Auto-immune conditions are of particular interest to FTWW, as data shows that around 80% of those living with these diseases are female. Historically – and globally – there has been little investment in research or services for benign health conditions which predominantly affect women, to the detriment of all patients affected, irrespective of gender.

Sadly, reports of feeling ignored, dismissed, and not being taken seriously are rife in women's accounts of healthcare services across the UK and beyond, and Lupus is no exception. Baroness Cumberlege's 'Do No Harm' report (2) laid bare widespread systemic failings in the healthcare system, where women's concerns were ignored and where patients, even now, are being put at risk. The inquiry blamed a 'disjointed, siloed, unresponsive and defensive' system. This without a doubt applies to lupus patients in Wales.

FTWW wants to see a change in mind-set which often sees these patients dismissed or misdiagnosed and languishing without recognition or adequate care, something that our research seems to disproportionately affect women and has been highlighted by the Covid19 pandemic, not least in the Welsh Government-commissioned report, 'Locked Out...' (3). Medically-speaking, the extreme immune response to COVID-19 is the same as an extreme lupus flare; the cytokine* storm is the same immune system reaction as is seen in lupus patients with severe disease activity.

Data clearly demonstrates how black and ethnic minority people are at a higher risk of serious complications, both from Covid-19 and Lupus (4) with low levels of vitamin D as factor in common (5). If any good is to come out of the pandemic, it is that the medical world

will have a much better understanding of the human autoimmune response to viruses and, consequently, improve understanding of lupus and other RAIRDs too.

*Cytokine Storm, where the body starts to attack its own cells and tissues rather than fighting off the virus.

The Aims of this Report are to

- **Highlight the current situation facing people living with lupus and rare auto-immune conditions in Wales**
- **Ensure that patient testimonies and experiences are central to the co-production of future strategy and improvements in care**
- **Make a number of evidence-based recommendations which are in line with existing and developing policy in Wales and the UK to improve patient outcomes**
- **Address the gender health inequalities which underpin inadequate service delivery in Wales.**
- **Apply an intersectional focus to the experiences of Lupus and other RAIRD patients, not least by referencing the Black Lives Matter movement.**
- **Consider patient experiences in the context of COVID-19.**

Part Two: Definitions

2.1 What are Rare Auto-Immune Rheumatic Diseases / RAIRDs?

Rare autoimmune rheumatic diseases (RAIRDs) can be split in two groups: connective tissue disorders (any disease that affects the parts of the body that connect the structures of the body together) and systemic vasculitis (inflammation of the blood vessels). RAIRDs develop when the body's own immune system starts to attack healthy parts of the body, leading to inflammation and damage in tissues or organs, which is often irreversible, and can be fatal. These conditions can affect many parts of the body (including joints, skin, lungs, kidneys and heart) and therefore often require similar cross-specialty medical expertise.

For many people living with these diseases, the effects may not be outwardly apparent, including nerve damage, kidney disease, and chronic pain. Many suffer from severe fatigue (as a result of both disease and treatment), and their health can vary from day to day making it difficult to meet the demands of a job and employer. Similarly, their personal and social life can be devastated, resulting in breakdown of relationships.

Unlike the vast majority of rare diseases, these conditions predominantly occur in adult life and do not have a simple genetic link.

According to RAIRDA around 9,000 people in total in the UK will be diagnosed with vasculitis (including Behcet's), scleroderma, lupus or primary Sjögren's Syndrome every year.

2.2 What is Lupus?

Lupus is short for Systemic Lupus Erythematosus. There are other versions of lupus too, for example Discoid Lupus, Sub-Acute Cutaneous Lupus Erythematosus, Drug Induced Lupus and Neo-Natal Lupus.

Lupus is an incurable immune system illness, probably genetic in origin and mainly suffered by females. Only one in ten sufferers are male. The body's own defences can attack almost any organ or bodily system, and therein lies the danger.

Worldwide, lupus is acknowledged as being more common than leukaemia, muscular dystrophy, and multiple sclerosis, but those conditions (and others) share a number of similar symptoms.

Lupus can produce many symptoms and general practitioners understandably often fail to recognise it. In its early stages, the condition is often misdiagnosed and mistaken for other chronic conditions (such as Multiple Sclerosis, Rheumatoid Arthritis, Rosacea and other skin conditions, Fibromyalgia or anaemia) Meanwhile, a number of major organs can be irreversibly damaged, principally the kidneys and the skin, but also the heart, lungs, and brain.

The most commonly reported symptoms are joint and muscle pain, and an extreme tiredness that will not go away no matter how much the person rests. Alongside this, rashes, depression, anaemia, feverishness, headaches, possible hair loss and mouth ulcers may all feature. Miscarriage, often recurrent, is another unhappy complication of lupus.

Notably, whilst pain and fatigue are invariably present, people with lupus can differ greatly in their symptoms and how the illness can affect them – it can be life-threatening for a few, very mild for some.

The main trigger for lupus is hormonal activity and change, and lupus can often develop after childbirth, at the menopause or during puberty and usually between the ages of 15 and 55.

Although life expectancy has vastly improved in the past 50 years, lupus remains a potentially life shortening and life-threatening disease in more severe cases, and the majority of patients experience life-changing symptoms and reduced quality of life.

The NICE-approved BSR Guidelines for the Management of Adults with SLE states, 'The disease is prone to relapses and remissions, resulting in considerable morbidity due to flares of disease activity and accumulated damage, and an increased risk of premature death, mostly due to infection or cardiovascular disease...About one-third of SLE patients in the UK develop Lupus Nephritis*' (6).

*Lupus Nephritis, lupus causing inflammation of the kidneys

Clearly, the key point of which to be aware is that lupus is not simply a musculoskeletal condition that effects only people's joints.

In 2019, Professor Caroline Gordon, Professor of Rheumatology at University of Birmingham and Lead Author of the BSR Guideline for the Management of Systemic Lupus Erythematosus in Adults, wrote a piece for the Chief Medical Officer for Wales's quarterly newsletter, where she emphasised the need for early diagnosis and expert treatment and monitoring:

'Patients with lupus nephritis died at a mean age of 40.3 years with an average of 7.5 years between the diagnosis of lupus nephritis and death...Most lupus patients die from infection or cardiovascular disease rather than active lupus disease in the UK. During life, lupus causes considerable morbidity as it may present with a variety of clinical symptoms and signs that progress slowly or in some cases rapidly, causing the accumulation of chronic damage if not promptly diagnosed, appropriately treated, and regularly monitored' (7).

Despite this powerful testimony and clear guidance, Lupus patients in Wales still report failures in diagnosis, treatment, and ongoing management.

Part Three: Wider Health Implications for Lupus Patients

As discussed, lupus is a systemic condition. This means it can have wide and varied impacts on different organs and their functionality as well as being linked with the development of other diseases.

There is a risk of organ damage being accrued while the patient is completely unaware. For example, renal damage is one of the most serious complications of SLE with up to 60% of adults with SLE developing renal abnormalities over the course of their disease. Additionally, osteonecrosis (loss of blood flow to the bones, causing eventual break-down and death of bone tissue) may be asymptomatic in patients with Lupus. Similarly, cardiovascular disease may also go un-noticed, although evidence shows that the prevalence of plaques in internal carotid arteries (fatty deposits clogging the blood vessels that deliver blood to the brain and head) is three times higher than in the regular population (8).

