Irish Society for Rheumatology

Spring Meeting

19-20 May 2022

Welcome Message from the ISR President Prof Geraldine McCarthy

Dear Colleagues and Friends

It is wonderful to be able to welcome you in person to this meeting in Sligo, our first on-site meeting since 2019. While we have had some excellent virtual meetings, they did not permit the real togetherness that our society usually enjoys. I have no doubt that we will make up for lost time in Sligo where our colleagues Carmel Silke, Bryan Whelan and Miriam O'Sullivan have put together an excellent program.

On Thursday evening, Professor Hector Chinoy will bring us up to date on the management of inflammatory myopathy. Then we will have the pleasure of Dr Ronan Kavanagh's insights into the 'Music of Rheumatology. On Friday morning, Dr David Kiefer will discuss the role of JAKi's in AS and PsA. I am delighted that my colleague from the Mater, Dr Kate O'Reilly will speak to us about interstitial lung disease in rheumatic diseases. She is a tremendous resource for our patients. We will then have presentations of clinical cases, which never fail to keep us thinking. After lunch, we will hear from Dr Nicola Goodson who will present virtually from Liverpool on co-morbidities in axial spondyloarthropathies. Finally, Dr Eoghan McCarthy will share his expertise in relation to biologic use in SLE.

I believe Rheumatology in Ireland is in a very positive place. There have been new Rheumatology posts generated country-wide via the National Clinic Program thanks to the excellent efforts of Professor David Kane. This year 6 candidates have been appointed to Rheumatology SpR positions and we have made excellent progress in harnessing Gainshare funds to the ultimate benefit of Rheumatology in Ireland and hence our patients.

I would like to offer special thanks to Michael Dineen and Marie Caston who have yet again worked extremely hard to make this meeting a success.

I am especially grateful to our colleagues in the Pharmaceutical Industry for the continued support of ISR and its members, especially during these challenging times.

Finally, my sincere thanks to the members of ISR board for all their continued support in 2022 and also since 2020, especially given all the challenges we have faced.

Please enjoy the meeting,

Prof Geraldine McCarthy
President ISR

ISR Spring Meeting
19-20 May 2022
Programme

Thursday, 19 May 2022

16.15 **CAG Meeting – Chaired by Professor David Kane**

17.25 **Opening Address – Professor Geraldine McCarthy, President, ISR**

17.30 **Prof Hector Chinoy**
Professor of Rheumatology and Neuromuscular Disease
at The University of Manchester, UK.
"Update in the management of inflammatory myopathy"

18.15 **Dr Ronan Kavanagh**
Consultant Rheumatologist, Galway Clinic and Bon Secours Hospital, Galway.
"The Music of Rheumatology"

Friday, 20 May 2022

09.00 **AbbVie Satellite Symposium**
Dr David Kiefer
Consultant Rheumatologist at the Rheumazentrum Ruhrgebiet Herne, Germany.
"The role of JAKi's in AS and PsA: reviewing the evidence for upadacitinib"

10.00 **Dr Katherine O'Reill**y
Consultant in Respiratory and Acute Medicine MMUH, Dublin.
"Interstitial lung disease in Rheumatic diseases"

10.45 **Coffee**

11.15 **Clinical Cases**

12.15 **STC Meeting**
Dr John Ryan,
Consultant Rheumatologist, CUH

12.45 **Lunch**

14.00 **Dr Nicola Goodson**
Consultant Rheumatologist, Liverpool University Hospitals NHS Foundation Trust.
"Comorbidities in Axial Spondyloarthritis"

14.45 **Dr Eoghan McCarthy**
Consultant Rheumatologist, Beaumont Hospital, Dublin
"Biologic - Use in SLE"

15.30 **Prize Giving and Close of Meeting**

Biographical Sketches

Speakers

Professor Hector Chinoy
Professor of Rheumatology and Neuromuscular Disease at The University of Manchester, UK.

Hector Chinoy is a Professor of Rheumatology and Neuromuscular Disease at The University of Manchester, UK, where he leads an active programme of translational research within the Manchester Myositis Research Group. At Salford Royal NHS Hospital Foundation Trust, he is an Honorary Consultant Rheumatologist and leads the Rheumatology Neuromuscular service. He has recently led the BSR myositis guidelines and is Chief Investigator of a study investigating JAK inhibition in patients with inflammatory myopathy.