There are a number of co-morbidities which need to be considered when discussing Lupus patients:

- **Diabetes**

Diabetes has been considered an important cause of heart attacks and strokes for a long time. While systemic lupus erythematosus (SLE) has been noted by experts to also cause heart attacks and strokes, it is generally not referenced in public information or even scientific leaders' lists of causative factors which currently include diabetes, high blood pressure, and high cholesterol. This is despite evidence from a 2020 study (9) which showed that Lupus patients had a 27% higher chance of having heart attacks and strokes than diabetics.

Heart attacks and strokes (i.e. cardiovascular disease) are the #1 cause of death in SLE patients, and these problems occur much earlier in life than they should. It is time for SLE to be added to the list of diseases that are most likely to increase the risk for having heart attacks and strokes, not least so both patients and their healthcare providers are aware and able to monitor individuals' health accordingly.

- **Cardiovascular Disease**

In 2019, the British Heart Foundation published its ground-breaking report, *Bias and Biology – How the gender gap in heart disease is costing women's lives* (10). The report provided alarming data on the number of women dying needlessly from heart attacks in the UK, or not making as good a recovery as they could, as a result of not receiving the same care and treatment as men.

The report found that women having a heart attack delay seeking medical help longer than men because they don't recognise the symptoms; that a woman is 50% more likely than a man to receive the wrong initial diagnosis for a heart attack; that women are less likely than men to receive a number of potentially life-saving treatments in a timely way, and that, following a heart-attack, women are less likely to be prescribed medications to help prevent a second heart attack.

Lupus patients are at 7- to 10-fold increased risk of developing cardio-vascular disease than the general population, whilst women with SLE have been found to be up to 50 times more at risk of developing heart disease (11). Given that 9 out of 10 lupus patients are women, gender bias against women receiving the correct diagnosis and treatment for a heart attack has a significant cumulative impact on this population's prognosis.

- **Kidney Disease**

Renal involvement is a major cause for morbidity and mortality in lupus patients. Renal disease is clinically silent and must be actively investigated and intervention started promptly to prevent renal damage.

One third of Lupus patients in the UK develop Lupus Nephritis. It is also well documented that half of all lupus patients will experience kidney involvement at some point in their lives.

A patient's blood tests can show normal kidney function, but they can still have inflamed kidneys (glomerulonephritis). Urine must be checked as it can provide further information by way of evidence of protein or blood. If detected, further tests can be performed. Kidney damage can be reversed if detected quickly enough. Early intervention is the key.

- **Neurological issues**

Lupus can present with a range of neurological issues, including migraine / headaches, brain fog, dizziness, balance issues, nausea, memory problems, lack of concentration, impatience, and other central nervous system symptoms. Too often, these issues are not referred on to Neurology services.

Neurological symptoms can be life-changing, very frightening, and have a pronounced impact on employment and well-being. Many lupus patients report feeling that they have been left to deal with symptoms on their own, which can be very distressing and result in a much-reduced quality of life.

- **Gastro-intestinal and hepatic features**

These issues commonly occur in lupus patients and are often not recognised as being due to lupus. Many lupus patients report having had gastro-intestinal issues and hepatic issues that have been ignored, or not connected to their lupus/auto-immune illness. Such symptoms should also be monitored, and an appropriate referral made if the patient is not responding to lupus medication. Stomach protectors for patients taking medications are often not addressed as quickly as they should be.

- **Ophthalmic manifestations**

These are rare but potentially sight-threatening and need to be carefully evaluated by an experienced Ophthalmologist. Many patients with RAIRDs will be taking Hydroxychloroquine, an anti-malarial medication which works by decreasing the activity of the immune system for their condition. The Royal College of Ophthalmologists deals with this in their Clinical Guidelines for Hydroxychloroquine (12) stating that detailed retinal screening is required for such patients. However, there are concerns that Wales does not have sufficient personnel to provide this service.

At the start of 2020, Hywel Dda University Health Board was advertising for no fewer than 6 Consultant Ophthalmologists, resulting in delays to receiving this essential service. With the additional backlog caused by COVID-19, patients are understandably anxious about damage to their sight, which can again be silent for some time. One Lupus patient reported taking Hydroxychloroquine for over twenty years and never yet having seen a Consultant Ophthalmologist.

- **Hypermobility**

A number of people with lupus have joint hypermobility. This is when joints are extremely flexible. It can lead to problems such as joint pain and stiffness, repeatedly getting strains and sprains, poor balance or coordination, thin, stretchy skin and bladder or bowel problems. In rare cases it can mean dislocation of joints – when joints pop out of place. The effects of this condition are regularly underestimated, with many patients reporting having its impact dismissed. In more serious cases, specialist physiotherapy should be available to help with many of the physical symptoms it incurs, including long term mobility issues. Hypermobility as an additional diagnosis to lupus is not discussed with the majority of the lupus patients.

- **Dermatological issues**

Many patients have skin involvement with their lupus. However, a considerable number are either not referred or do not have regular appointments with a Dermatologist. In Centres of Excellence there are frequent combined Rheumatology and Dermatology appointments,

where lupus patients can be reviewed by a Dermatologist with the requisite experience, in the same room as their Rheumatologist / Lupus Expert. New lesions or changes to existing lesions can be assessed, and those patients on medications which may incur additional risk of skin cancer can be monitored. However, as it stands, many patients in Wales report never having had dermatology involvement in their care. For others, delays mean they will be waiting far longer than a year for their 'annual' skin cancer checks.

- **Pregnancy**

Patients planning or becoming pregnant need careful and regular monitoring, both because of the increased risk of miscarriage and also because of the link between hormone changes and disease activity. One patient (who has both Lupus and APS) reported going through an entire pregnancy with no monitoring appointments at all from Rheumatology, whilst another described only having one appointment during her whole pregnancy.

Part Four: Rationale for this Report

4.1 The Covid Context

The medical landscape for patients in Wales with RAIRDs has been fraught during the pandemic, highlighting and exacerbating pre-existing problems in accessing specialist advice and information specific to these conditions.

In the first instance, many patients were unsure of their shielding status, having not received letters from Wales's Chief Medical Officer when they felt they met the criteria. In seeking clarification, patients were sent from hospital-run rheumatology helplines to the GP and back again, with no clear resolution.

During this period, the RAIRDA collated a report, *The Impact of COVID-19 on people with rare autoimmune rheumatic diseases* (13) which demonstrated that:

- 54% of respondents were currently shielding.
- 80% had experienced a change in their care and treatment.
- 37% said that their ability to manage their condition had been affected as a result.
- 33% said that the pandemic had adversely impacted their household finances compared to 23% of the general public.
- 34% said that advice from the health service or the government was "unclear and contradictory".
- Some raised concerns about accessing healthcare due to not wanting to add to the burden on the NHS or fears of infection with COVID-19. However, many more commented that they felt 'abandoned' due to a lack of clear information about their level of risk or the way their treatment plan had been changed.

When COVID-19 eventually recedes as a public health concern, RAIRDs will still be here. The testimonies of people with RAIRDs in this report and throughout FTWW's engagement with

patients during and beyond the pandemic clearly demonstrate the need to improve communication and care for this population.

4.2 Policies Relating to Lupus Patient Care in Wales

Research conducted with lupus patients across Wales reveals that a significant number feel that the NHS system here is failing them. Many gave accounts of negative experiences and barriers to accessing care, with symptoms repeatedly dismissed and treatment inadequate or unavailable. A large proportion of them described feeling hopeless, abandoned, and worthless. For information on the wider policy landscape and its implications for patients in Wales, see Appendix 2.

- **National Guidelines**

A number of patients' experiences exemplified a lack of compliance with the BSR Guidelines including not having Rheumatology reviews often enough; baseline tests not being conducted; urine samples not being taken or sometimes disposed of without checking; not having a Dermatologist referral or any sort of multi-disciplinary team approach; not seeing an Ophthalmologist for eye checks, and vitamin D levels not being monitored, or deficiency addressed. Meanwhile, the *Rheumatology in Wales: State of Play* 2016 report (14, see Appendix 3 for more detail) confirms that there is significant variation in terms of meeting approved guidelines across Wales.

- **Accessing Second Opinions**

Currently, in Wales, lupus is not always considered serious or complex enough to warrant access to specialist expertise, particularly in a tertiary setting. It is felt that a general rheumatology department can provide adequate care. NHS Wales policy does not guarantee access to an independent second opinion or an appointment with the patient's chosen clinician. If a second opinion is permitted, it is usually from a general Rheumatologist within the same Health Board and department as the original consultant. This policy results in a clear conflict of interest for the clinician and a lack of independent expertise available to the patient.

- **The Individual Patient Funding Request**

For those patients who want to see a specialist in their condition outside of their health board, NHS Wales operates an Individual Patient Funding Request (IPFR) system. However, based on findings from a series of Freedom of Information Requests submitted to the seven health boards in Wales, it is clear that very few lupus patients attempt this mechanism. This is likely because it is the local consultant who is required to complete the IPFR rather than the GP or patient themselves.

- **Tertiary Care**

Wales's population is relatively small, with only around 3 million residents. Healthcare is a devolved matter, with funding for services being proportionate to that population. The

services that are available are the responsibility of each individual health board, with health conditions prioritised according to local need.

Where RAIRDs are concerned therefore, it would be unrealistic to expect every health board to provide a tertiary Centre of Excellence for what will be a relatively small number of people. However, as it stands, the absence of local provision is proving to be an insurmountable and unnecessary barrier for patients needing to access specialist care, demonstrating the inflexible nature of policies which are not patient-centred.

Prior to the pandemic, children with Juvenile Lupus were referred to Centres of Excellence in England as there was no Paediatric Rheumatology Service in Wales. There is now a service provided in Cardiff. However, the situation for adult patients seeking access to tertiary-level care remains fraught with difficulty as will be explored later in this report.

- **Participation in Research**

Whilst there are no Centres of Excellence for Lupus in Wales, the Royal Glamorgan Hospital within the auspices of Cwm Taf Morgannwg University Health Board, is a BILAG Centre, (British Isles Lupus Assessment Group). This is a way of administering biological treatments to lupus patients and adding them onto the BILAG Biologics Register, an academic study looking at the safety of biologics in lupus patients. However, being on the BILAG BR register is not equivalent to the standards operating in a Centre of Excellence. No other hospitals in Wales are on the BILAG BR register.

With regards to funding applications made to Lupus UK for research, BILAG has informed the authors of this report that they have no knowledge of any research into lupus or rare auto-immune diseases taking place in Wales.

4.3 Diagnostic Delay

Prompt diagnosis is a major issue for all RAIRDs. Many patients report being dismissed or misdiagnosed for several years, the average time to diagnosis being around 6.4 years (15). The BSR Guidelines make clear that both initial diagnosis and ongoing assessment of patients are dependent on taking a careful history and examination. Blood tests are only one part of a range of investigations necessary to establish the degree of disease activity, accumulation of chronic damage, and identify other complications or co-morbid conditions that will influence the patient's treatment plan. Far too many patients report having blood tests which, when eventually facilitated, are interpreted erroneously, or used – wrongly – as a definitive diagnostic tool when, for several patients, they may be inconclusive, especially if looked at in isolation.

Assuming a patient is referred for an appointment with a general rheumatologist, fibromyalgia, traditionally perceived within the medical establishment as a condition with no physiological cause will often be determined. Under current protocols, this sees most patients discharged from secondary care and sent back to primary care for GP management, leaving many without access to adequate testing, treatment, or monitoring. However, there

is a growing body of evidence positing fibromyalgia as an autoimmune condition, either a 'new' addition to the spectrum of auto-immune diseases or simply one of the many autoimmune diseases that has gone undiagnosed for several years, lupus amongst them (16).

The American College of Rheumatology Classification Criteria for SLE makes clear that a patient may be diagnosed with lupus if they have 4 or more of 11 criteria present (17; see Appendix 4 for a list of criteria). However not all patients who meet four of the criteria have a lupus diagnosis. This may be because the rheumatologist is inexperienced with lupus and worried about getting the diagnosis wrong. The symptoms for these criteria may appear and disappear over time, making it even trickier for non-experts. This complexity makes it all the more important that specialist input is sought early on in the diagnostic process, something that continues to be an issue for many patients in Wales, for whom accessing a rheumatologist specialising in RAIRDs, can be particularly problematic.

When patients attend an appointment in General Rheumatology, they will likely have waited an extended period and symptoms may have subsided. The lack of understanding of the typical 'ebb and flow' of Lupus results in many patients feeling disbelieved and their diagnosis questioned. The impact on mental health caused by this experience must not be under-estimated. One 2020 Study of Lupus Patients' Experiences states that, 'although many patients have eventually built secure, trusting medical relationships with individual physicians, distrust in the wider medical profession often remains. (This) can be improved by physicians...being more aware of the ongoing impact of persisting psychological damage from previous misdiagnoses' (18).

Both the NICE BSR Guidelines and RAIRDA's 2018 report, **Reduce, Improve, Empower** (19) emphasise the importance of making the earliest possible diagnosis, so that the correct treatment can be administered. The evidence clearly indicates that referral to a lupus expert at the diagnostic stage would both expedite diagnosis and treatment and reduce the impact of any long term, permanent physiological damage.

On the topic of diagnostic delay, patients in Wales said:

I have not formally been diagnosed with Lupus. I believe I have all the symptoms except the bloods. I have Anti Phospholipid Syndrome as well as Sjogren's Syndrome... My kidneys are also not performing as they should. I can go from morning time to teatime without going to the toilet, despite drinking plenty. I often get blood in my urine which is visible. I get no restorative sleep and haven't had for over 10 years. I was discharged from my work due my health aged 54...If I try to do much, I get breathless and have to stop. My condition basically rules my life. I cannot plan anything because of the unknown quantity that is my health. I asked to be referred but was told there is sufficient expertise within Wales. I was diagnosed with Ankylosing Spondylitis at St Thomas' in London but that was refuted once in Wales and my Humira that was prescribed for it was taken away. I have suffered as a result of the poor care I have received which makes me disappointed, frustrated and

angry.

I was diagnosed with SCL in November 2013, but I had my doubts that this was the correct diagnosis from the very beginning. In my mind I met the diagnostic criteria for a Systemic Lupus / SLE diagnosis, having read and researched this endlessly. I was denied a second opinion on my SCL diagnosis from a lupus expert, via the NHS, on many occasions.

The first symptoms of lupus that I had such as skin rashes, nose bleedings, tiredness and anaemia despite diet, started when I was 7-8 years old. These continued during childhood. When I was 14 years old, I started having joint and muscle pain which got me a lupus diagnosis. At first doctors didn't know if it was lupus or connective tissue disease. Years after, it was clear that it was lupus.

I am originally from Cardiff, where it took many years for me to be diagnosed with Sjogren's Syndrome and eventually Lupus, then Nephrotic Syndrome. I believe I also have several other undiagnosed autoimmune / connective tissue / hypermobility conditions, but this is never discussed or explained.

4.4 Lack of Specialised Lupus Care in local Rheumatology services

With current waiting lists for new patients to general rheumatology in Wales reported to be somewhere between a year and two years long and the expectation that lupus can be managed locally, it is very rare for a lupus patient in Wales to access a second opinion regarding either a diagnosis or a treatment plan from a lupus expert, either within Wales, or a Centre of Excellence in England. As this report came to publication, the number of such centres in England has increased to ten. There are none in Wales.