Dr Nicola Goodson
Consultant Rheumatologist, Liverpool University Hospitals NHS Foundation Trust.

Dr Goodson trained in Medicine at the University of Liverpool. She obtained a PhD in Epidemiology for her work exploring cardiovascular morbidity and mortality in patients with early inflammatory polyarthritis.
After completing Rheumatology training she was employed as a Senior Lecturer in Rheumatology at University of Liverpool. She is currently employed as a Consultant Rheumatologist at Liverpool University Hospitals NHS Foundation Trust.
She runs the Axial Spondyloarthritis service and high cost rheumatology therapy service at this site. Dr Goodson has research interests in work, epidemiology, pharmacoepidemiology and comorbidity of inflammatory rheumatic conditions. She Chairs the British Society of Rheumatology Biologic Registers Committee.

Dr Ronan Kavanagh
Consultant Rheumatologist, Galway Clinic and Bon Secours Hospital, Galway.

Dr Ronan Kavanagh is an easily bored rheumatologist working in full time private practice in Galway. He is interested in the space where medicine meets the rest of the world - particularly the arts and humanities, but also in the meaningful use of technology. He is the founder of the award winning dotMD Festival of Medical Curiosity, is interested in what rheumatologists can do for musicians, but also in what rheumatologists can learn from them about the art of the practice of medicine. He is also a former president of the ISR.

Dr David Kiefer
Consultant Rheumatologist at the Rheumazentrum Ruhrgebiet Herne, Germany

Dr David Kiefer is a consultant rheumatologist at the Rheumazentrum Ruhrgebiet Herne and the Ruhr-University Bochum, Germany. He studied Human Medicine at the university of Essen, receiving his official Board Degrees in Internal Medicine and Rheumatology in 2014.
His research interests are focused in the field of spondyloarthritides, with special emphasis on patient outcomes and physical function and imaging. In addition, he is part of a team conducting clinical trials in these areas. In collaboration with AbbVie, he designed the SPINEtronic study which focuses on spinal mobility impairments in patients with axial spondyloarthritis with initial results recently published.

Dr Eoghan McCarthy
Consultant Rheumatologist, Beaumont Hospital, Dublin

Eoghan McCarthy is a consultant rheumatologist and general physician at Beaumont Hospital, Dublin and senior lecturer at Royal College of Surgeons in Ireland. He graduated from UCC in 2003 and completed his specialist training in Ireland. During this time he completed a PhD investigating the role of monocyte dysfunction in SLE as well as a Masters in Medical Education through NUI Galway. He subsequently undertook a fellowship in Lupus and vasculitis in the Kellgren Centre for Rheumatology, Manchester following which he was appointed as a consultant rheumatologist with a special interest in CTD/Vasculitis in the Unit. Prior to his return to Beaumont he was clinical lead for rheumatology as Manchester Royal Infirmary. His research interests in the safety and effectiveness of biologics in SLE as well as endothelial dysfunction in CTD.

Dr Katherine O'Reilly
Consultant in Respiratory and Acute Medicine MMUH, Dublin.

Dr Kate O'Reilly is a Consultant Respiratory Physician and Associate Clinical Professor based at the Mater Misericordiae University Hospital in Dublin. She has a longstanding interest in interstitial lung disease (ILD) including connective tissue disease related ILD and leads the Mater's ILD service and chairs the ILD multi-disciplinary meeting. With her colleague, rheumatologist Dr John Stack she set up a combined respiratory –rheumatology clinic based at the Mater. Research interests include mechanism of fibrosis and matrix biology, exercise physiology and pulmonary rehabilitation in ILD and she is principal investigator on a numerous of clinical trials in IPF and Scleroderma associated ILD.