If a Rheumatologist has agreed to a referral to a Centre of Excellence in England, it is usually in very limited circumstances, firstly where there is kidney involvement and/or where the biological infusion, Rituximab, is being considered. Unfortunately, this is often extremely late in the patient journey, when disease may be too advanced to prevent long-term damage.

As this report was collated, no Rheumatologists from mid & South Wales have made enquiries with Lupus UK expressing an interest in setting up a Centre of Excellence in Wales. There is a single recognised lupus expert working in South Wales and one in North Wales, both of whom are recognised by Lupus UK as Lupus Specialists (for more information, see Appendix 5).

Unfortunately, patients outside of the specific health boards in which the two experts are located and who have requested a referral have had their requests rejected. There have, however, been three enquiries made to the charity regarding the possibility of applying for five years of funding for a Lupus Specialist Nurse (see Appendix 6 for more information on this role). To date, no actual applications have been submitted.

In 2018, Lupus UK offered to fund a Specialist Lupus Nurse to assist the Rheumatology Department in Hywel Dda UHB, at a cost of £250,000 over five years but, unfortunately, this offer was rejected. At the time, the health board gave the service's lack of capacity to develop and fully support the addition of a Lupus Specialist Nurse to the rheumatology team as the reason. This is surprising, considering Lupus UK's findings which show that all of the charity's UK-funded Specialist Lupus Nurses are subsequently adopted by their Trusts after the initial five years of funding expires. Evidence shows that they can recoup their employers this initial outlay and more, as well as play an integral role in improving patient health and well-being.

On speaking to patients in various parts of Wales, it has become apparent that there is little understanding of lupus / connective tissue disease within some local Health Board(s) and therefore no understanding of how an expert opinion, treatment plan, and monitoring have the potential to dramatically improve a lupus patient's quality of life or increase their life expectancy. It seems that Lupus is often miscategorised as a musculoskeletal condition, like osteoarthritis, and this should not be the case. Lupus does not affect only the patient's bones; it is a systemic illness that can affect any organ.

Another issue with not being able to access RAIRD specialists is an apparent hesitancy in appropriately managing patients medically. Several Lupus patients have reported their general rheumatologist being reluctant to increase medication levels, add new drugs into their treatment plan or combine certain medications. When patients have sought private expert opinions, they have been told that they are under-medicated. This resistance can potentially be attributed to a lack of knowledge / experience / confidence with lupus and the drugs required. The problem is that if lupus is left under-medicated, it smoulders away and can cause irreversible damage (20).

Not only is medication proving to be problematic, patients also report a lack of appropriate monitoring, with the important role of urine analysis, vitamin D levels, and DEXA scans not fully appreciated or utilised in their disease management. Tests do not appear to be repeated at sufficiently regular intervals which makes it more difficult to keep a close eye on disease activity and intervene if organ damage is being sustained.

Despite the best of intentions, Rheumatology Helplines in Wales do not always work as effectively as they should. Patients report that telephone calls are not always returned. For Lupus patients, who can experience severe and unexpected disease flares, advice from the Helpline is an essential tool in enabling self-management, including coping with symptoms, ability to maintain employment, and carry out daily tasks. It is most important that the Helplines are adequately resourced, staffed, and in possession of relevant information for those living with RAIRDs.

On the topic of Lack of Specialised Lupus Care in local Rheumatology services, patients in Wales said:

There are a few of us with lupus who have attended the London Lupus Centre privately for help and some who have attended a Sjogren's expert in Swindon privately too...Lupus

patients with serious organ involvement, kidneys, lungs, or liver...Some (are) so desperate for help they've taken out loans, used credit cards or in the worst case, taken out a pay day loan to access a specialist lupus doctor.

I do have monitoring appointments in Cardiff, but I am not confident with that care. I constantly have very high proteinuria and previously had kidney involvement with my lupus, but despite this, when I asked my Consultant Rheumatologist if I could be referred to a Nephrologist, he refused and told me that it is not necessary. I STRONGLY disagree. This is why I am not confident in his care. If he understood lupus, he would have referred me to a Consultant Nephrologist a long time ago. Ignoring these signs frightens me very much.

I feel that if I'd been listened and the appropriate action taken, i.e. scans, medication, monitoring, proper regular follow ups, looking at the whole picture not a specific area, connecting the symptoms, I wouldn't have had two heart attacks...or end(ed) up on the kidney transplant list, I firmly believe these things could have been prevented or and least delayed by many years, saving the NHS thousands upon thousands of pounds, never mind the emotional and physical cost to myself and my family.

About 2 years ago, I underwent surgery to fit a cannula into my arm to be used for dialysis. I have never started dialysis. My son said he wanted to donate me a kidney. We went through everything that was asked of us...At the last minute (they) told us that (he) could not donate, his BMI was too high...He is as fit as a fiddle...Am I having a transplant? Or am I having dialysis? I need to know. I feel like I'm being left to die. No one has been in touch with me to tell me what the plan of action is for me.

Despite my kidney function being about 9% currently, due to my lupus nephritis, this new doctor told me that my lupus is not an issue for me, at my age, as it's most likely inactive now. He told me that post-menopausal women do not have active lupus...How can my lupus be inactive? I am covered in a rash and my kidney function is 9%. My lupus is active. My age doesn't matter.

My health was declining rapidly, my lupus was in a massive flare for months. Medication clearly wasn't working well, but despite my pleas every time I saw the nurse my suffering was dismissed...I ended up in a wheelchair due to the horrible pain of swollen red-hot joints. I had locked joints and muscle pain so badly I cried when I had to make few steps to walk to the loo. My fever was consistent. My butterfly rash came back with vengeance...My mother called the ambulance and they rushed me to University Hospital of Wales. My organs were shutting down. I had heart, lung, and kidney failures. All which were overlooked by my rheumatology care. Unfortunately, all the damage and scarring which lupus caused my kidneys during a whole year of rheumatology team not noticing my decline caused my kidney failure. Now I am awaiting a kidney transplant / dialysis. I truly believe that all this could have been avoided with proper care and treatment.

4.5 Lack of joined-up care

Lupus is a multi-system disease. However, patient testimonies suggest that referrals to other specialties in Wales, including Gastroenterology, Dermatology, Nephrology, Cardiology, Haematology, Immunology, Urology, Orthopaedics, and other relevant departments are either terribly slow or, worse, sometimes non-existent.

Patients report their local rheumatologists being reluctant to make onward referrals and advising that their GP must make them instead. This delays the process for patients and is not a good use of GP time. It also means that care is fragmented, with consultants working in silos and no overall plan for the patient's care and well-being.

The BSR Guidelines for the Management of Adults with SLE clearly state that a multi-disciplinary team approach should be taken, where 'management may involve a variety of specialists...working as part of collaborative clinical networks involving regional specialist centres, local hospitals and GPs' (21). As it stands, the responsibility to co-ordinate care often falls to the seriously ill patient, who is required to facilitate information-sharing between parties. This is made all the more challenging when patients in Wales will not automatically receive test results or consultants' letters and have limited access to their electronic patient records.

Communication between specialities and patients is particularly important when the patient is complex and has organ involvement or is taking a number of medicines, including Disease-Modifying Anti-Rheumatic Drugs (DMARDs). For Shared Care Agreements to be meaningful, the prescribing consultant (usually the rheumatologist) needs to be involved and advising both the GP and the patient on next steps.