ISR Board members

Professor Geraldine McCarthy
President
Consultant Rheumatologist
Mater Misericordiae University Hospital Dublin and Full Clinical Professor of Medicine University College Dublin

Geraldine McCarthy graduated in Medicine from NUI. She received her Fellowship in Rheumatology at the Medical College of Wisconsin. Her research has focused on the biological effects of calcium-containing crystals in degenerative joint disease as well as in atherosclerosis and breast cancer. Promoted to Associate Professor of Medicine at the Medical College of Wisconsin in 1996 where she remained until her return to Dublin.
Prof McCarthy was appointed Consultant in Rheumatology at the MMUH and Cappagh National Orthopedic Hospital Dublin in 1999 where she continues to run a busy clinical practice. She teaches as part of the University College Dublin Faculty of Medicine where she was the first clinician to be appointed Full Clinical Professor of Medicine through the Clinical Pathways in 2009.
Geraldine has current international collaborations in the UK, USA, Europe, Australia, New Zealand and Canada, particularly in relation to calcium crystal deposition diseases as well as gout. She continues her involvement in bench research related to the pathogenesis of basic calcium phosphate crystal–induced joint disease and participates in and contributes to numerous international collaborations related to gout. Other research interests include platelet activation in inflammatory arthritis and its role in enhanced cardiovascular risk. She also participates in collaborative studies of the pathogenesis of giant cell arteritis and HIV-associated bone pathology.
Author of over 130 publications, including original manuscripts, editorials, reviews and book chapters and has spoken at many national and international meetings. She has been winner of several research and teaching awards and has mentored many medicine and science graduates in clinical practice and in research.

Dr Claire Sheehy
Honorary Secretary
Consultant Rheumatologist
University Hospital Waterford

Dr Claire Sheehy is a Consultant Rheumatologist in University Hospital Waterford. A graduate of Trinity College Dublin, she completed the higher specialist training in rheumatology and general medicine, and was awarded an MD for work exploring the role of anti TNF therapy in early rheumatoid arthritis. She undertook a fellowship in connective tissue disease and vasculitis between Norfolk and Norwich University Hospital, and Addenbrookes Hospital. She took up her post in 2012; her current clinical interests include early inflammatory arthritis and connective tissue disease.

Dr Shawn Chavrimootoo
Honorary Treasurer
Consultant Rheumatologist,
Our Lady's Hospital,
Navan, Co Meath.

Shawn Chavrimootoo is a Consultant Rheumatologist at Our Lady's Hospital, Navan, Co. Meath. He graduated in Medicine from RCSI, Dublin in 2002 and developed an interest in Rheumatology during his Senior House Officer years in Connolly Hospital, Blanchardstown. Following this, he completed higher specialist training in Cork University Hospital, Kerry General Hospital, Connolly Hospital and St Vincent's University Hospital in Dublin. He was appointed to his Consultant Rheumatologist post in 2013 when he joined Dr Ramakrishnan at Our Lady's Hospital, Navan, from where they currently provide a regional Rheumatology service for the North East of Ireland. His clinical interests include osteoporosis as well as gout, inflammatory arthritis, spondyloarthritis, connective tissue disease and vasculitis.

Dr Nicola Ambrose
Consultant Rheumatologist,
Blackrock Clinic, Co Dublin

Dr Nicola Ambrose is a graduate of Trinity College Dublin. She completed her specialist training in rheumatology and general internal medicine in Ireland, before obtaining an Arthritis Research UK (ARUK) fellowship to undertake a PhD at Imperial College London, studying inflammation in Behçet's Syndrome. She then obtained a Richard Steeven Fellowship from the HSE to undertake a Clinical Fellowship at the ARUK Adolescent Rheumatology Centre of Excellence at University College London Hospital (UCLH). She stayed at UCLH as an Adolescent and Adult consultant rheumatologist, and was the Clinical Lead for Adolescent Rheumatology. Special interests: Adolescent and Young Adult Rheumatology including JIA; Behçet's Syndrome; SLE and dermatomyositis; Gout Osteoporosis and fracture secondary prevention; Inflammatory arthritis.
She has published 23 peer review papers as well as 6 book chapters.

Dr Elizabeth Ball
Consultant Rheumatologist
Musgrave Park Hospital/
Belfast City Hospital

Dr Liz Ball is a graduate of Queen's University Belfast and was appointed as a Consultant Rheumatologist at Musgrave Park Hospital/ Belfast City Hospital in 2014. She is also an Honorary Lecturer at Queen's University Belfast. She has a special interest in autoimmune disease and lupus and was awarded an MD entitled *'A study of hand arthritis in Systemic Lupus Erythematosus from a Clinical, Imaging and Cytokine Perspective'* from Queen's University in 2013. She is involved in postgraduate medical education and holds a Training Programme Director role within the Northern Ireland Deanery and is currently completing a Masters in Clinical Education. She is a musculoskeletal ultrasound tutor and regularly teaches regionally and nationally.

Dr Andrew Cairns
Consultant Rheumatologist,
Musgrave Park Hospital, Belfast

Dr Andrew Cairns graduated in Medicine from Queen's University Belfast in 1995. He completed specialist training in Belfast and also at the Rheumatic Diseases Unit in Edinburgh. He was awarded an MD by thesis entitled "Leucocyte surface receptor expression of relevance to apoptotic cell clearance in systemic lupus erythematosus" from Queen's University Belfast in 2001, and an MSc in Sport and Exercise Medicine from the University of Ulster in 2008. He is a Consultant Rheumatologist at Musgrave Park Hospital Belfast where he was appointed in 2004, and also provides rheumatology clinics at Belfast City Hospital and Lagan Valley Hospital Lisburn. He is a Fellow of the Royal Colleges of Physicians of Ireland, London and Edinburgh. He has published in a wide range of rheumatic diseases and is an enthusiastic proponent of musculoskeletal ultrasound.

Dr Michele Doran
Consultant Rheumatologist and General Physician, St James's Hospital Dublin

Dr. Michele Doran has been working as a Consultant Rheumatologist and General Physician at St. James's Hospital, Dublin since 2003. She graduated in Medicine from UCD in 1993, and completed her clinical training in General medicine and Rheumatology in Dublin and Bath, UK. She undertook a 2 year Research Fellowship at Mayo Clinic, Rochester, USA, where she completed an MD degree with research relating to the Epidemiology of Rheumatoid Arthritis. During this time she completed a Master's Degree in Biomedical Sciences, Clinical Research, in the Mayo Clinic Graduate School. She was involved with the establishment of and is on the steering committee for the Rheumatoid Arthritis Biologics Registry of Ireland (RABRI).

Professor Ursula Fearon
Head of Molecular Rheumatology,
School of Medicine,
Trinity Biomedical Sciences Institute,
Trinity College Dublin.

Professor Ursula Fearon is head of Molecular Rheumatology, School of Medicine, Trinity Biomedical Sciences Institute, Trinity College Dublin. Professor Fearon's research is a bench-to-beside translational approach, focusing on understanding the underlying mechanisms that drive disease pathogenesis; her team specifically examine components of joint inflammation at a cellular and molecular level to dissect out the signalling and gene pathways that are involved in the pathogenesis of inflammatory arthritis and rheumatic diseases. She has established strong collaborative research networks across Europe, USA and Singapore. Professor Fearon, has been awarded significant research funding from Arthritis-Ireland, Health Research Board, Science Foundation Ireland, IRCSET, European-ASPIRE, JU Innovative Medicines Initiative (IMI) and Maeve Binchy Funding for Arthritis Research, in addition to industry collaborative partnerships. She has published extensively in high impact peer-reviewed journals, and her research has been awarded several National/International awards.

Professor David Kane
National Lead for Rheumatology
HSE Clinical Programme
Consultant Rheumatologist,
Tallaght University Hospital, Dublin

Prof David Kane attended medical school at Trinity College, Dublin, Ireland and was conferred MB BCh BAO BA in 1991, PhD in 2002 and FRCPI in 2006. He has trained in rheumatology with Prof. Barry Bresnihan and Prof. Oliver FitzGerald at St. Vincent's University Hospital, Dublin, Ireland and with Prof Roger Sturrock, Prof Iain McInnes and Dr Peter Balint at Glasgow Royal Infirmary, Glasgow, United Kingdom. He was appointed as Senior Lecturer in Rheumatology at the University of Newcastle (2003-2005) and is currently working as Consultant Rheumatologist at the Adelaide and Meath Hospital and Clinical Professor in Rheumatology at Trinity College Dublin. His special interests are musculoskeletal ultrasound, spondyloarthopathy and synovial inflammation. He is a member of the European Working Party on Musculoskeletal Ultrasound and the OMERACT special interest group on musculoskeletal ultrasound, previous organiser of the BSR Musculoskeletal Ultrasound course and is Faculty member of the EULAR Musculoskeletal ultrasound course. He has served as a Board member of the Irish Osteoporosis Society, as President and Treasurer of the Irish Society for Rheumatology and is currently a Board member of Arthritis Ireland.