As described in Section 3.2. the increased risk of premature death in Lupus patients is caused by infection or cardiovascular disease, as referenced in the BSR Guidelines. A recent study (22) confirmed that lupus patients have a 27% increased risk of heart attacks or strokes, compared to diabetic patients. However, patients report inadequate monitoring of cardiovascular issues. Most will have had the baseline tests undertaken at diagnosis; however, for a considerable number, tests do not seem to be repeated at any point.

Careful and consistent monitoring of cardiovascular risks and health in Wales's lupus patients could prevent these potentially catastrophic events and constitutes an essential part of patient-centred care. The same applies to those with kidney involvement which, for Lupus patients, can only be classified by kidney biopsy. This requires sufficiently specialised rheumatologists working alongside nephrologists to advise and establish joined-up monitoring and treatment plans.

Unfortunately, despite the serious implications of Lupus and clear patient need, the Welsh Renal Network has informed the author that lupus patients are not eligible for their services. This is particularly concerning as the network can provide invaluable practical and emotional support for patients. For Lupus patients in Wales, where there can be a real absence of guidance, it is a significant missing link.

A significant number of patients report that, even when they have gone to the trouble and expense of seeking an expert opinion, there is no guarantee that the diagnosis or treatment plan devised will be utilised when the patient returns to local service providers, even if the care offered locally doesn't comply with guidelines. For example, Lupus patients who have active disease should be seen every 1-3 months, more often if necessary. Those with mild and / or stable disease should be seen every 6-12 months. Even before the pandemic, this often was not the case.

Members of LUPUS UK in Wales who have been admitted to hospital with lupus-related issues report either waiting several days for a Rheumatologist to visit them, or, worse, being discharged after over a week as an inpatient without being seen by a rheumatologist at all. This can cause dis-jointed care and conflicting treatment plans.

The NHS in Wales is moving towards a system of patient-initiated, rather than automatic, follow-up which could have negative implications for those with on-going health needs, particularly those with chronic, complex conditions like Lupus. According to the BSR Guidelines, better outcomes are achieved if Lupus in-patients are overseen in multi-disciplinary centres with experience of managing the complex nature of the disease. It is unclear which hospitals in Wales have dedicated Rheumatology beds available, but this is an important consideration for complex patients going forward.

On the topic of lack of joined-up care, patients in Wales said:

I have never seen a Consultant Ophthalmologist for an eye-check, and I have been on hydroxychloroquine for 15 years now.

I need someone to take control of my case. I need a multi-disciplinary meeting, with rheumatology, nephrology, dermatology, and the transplant team. I've completely lost faith with everyone involved in my case. All these doctors and all these hospitals are not co-ordinating anything. It's getting too much for me to deal with and coordinate myself. I do not have the energy. I am so, so very tired of all of this and that scares me.

Lupus is a 'systemic' disease...consequently, the patients find themselves going to an assortment of hospitals, to see various Consultants, each specialising in a different 'speciality' e.g Rheumatology...Renal...Immunology...Gastro...and so on! Increasingly I detected...a lack of communication between all the doctors of a patient with Lupus...I was/am doing my own 'Patient's Admin'

Meanwhile...

Patient K was admitted to hospital and diagnosed with lupus. They were told that they had kidney involvement immediately. They were referred to a Centre of Excellence in England for an opinion and new treatment plan. The Lupus expert they saw told them a treatment plan could not be devised until the extent of the kidney involvement was confirmed via a kidney biopsy. A letter was sent to Patient K's rheumatologist in Wales informing them of this proposed course of action. The letter also advised that Patient K could have the biopsy at the Centre of Excellence, if that was the preferred option. Instead,

Patient K was referred to a local Nephrologist in Wales, who refused to perform the biopsy as, despite the Lupus expert's opinion, they did not consider it necessary. As a result, more than two years later, Patient K still has no specialist lupus treatment plan as no biopsy to assess kidney involvement and decide on a drug regimen has been performed. Patient K wonders why they were sent to a lupus expert out of area if the doctors at home had no intention of adhering to the specialist's opinion?

Patient L is actively advertising for a live donor, whilst Patient M, another lupus patient requiring a transplant, says that they feel they have been left to die.

Patient G was sent via an NHS referral to see a lupus expert at the Birmingham Centre of Excellence for Rituximab infusions. The lupus was severe and not being controlled. The lupus expert agreed Patient G should start Rituximab. However, locally, other treatment options were tried instead. By Christmas 2019, Patient G was bed-bound and covered top to toe with a lupus rash and had never had such a long or severe lupus flare before.

Patient H is 27 years old and has lupus, APS and Primary Sclerosing Cholangitis. Patient H suffered two strokes caused by APS but was told that treatment wasn't needed. Patient H has not seen a Rheumatologist since 2019. A Gastroenterologist has seen Patient H and written to their Rheumatologist on three or four occasions but has never received a reply. There is no multi-disciplinary team approach. The Gastroenterologist is now leaving and is concerned about who will look after Patient H. Patient H has been informed that their life expectancy is limited. Patient H is a single parent to two young boys.

4.6 Issues in general practice

For the most part, patients report that their GPs are doing their utmost to help, but that many have voiced feeling out of their depth and that lupus patient care should be led by rheumatology.

Unfortunately, a significant number of patients report having been discharged from rheumatology care, despite having a diagnosis of SLE or other Connective Tissue Diseases and taking strong medication including DMARDS / immune suppressants. This raises concerns about the standard of care provided, as the BSR Guidelines clearly state that, even if a patient's disease is considered stable and/or they have low disease activity, they should still be monitored by a Rheumatologist every 6 – 12 months. To do otherwise would be considered a breach of the patient's Shared Care Agreement.

Dialogue between Senedd Members and Health Boards suggests that the main justification for not following these Guidelines in Wales is, simply, a lack of resources.

On the topic of Issues in General Practice, patients in Wales said:

I had some fatigue and neck pain and stiffness in the morning. I didn't pay much attention, thinking it was posture, sleeping position or stress. A few months later, my joints started hurting a lot. Then they started swelling up really bad. I got a mild fever. My GP ordered some tests. I was anaemic and had raised inflammation levels, also low

vitamin D. My GP also noted a strange rash, which I hadn't noticed myself and that's when she suggested for the first time that it could be lupus. By that time, I was in so much pain I could hardly walk. I was crawling, using crutches. It took 4 trips to A&E via ambulance when I couldn't walk, eat or drink for many days as I would bring everything up. By that time, I was in renal failure and multi organ damage due to massive inflammation in my body.

As Lupus is thought to be linked to female hormones, a Rheumatology Consultant predicted that as hormones lessened over years, hopefully, my many systemic Lupus symptoms...would gradually lessen. This did not happen to me.

4.7 Barriers to tertiary care

In Wales, all health boards offer a General Rheumatology service. Most will not have a Lupus or RAIRD expert. There still tends to be the perception that Lupus is purely a musculoskeletal disease, for which specialist care is unnecessary. As a consequence, accessing a Lupus / RAIRD expert 'out of area' is not automatically – or straightforwardly - facilitated.

Patients report feeling as though their Lupus is not considered serious enough to warrant expert input. This approach in Wales is in opposition to the direction of travel taken by NHS England. In line with the NICE-approved BSR Guidelines and LUPUS UK's recommendations, NHSE continues to invest in increasing provision of Centres of Excellence in key locations, where multi-disciplinary expertise can be accessed in one place at one time, whilst Lupus Specialist Nurses provide care closer to home to maintain local monitoring and support.

Some patients report their being denied a referral to a Centre of Excellence due to a misconception that the hospitals offering these Centres are 'not NHS'. Despite LUPUS UK correcting this factual inaccuracy and providing an up-to-date list of the NHS Centres of Excellence, rejections have not been reconsidered to date.

It is important to note that when Wales became devolved, the Health Minister at the time, Mark Drakeford, gave clear confirmation that this should not be a barrier to patients accessing the expert care they needed over the border (in England) (23). Despite those assurances, this is indeed what has happened to many patients, with Lupus no exception.

As discussed in Section 4.2, where specialist care is unavailable locally and out-of-area referrals to tertiary care not part of a pre-existing care pathway, an Individual Patient Funding Request may be utilised if the local consultant is willing to facilitate one. This can be problematic for a number of reasons, not least that many Lupus patients are under the care of a GP only. Unfortunately, a GP's signature is considered insufficient by IPFR decision-making panels.

Furthermore, Welsh Government and Health Board policy is very much focused on the delivery of 'care closer to home', where monies – and patients – remain within the local area. Whilst, generally-speaking, this approach may be more convenient for the patient in terms of reducing travel costs and time, it does not necessarily ensure best care for those

patients with more complex needs and / or 'rarer' conditions which would be better-served by accessing a specialist.

Every Lupus patient who responded to the call for evidence for this report said that they would prefer to travel to see a Lupus Expert at a Centre of Excellence and have the resulting treatment plan monitored locally. For them, this approach would be most helpful in maintaining a satisfactory quality of life, remaining in employment, and playing a fuller role in their family life and communities.

A UK-wide study of patient experiences of Lupus and related systemic autoimmune diseases from 2020 concluded that, 'participants in Wales had the poorest perception of overall medical support. Great concerns over local barriers to accessing specialist care were detailed by those living in Wales' (24). For a nation which prides itself on listening to patients and has legislation which enshrines wellbeing and co-production, this is a damning indictment on Wales's NHS policy and its impact on those patients living with chronic illness.

On the Topic of Barriers to Tertiary Care, Patients in Wales said:

Current lupus care in Wales, by my experiences, is poor. The IPFR system is flawed. I would prefer to travel to see a lupus expert at a Centre of Excellence, as from my care in Leeds, I know proper care and management of lupus gives a better quality of life and lessens the accumulative damage to your body. Currently, my care leaves me feeling scared and quite daunted.

Lupus care in Wales has failed me miserably and I want to see radical changes to ensure Lupus patients are treated properly from here on in. The IPFR system does not work for us. The care is not good enough. We deserve better. The massive effect this chronic illness has on us needs to be officially acknowledged and NHS Wales and the system need to know that we are willing to travel further afield for better care and treatment, for a better quality of life.

I asked to be referred for a second opinion via the NHS in Wales and was told that it would never be allowed, so there was no point in even completing the paperwork. So, these initial denials were verbal and no actual IPFR paperwork was completed or submitted.

PART FIVE: Service Comparison and Centres of Excellence

5.1 Medical management of RAIRD and Lupus Patients

The research underpinning this report suggests that biological medicines and infusions are not generally available to lupus patients in Wales, whilst in other parts of the UK, LUPUS UK confirms that infusions are being widely used. Even more frustratingly, from enquiries made, infusions of biological treatments are widely used locally for Rheumatoid Arthritis, Psoriatic Arthritis, Colitis and Ankylosing Spondylitis.

There are Lupus patients in Wales for whom DMARDS have not worked but who are not offered biological treatments, despite the possibility of their giving them an improved

quality of life. Whether this is a financial decision, with limited resources directed towards the higher number of patients with Rheumatoid Arthritis, or simply down to a lack of expertise in managing Lupus is unclear, but it is a situation which needs addressing urgently.

Covid-19 has served to further highlight the discrepancies between the different National Health Services' approach to managing Lupus patients. Those in England who were receiving Belimumab by infusion, were fast tracked to a self-inject subcutaneous version due to doctors wanting to reduce the lupus patient's risk of Covid-19 exposure at hospitals. However, the report author is unaware of any patients in Wales who have been offered the option of self-inject subcutaneous Belimumab treatment.

5.2 Centres of Excellence

The LUPUS UK Centres of Excellence award scheme is designed to recognise and reward specialist Lupus Clinics that offer a high quality of care for people with lupus and to ensure that these standards are maintained. There are various criteria to obtain this status, including:

- A dedicated Lupus clinic with three or more consultants covering specialities e.g. Rheumatology / Nephrology / Dermatology that are available to see patients, though not necessarily all on duty at the same time
- At least one consultant with specialist knowledge of Lupus, with others having a good 'working knowledge' of Lupus
- A dedicated specialist Lupus nurse with other nursing staff having 'working knowledge' of Lupus.

These Centres of Excellence have been shown to improve patient outcomes, and rheumatologists at the Centres take great pride in achieving the accolade. There is no doubt that the patient care offered therein, and the research undertaken by personnel, has helped improve Lupus patient care in general.

Currently, there are ten LUPUS UK Centres of Excellence, all of which are in England (see Appendix 7) and, under NHS England policy, a patient has the right to be seen there if this is their preference. Many patients in England will only access lupus experts when required, with the patient's local rheumatologist helping to implement the lupus expert's treatment plan and monitoring the patient between consultant appointments, a team approach recommended in the BSR Guidelines.

The key word is choice. Not everyone in England with lupus asks to be seen by a lupus expert or at a Centre of Excellence. They might have mild, well controlled lupus or they may be happy with their local rheumatologist's knowledge and care. Perhaps they do not – or cannot - travel further. Whatever their reason, they still have choice, something that patients in Wales are denied.

PART SIX: Concluding Remarks

A series of Freedom of Information requests made by the report author reveals that there is considerable variation and inconsistency in what data is being collected, and how Welsh Government's policies are applied by individual health boards. There seems little oversight or accountability to address these variations.

The extensive research underpinning this report has revealed that some clinicians and health boards use the IPFR application process, whilst others say that this is not relevant, and a prior authorisation process can be used. Either way, it is unrealistic and unfair to expect seriously ill patients to navigate their way through such an administrative minefield, where there is no consistency in the use of IPFRs or even in the application of the BSR Guidelines.

What is clear is that expert opinions from lupus experts and / or LUPUS UK Centres of Excellence are not being obtained often enough, or when they are, early enough. Lupus patients need earlier intervention to achieve a better outcome. The options for treatment and medication offered to lupus patients in Wales are being significantly hampered by this lack of dialogue with specialists.

In the 1950s, most people with an SLE diagnosis died within five years. Thankfully, things have progressed for lupus patients and lupus is no longer considered a terminal illness. It can however be life-shortening and is often life-limiting. Quicker diagnosis and treatment can mean that a lupus patient is able to lead a more normal life, hold down their chosen career, play an active role in their community, and maintain their personal relationships, all of which can be damaged by living with lupus. Dedicated support for these patients, to enable them to maintain the best quality of life possible, is vital.

On the topic of Quality of Life with Lupus, Patients in Wales said:

I currently work from home. but working time has been reduced, and there has to be plenty of flexibility. It's tough. I have a lupus 'glass ceiling'. I know that even if I'm not flaring, I am physically limited due to some amount of pain and tiredness...Every day I have to choose some activities that are priority, and that's all I can do.

The time when I'm feeling relatively well is becoming scarcer. Healthy diet, good regular rest and relaxing are things that help but they are not a cure. Treatment is essential. Treatment has not only saved my life in multiple occasions, but it has allowed me "to be"...Lupus has affected every part of my life. My work, my income, my hobbies, my family life.