Dr Bernadette Lynch
Consultant Rheumatologist,
University Hospital, Galway.

Dr Bernadette Lynch graduated from the Royal College of Surgeons in Ireland in 2003. She completed her higher specialist training in Rheumatology and General Medicine in 2013 having worked and studied in Dublin, Galway and London. She was awarded an MD from University College Dublin in 2011 for work on IL-22 and musculoskeletal ultrasound in Inflammatory Arthritis. She undertook a fellowship in Scleroderma and Vasculitis at the Royal Free Hospital Hampstead under Professor Chris Denton and Dr Aine Burns. During this time, Bernadette was part of the UK Scleroderma Study Group (UKSSG) which developed the national guidelines on the management of complications of Scleroderma. She took up her current appointment as Consultant Rheumatologist and General Physician in University Hospital Galway in 2015. Her principal clinical and academic interests are Scleroderma and Inflammatory Arthritis.

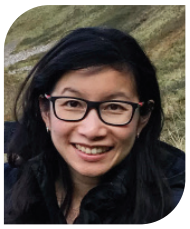
Dr John Ryan
Consultant Rheumatologist,
Cork University Hospital, Cork

Dr John Ryan is a graduate of the Royal College of Surgeons in Ireland, he completed his higher medical training in rheumatology and general internal medicine in Ireland. He undertook a fellowship at the National Institute of Arthritis and Musculoskeletal and Skin Diseases (NIAMS) in Bethesda, Maryland. During this time he undertook translational research into disordered innate immunity manifesting as recurrent fever syndromes. He joined Dr Sinead Harney in the Rheumatology service at Cork University Hospital in 2010. The Rheumatology department has since expanded to include Dr Grainne Murphy. In July 2017 he took up the post of National Specialty Director for Rheumatology.

Dr Emma Jane MacDermott
Consultant Paediatric Rheumatologist,
CHI Crumlin

Emma Jane MacDermott, is a Consultant Paediatric Rheumatologist in CHI Crumlin where she joined the team in 2012 and has helped oversee the ongoing growth and development of the paediatric rheumatology department into a dynamic national service, now including a growing research and education component. With a special interest in education she enjoys working with patients, parents and medical providers to raise the profile and understanding of rheumatologic disease. She works with the national advocacy groups continuing to raise the profile for Irish paediatric rheumatology patients. Areas of interest include Juvenile arthritis, Paediatric Lupus and autoinflammatory disease.
A graduate of University of Dublin, Trinity College Medical School she pursued her post graduate training in paediatrics, becoming a member of the Royal College of Physicians in 2001. She subsequently moved to New York, where she completed a fellowship in Paediatric Rheumatology, from Weill Cornell Medical School, working at Hospital for Special Surgery and the Cornell Campus of New York Presbyterian Hospital as Assistant Attending in Paediatric Rheumatology at Hospital for Special Surgery and Assistant Professor of Paediatrics at Weill Cornell Medical School until her return to Ireland in 2012.
Emma is a member of the Royal College of Physicians of Ireland, the American College of Rheumatology, the Irish Rheumatology Society, the British Society of Adolescent and Pediatric rheumatology.

Dr Wan Lin Ng
SpR Rep on ISR Board



Dr Wan Lin Ng is a medical graduate from the Royal College of Surgeons in Ireland (RCSI). She has completed her basic specialist training in Ireland and is currently in the higher specialist training programme in Rheumatology. She is a recipient of the StAR MD scholarship from RCSI and the ISR Rheumatology Patient Initiative Fund. With a keen passion in teaching and education, Dr Ng was previously an affiliate tutor with University of Limerick and RCSI. Her clinical interests include connective tissue disease related interstitial lung disease and musculoskeletal ultrasound.

Dr Bryan Whelan
Consultant Rheumatologist
Our Lady's Hospital, Manorhmailton,
Co Leitrim

Dr Bryan Whelan is a Consultant Rheumatologist in Our Lady's Hospital in Manorhmailton, Co Leitrim and an Honourary Senior Lecturer in Medicine in NUIG. He qualified from UCD in 2000 and completed BST in the Mater Hospital in Dublin. He completed SpR training in Rheumatology in CUH, the Mater Hospital and University College London. He has an MD and Masters Sports and Exercise Medicine from UCC and an MSc in Epidemiology from the London School of Hygiene and Tropical Medicine. He is currently a board member of Arthritis Ireland, the SUH Research and Education Foundation, a member of the Academic Committee of the FSEM and a member of the Advisory Committee for Human Medicines Clinical Trials Subcommittee of the HPRA. His current research interests include muscle disease, exercise in rheumatology and osteoarthritis.

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Consultant Rheumatologist
Mater Misericordiae University Hospital Dublin
and Full Clinical Professor of Medicine
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Return to Normality

As we approach our first 'face to face' meeting in over two years, we reflect on the pandemic period, the negatives, and some positives. The meetings were better attended in some part, many members just joined in for the presentations which they were really interested in, some logged in and out on a few occasions during the day and meetings were slightly less expensive to organise.

A huge difficulty was to develop a way of accommodating the industry and how to allow them some means of participating in a meaningful way. The industrial videos filled this gap and by and large the industry was happy with this. Some of the videos were novel and even inspirational.

A big plus for ISR was the increase in our membership. While we did not impose a registration fee for logging in to the meetings, we insisted that in order to obtain the login details one needed to become a member of ISR, which brought our membership of the Society over the 200 mark for the first time. All membership will in future be routed through our website and no longer be dependent on outdated banking routes. Membership of ISR will as always be an annual deduction.

The big negative especially for our senior members was the inability to network. While the educational benefit of our conferences cannot be undervalued, the lack of meeting face to face, squeezing the flesh and swopping experiences cannot be overstated.

I feel that going forward we will always have some virtual element in our future meetings especially where a principal speaker is unable to travel and may have to withdraw at the 11th hour, it may be easier to attract a late substitute to present virtually.

I should also mention that for obtaining CPD certificates the ground rules are changing. One of these is that RCPI will now require a meeting evaluation form from delegates before a CPD certificate may be issued. Obtaining CPD from UK centres will also be far more stringent in the future.

Recently we have heard lots of ideas and discussions of how best to help our friends in Ukraine. One consideration is how we might extend our training and expertise to trainees in their country. The IT and Translation facilities are available to us.

Finally we are indeed well served by our dedicated officers and board members under the capable leadership of Prof Geraldine McCarthy.

Kind Regards,

Michael Dineen
Chief Executive ISR

CLINICAL CASES

22S101

A Flare of Lupus Nephritis Leading to a Diagnosis of Polycystic Kidney Disease: Case Report

Author(s)
Moollan N1, Dorman A2, Magee C3, Durcan L1

Department(s)/Institutions
1Department of Rheumatology, Beaumont Hospital, Dublin
2Department of Histopathology, Beaumont Hospital, Dublin
3Department of Nephrology, Beaumont Hospital, Dublin

Introduction
Lupus nephritis is an immune complex glomerulonephritis which typically presents with proteinuria/hematuria and decline in renal function. By contrast polycystic kidney disease (PKD) is a hereditary kidney disease that usually presents with abdominal pain and fullness, hypertension and renal failure. Both of these conditions are rare, with only two reported cases of combined lupus nephritis and PKD.

Results
We present a case of a patient with known lupus nephritis, who presented with rising creatinine levels, nephrotic range proteinuria and resistant hypertension. He was managed as a lupus nephritis flare. However, multiple bilateral renal cysts were identified prior to percutaneous renal biopsy. On further history, it was discovered that he had a positive family history for polycystic kidney disease. Based on the laboratory and ultrasound results, and family history, a diagnosis of combined PKD with lupus nephritis was made.

Conclusions
Lupus nephritis with PKD is a rare presentation. However, this case highlights the importance of keeping an open mind when evaluating patients with lupus nephritis and worsening renal indices. It also makes us consider the value of re-addressing family history, even in longstanding patients with established diagnoses.

22S102

A Tale of Two Pregnancies

Author(s)
Moollan N 1, Durcan L 1

Department(s)/Institutions
1 Department of Rheumatology, Beaumont Hospital, Dublin

Introduction
Systemic Lupus Erythematosus(SLE) primarily affects women of childbearing age. Although SLE does not adversely impact fertility, it is associated with higher rates of preterm birth, preeclampsia, growth restriction, fetal loss, and neonatal lupus. Predictors of adverse pregnancy outcomes include active disease, prior nephritis, hypocomplementemia, anti-dsDNA antibodies, antiphospholipid antibodies, thrombocytopenia, and antihypertensives.