PART SEVEN: Recommendations

- Listen to and work with patients / patient advocates on implementing the following recommendations, in line with the principles of co-production enshrined in Welsh legislation and policy, not least the Wellbeing for Future Generations Act. Where Lupus specifically is concerned, Lupus UK should be a recognised third

sector stakeholder in future healthcare discussion and planning in Wales regarding lupus patient care.

- **As per the ‘Rheumatology in Wales: State of Play’ report, local Rheumatology Departments should hold more information and leaflets on RAIRDs, including Lupus. Further, signposting to dedicated support groups is warranted due to the important role they play in improving patient well-being. Evidence shows that patients who attend support groups are better informed about their condition and feel less isolated.**
- **Commit to building a strong working relationship between relevant charities, including Lupus UK and FTWW, Senedd Members, and Welsh Government.**
- **Welsh Government should write to all Health Boards to ensure they understand that they are expected to follow the NICE-approved BSR Guidelines for the Management of Adults with SLE. This includes the recommendation that lupus patients should have access to both a local rheumatologist and a Centre of Excellence from the start of the diagnostic process to ensure continuous and expert management of their condition and avoid long-term damage to physical and mental health.**
- **Advise health boards in Wales that, if a patient is admitted to hospital with Lupus-related health issues, they should be seen by a rheumatologist during their stay to review and adjust their treatment plan before discharge.**
- **Ensure that existing Quality Statements for those major health conditions relevant to Lupus patients, including Diabetes and Heart Conditions, refer to SLE and that this information is available to all diabetes and cardiology nurses / practitioners in Wales.**
- **Consider creating a RAIRD Implementation Group in Wales to co-produce a Quality Statement and Delivery Plan specifically for Lupus and other RAIRDs which would encompass compliance with existing and emerging guidance on the diagnosis and management of Lupus / Mixed Connective Tissue Disease / Undifferentiated Connective Tissue Disease / Rare Auto-Immune Rheumatic Disease across Wales. Clear pathways, with timelines, in the vein of Wales’s Cancer Pathway would be the recommended way forward and should include auditable Patient Reported Outcome Measures.**
- **Implement improved mechanisms for data collection on incidence of RAIRDs, including Lupus**

- **Consult with patients and established lupus / RAIRD specialists on how the principles of prevention and early intervention can and should apply for these disease areas, including those who are suspected of having kidney involvement. Early and expert intervention can prevent organ damage which incurs great personal and financial cost to patients and loved ones and makes economic sense for healthcare providers.**
- **Work with patients / patient advocates to map out what constitutes a multi-disciplinary team approach to the management of Lupus and other RAIRDS and advise various specialties of their potential role in diagnosis and treatment of patients affected, including specialised physiotherapy and co-design of self-management programmes. Additionally, consider the development and implementation of a 'red flag' system which highlights when interventions need to be initiated and / or expedited, for example in pregnancy.**
- **Ensure that all Lupus patients have both an Emergency Treatment Plan and a Formal Care Plan. This ensures that when patients experience flares of their disease, they and any / all treating clinicians understand the implications of Lupus and how best to manage it, even at those times (like weekends and public holidays) when the Rheumatology Helpline or GP is unavailable. These enable informed and shared decision-making and improve care.**
- **Consider making funding for specialist services for Lupus and other RAIRDS the responsibility of the Welsh Health Specialised Services Committee to ensure equitable access for all patients in Wales and avoid postcode lotteries.**
- **Allow patients access to lupus expertise and/or Centres of Excellence in England at agreed stages of their illness whilst there exists no such provision in Wales. The NHS system in Wales should not be a blunt instrument to deny patients and their clinicians choice, where to do so will have a negative impact on patient outcomes.**
- **In line with Welsh Government's Covid19 Clinical Recovery Plan, 'Looking Forward', which emphasises innovative approaches and collaboration, work with Lupus UK on facilitating the employment of specialist lupus nurses in every health board or, if population need is insufficient, for nurses to be a shared resource between neighbouring health boards.**
- **Agree on a plan and timescale to establish Wales's own Centres of Excellence for lupus patients. To ensure geographical coverage, Centres of Excellence should be initiated in both North and South Wales. Whilst the numbers of lupus patients in Wales may not warrant centres offering full-time provision, this should not be a barrier to their development. Equivalent recognition and support for people living with other rare auto-immune diseases should also be provided.**

- **Formally recognise that lupus is a sub-specialist area of rheumatology medicine, and not within the remit of general rheumatology. Consider facilitating a funding scheme that would give general rheumatology clinicians in Wales the opportunity to apply for training and development of expertise as part of a formal, holistic care pathway to include tertiary centres of excellence as developed and endorsed by Lupus UK and in line with 'Innovate to Save' principles.**
- **Work with GP Trainers and GP Surgeries to improve management and care for patients with lupus and other rare auto-immune diseases between consultant appointments. GPs should have the autonomy to refer patients directly to Centres of Excellence where there is clear evidence of patient need and preference, in line with NICE principles of Shared Decision-Making.**
- **Recognise and act to address urgently the equality implications of not revisiting lupus and RAIRD diagnosis and management in Wales. Welsh Government has declared itself a 'Feminist Government' and Wales an 'anti-racist' nation. However, the existing service failures for these conditions are having a particularly negative impact on females and BAME people who make up the majority of patients and patients with severe disease.**
- **Undertake research on the implications Covid19 has for wider understanding of auto-immune diseases. Note that Long-Covid has an 80% female prevalence, much like RAIRDs, ME, CFS, and Fibromyalgia and commit to enhanced and co-produced medical education and training on these conditions to improve care for sufferers.**
- **Reconsider 'Care Closer to Home' as a blanket precept for all patients in all circumstances and with all services. As it stands, it is in direct opposition to the principles of individualised care, with the patient at the 'centre'. Recognise that patients' own statement of need must be central to decision-making and that for some patients there will be a preference to travel to NHS Centres of Excellence for their condition rather than repeatedly access less specialised care locally.**
- **Revisit the IPFR mechanism to ensure it is fairly applied to patients requiring tertiary specialist care. Patients and GPs should be empowered to utilise this mechanism without necessarily requiring a local consultant's authorisation. A full public consultation on the use and implementation of IPFRs should be conducted and a Task & Finish Group made up of patients, clinicians, and NHS executives created to review its results and oversee development of best practice.**

PART EIGHT: Appendices

1) A petition with Change.org to raise awareness of our plight, again with the assistance of Lupus UK. It currently stands at 2644 signatures (25)

2) Wider policies impacting on patients

- **Prudent Healthcare**

Auto-immune and lupus services are currently not in line with Welsh legislation or policy, including the Prudent Principles of ‘patients and (healthcare) professionals as equal partners’, ‘do no harm’, and ‘reduc(ing) inappropriate variation using evidence-based practices consistently and transparently’ (26)

- **Patient and professionals as equal partners in decision-making**

NICE published its Guideline on Shared Decision Making in May 2021 (27), the implementation of which, like other NICE guidelines, is a commitment made by Welsh Government. The NHS system in Wales, where ‘care closer to home’ underpins healthcare policy can actually serve to undermine a commitment to shared decision-making because it acts as a barrier to specialist tertiary care out of area which is often the preference of patients with complex and chronic health conditions.

- **Care Closer to Home**

In 2019, Welsh Government published its long-term action plan for health and social care in Wales, ‘A Healthier Wales’ (28). As early on in the document as page 3, Welsh Government states its commitment to ‘care closer to home’.

A 2021 survey conducted by FTWW of its online group membership revealed that 100% of respondents had never been offered a choice between accessing standard care closer to home or specialised care farther afield. Further, 100% of respondents said that they would prefer to travel to a tertiary clinic offering specialised multi-disciplinary approaches for their condition. Respondents said that they would prefer to access care closer to home for more ‘minor’ or commonplace issues.