Aims/Background
We present a case of a patient diagnosed with SLE during pregnancy, resulting in lupus nephritis, APS, stroke, hypertension, seizures, and early foetal demise, who after achieving disease remission, proceeded to have a successful pregnancy.

Results
A 35-year-old of 20 weeks gestation presented following a generalised tonic-clonic seizure, she was found to be hypertensive, proteinuric, and as such was treated as pre-eclampsia. Her background included an IVF pregnancy, miscarriage at 10 weeks gestation, primary infertility, and previous ITP. Medications included aspirin and prophylactic clexane. On admission she had thrombocytopenia, elevated creatinine levels, low C3/C4, dsDNA antibody and triple antiphospholipid antibody positive. MRI brain showed old infarcts. Renal biopsy confirmed active lupus nephritis. For nephritis she was commenced on azathioprine, hydroxychloroquine and prednisolone, for APS she was commenced on therapeutic clexane and aspirin, for hypertension she was commenced on labetolol, and for seizures she was commenced on levetiracetam. However despite this treatment, her baby died in-utero at 25 weeks gestation. Despite a pregnancy complicated by lupus nephritis, APS, stroke, hypertension, seizures, and early foetal demise, our patient had a strong maternal wish for a further pregnancy. Azathioprine was switched to mycophenolate, labetolol to ramipril, and clexane to warfarin. At review 1 year later, she had normal BP, renal function and platelet count, with improvement in dsDNA antibodies, C3/C4 counts. She was switched back to azathioprine, continued prednisolone and hydroxychloroquine, ramipril was stopped, she was switched to therapeutic clexane and continued on aspirin, she was given 2 cycles of rituximab, and continued on levetiracetam. The IVF cycle was successful and she remained normotensive, non-proteinuric and seizure free throughout the pregnancy. She delivered a healthy baby at 37 weeks gestation.

Conclusions
Our case highlights the importance of preconception planning and achieving disease remission in patients with SLE in order to optimise pregnancy outcomes.

22S103

Confusion with a positive antinuclear antibody test.

Author(s)
Majeed Haider, Joe Devlin, Alwin Sebastian, Alexander Fraser, Fahd Adeeb, Kieran Murray

Department(s)/Institutions
Department of Rheumatology, University Hospital Limerick, Ireland

Introduction
Approximately one third of Systemic Lupus Erythematosus (SLE) patients develop neuropsychiatric syndromes attributed to SLE. Most of these neuropsychiatric events occur following the diagnosis of SLE and are accompanied by increased disease activity. However, in a minority of patients, neuropsychiatric events precede SLE diagnosis which can make the attribution of these events to SLE more challenging.

Aims/Background
We describe a case of acute encephalitis associated with a positive antinuclear antibody (ANA).

Method
Case Presentation
A 33-year-old Brazilian female presented with behavioral changes for one day followed by witnessed generalized tonic-clonic seizures and confusion. On admission, she was febrile, and Glasgow Coma Scale (GCS) was 14/15.
Initial investigations revealed a leukocytosis with raised C-reactive protein. Cerebral spinal fluid analysis showed pleocytosis and normal protein. Brain Magnetic Resonance Imaging with contrast was unremarkable, but electroencephalogram showed diffuse slowing consistent with non-specific encephalopathy.
She was initially managed as infectious encephalitis without response to antimicrobial therapy. On day four of admission, she deteriorated with a GCS of 3/15 requiring intubation. An infectious etiology was not detected. ANA, however, was 1/1600 speckled pattern with negative extractible nuclear antigen panel and normal complements. Further collateral information from patient's partner revealed history of alopecia, fatigue, arthralgia, and rash along with positive ANA serology suggestive of lupus which had not required treatment at the time.

She was treated with intravenous methylprednisolone, intravenous immunoglobulins, and cyclophosphamide for possible neuropsychiatric lupus. However, subsequent investigations revealed a positive anti-NMDA receptor (NMDAR) antibody consistent with autoimmune encephalitis. Investigations for triggers of NMDAR autoimmunity led to the finding of an ovarian teratoma.