- **The Social Services and Wellbeing (Wales) Act 2014** (29) has ‘voice, choice, and control’ as one of its central tenets. However, the NHS system in Wales, which denies patients the choice of where and how to access health services largely takes away the ability to control one’s own health and wellbeing.

- **The Wellbeing of Future Generations (Wales) Act 2015** (30) commits public bodies to fully consider the impact their decisions could have on people living in Wales now and in the future. Five principles underpin decision-making: long term; prevention; integration; collaboration; & involvement. The situation for Lupus & RA patients at present falls short on all 5 counts and there seems to be no forward-planning to improve things.

On the Future Generations website, commentary states that, ‘The Commissioner’s current analysis is that there is compelling evidence that we are not investing in the best balance of services to keep people well and to enable them to live healthy and fulfilled lives. Many well-being objectives and steps acknowledge the need to shift

towards prevention and wellness, but need to go further in how they fund these interventions and transform the way they deliver services' (31).

- **NHS Wales Clinical Recovery Plan, 'Looking Forward'** (32)
Published in 2021, Welsh Government's plan for NHS services across Wales post-pandemic highlights collaboration between health boards by way of 'regional' diagnostic hubs, and additional training for clinicians for them to meet the healthcare needs of complex patients. It also posits the development and implementation of Quality Statements for major conditions. Where Lupus and other RAIRDs are concerned, all of these commitments could serve to improve care available. Additional training for local practitioners and provision of advanced skills nurses could meet the 'care closer to home' objective, whilst investment in regional hubs, bringing together an appropriate multi-disciplinary team, could fit requirements of a Lupus Centre of Excellence.
- 3) The **Rheumatology in Wales: State of Play Report 2016** (33) highlighted significant variations in meeting the standards of care developed by the British Society for Rheumatology (BSR), NICE, and the Welsh Government. Concerns raised included practical barriers to accessing care and the IPFR system's lack of accountability and time-consuming nature.

The report's evidence base included patient testimonies regarding their recent experiences of being denied access to expert lupus care, with many reporting feeling as though the system was deliberately complex and impenetrable so that they give up seeking care.

The report also expresses concerns about NHS Wales's focus on encouraging more doctors to be generalists rather than specialists, a concern shared by patients. Policies in Wales, far from encouraging innovation in service provision or improved patient outcomes, are serving more to prevent meaningful progress in lupus patient care in the region.

- 4) American College of Rheumatology - List of Criteria for Lupus Diagnosis (34)
- Anti-nuclear Antibodies;
 - Fever
 - Leukopenia
 - Thrombocytopenia;
 - Autoimmune Hemolysis
 - Delirium;
 - Psychosis;
 - Seizure;
 - Non-scarring Alopecia;
 - Oral ulcers;
 - Subacute Cutaneous or Discoid Lupus;

- Acute Cutaneous Lupus (includes Malar Rash)
 - Pleural or Pericardial Effusion;
 - Acute Pericarditis;
 - Joint Involvement;
 - Proteinuria (greater than 0.5g / 24 hours)
 - Class II or V lupus nephritis on renal biopsy according to ISN/RPS 2003 classification
 - Positive Antiphospholipid Antibodies;
 - Low C3 OR Low C4;
 - Low C3 AND C4;
 - Anti-dsDNA antibodies OR anti-Sm.
- 5) As of writing this report, there are 2 Lupus Specialists in Wales recognised as such by Lupus UK:
- South Wales - Dr Julian Nash Consultant Rheumatologist, BSc, MB Bch, PhD, MRCP, who has a PhD in Systemic Lupus Erythematosus and studied in Imperial College Hammersmith. Hammersmith Hospital was Europe's first dedicated Lupus Clinic, established in 1993 by the UK's leading lupus specialist Professor Graham Hughes.
- North Wales - Dr Yasmeen Ahmad who has developed a specialist interest in Lupus after undertaking a PhD which examined the Traditional and Novel Cardiovascular Risk Factors as predictors of cardiovascular disease in SLE.
- 6) Lupus UK lists the following criteria for the role of specialist Lupus Nurse (35):
- To run patient education clinics and drop-in sessions
 - To provide longer patient appointments
 - To offer advice through an email and telephone helpline
 - To assist with lupus research
 - To run 'fatigue management' workshops (almost 90% of people with lupus experience this difficult to treat symptom)
 - Support any local lupus groups

The charity is currently funding specialist lupus nurses in:

- Belfast, Northern Ireland
- City Hospital, Birmingham
- Edinburgh
- Lanarkshire, West of Scotland
- Leicester
- North Wales
- Queen Elizabeth Hospital, Birmingham
- Royal Free Hospital, London
- St George's Hospital, Tooting

- 7) At time of writing, Lupus UK (36) has 10 approved Centres of Excellence, all in England, as follows:
- Alder Hey Children’s Hospital in Liverpool, a Paediatric Rheumatology Service for Juvenile SLE patients, opened in 2010,
 - City Hospital / Queen Elizabeth Hospital / Birmingham University opened in October 2005,
 - Kellgren Centre at Manchester Royal Infirmary opened in October 2007,
 - Royal National Hospital for Rheumatic Disease in Bath opened in March 2016,
 - Louise Coote Lupus Unit, the largest in Europe, opened September 2014,
 - The Centre for Rheumatology at UCLH in London, opened in July 2012,
 - Addenbrookes in Cambridge, opened in July 2018
 - University Hospital Southampton, under Professor Chris Edwards, 2020,
 - Barts Lupus Centre, Mile End Hospital, London, 2020 and
 - Great Ormond Street Hospital, London – second ever Paediatric Lupus Centre of Excellence, 2020.
- 8) **The UK Strategy for Rare Diseases 2020** is a plan to which Welsh Government has committed itself, with consultation on its delivery under way presently. It follows on from the 2017 Welsh Rare Diseases Implementation Plan (37), produced by the Welsh Rare Disease Implementation Group (part of Welsh Government).

Politically, the Senedd Cross-Party Group on Rare Diseases has committed to keeping a watching brief on how the Rare Diseases Plan will be delivered in Wales. Most notable is the need for auditable outcomes based on patient reported outcomes, along with clear targets and time-scales for delivery. This follows what many patients and advocates consider the failure of the 2017 Rare Disease Strategy for Wales with concerns expressed at the lack of tangible progress in delivering its recommendations, namely:

- Equity of access, incorporating clear, well defined care pathways, providing high quality services between primary, secondary, and specialised care appropriately and without delay for every individual.
- A patient centred, coordinated approach to treatment services, specialist healthcare and social care support
- Local service providers aware of their limitations and being prepared to refer on, or seek advice, from specialist centres or experts.
- Utilising specialist providers in England.
- Implementing changes to the Individual Patient Funding Request (IPFR) process, as recommended by a 2016 review which allows for interventions for rare

diseases to be assessed on the basis of whether they offer significant clinical benefit and reasonable value for money.

- 9) Part of the research underpinning the 2020 UK Rare Disease Strategy is the **CONCORD (CoOrdINated Care of Rare Diseases) Study** (38) in which FTWW members participated.

The study is looking at how care services for people with rare diseases are coordinated in the UK, and how people affected by rare diseases and health care professionals who treat rare diseases would like them to be coordinated. It is a collaboration between patients and carers affected by rare conditions, health care professionals with expertise in rare conditions, and health services researchers. One of their key recommendations was around the need for dedicated Care Co-ordinators, to assist patients in navigating a complex and disjointed health and social care landscape.

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PART TEN: Acknowledgements

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FTWW