Results

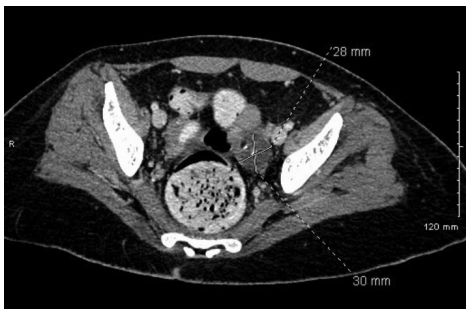
Discussion

Differentiating neuropsychiatric lupus from other autoimmune neurologic syndromes remains challenging despite recent advances. There is scientific evidence for a potential link between anti-NMDAR encephalitis and the development of autoantibodies or autoimmune diseases. Current literature also points toward a possible role for anti-NMDAR antibodies in the pathogenesis and diagnosis of neuropsychiatric lupus. Additional studies suggest an association between ovarian masses and ANA positivity.

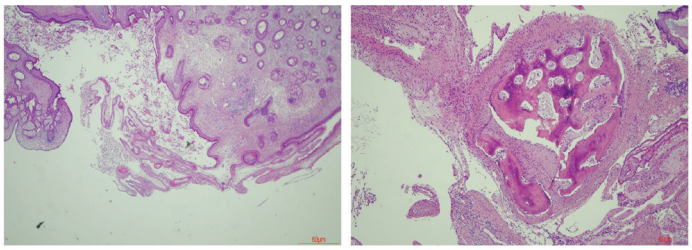
Conclusions

This case highlights the importance of maintaining a broad differential for neuropsychiatric events in patients with suspected SLE and the importance of careful interpretation of a positive ANA.

Figure



Figure



22S104

"Identification of NLRP6 mutation in a Patient with an Autoinflammatory Disorder."

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Introduction

Autoinflammatory disorders, characterized by recurrent inflammatory episodes driven by the innate immune system, are classically present in childhood 1. Several inflammasome mutations are associated with autoinflammatory conditions, including nucleotide-binding oligomerization domain (NOD)-like receptors (NLR) mutations. NLRP6 is a recently discovered inflammasome, which has very few reports linking it to autoinflammatory disease 2. NLRP6 deficiency negatively regulates inflammasome assembly, leading to a localized inflammatory response 3. We report a case of "complex autoinflammatory disorder with an NLRP6 mutation."

Aims/Background

A 20-year-old female with recurrent erythema nodosum / panniculitis, responsive to moderate dose steroids and Adalimumab, since the age of 6 months. Biopsy of liver confirmed lobular granulomatous hepatitis. She suffered chronic anorexia and was on the 50th centile for weight and height in 2008 and the 2nd-9th centile in 2013. Before age 12, she had several episodes of severe left knee pain and swelling relieved by high dose oral steroids. CRP and ESR were raised despite steroids, Adalimumab, sulphasalazine and 6-mercaptopurine. She had subtle endoscopic changes of inflammatory bowel disease with normal histology. In 2013, she had reduced diffusion on pulmonary function tests. An NLRP6 gene mutation was isolated- ADA2 levels were normal. The patient was trialled on Tocilizumab but discontinued due to persistently raised serum amyloid A in 2019, triggering panniculitis, arthritis and malaise requiring hospitalisation, and at that stage, Adalimumab was restarted. Her disease is clinically in remission at present.

Method

Discussion:

NLRP6 mutation has mainly been investigated in rodents and has very few notable case reports. This case has been classified as a complex autoinflammatory disorder. Even though this patient had elevated IL-6 levels, Tocilizumab caused the recurrence of symptoms.

Results

Learning Point(s): An important learning point from the case is to treat the patient, not the lab result. This patient's symptoms remain under control on Adalimumab.

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22S105

An insidious case of an Igg4 related disease mimic

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Introduction

A 57 year old man was referred to inpatient Rheumatology services for consideration of an autoimmune aetiology for a pericardial effusion noted during diagnostic workup for an acute stroke

His past medical history was remarkable for a recent admission with critical limb ischaemia. He also had depression, type 2 diabetes and psoriasis.

Aims/Background

Diagnostic Testing

MRI brain confirmed brainstem stroke with no evidence of a vasculitic cause. TTE revealed an incidental large pericardial effusion of 2.5cm which evolved into tamponade requiring pericardiocentesis with drainage of 750mls of exudative effusion. Cardiac MRI showed no evidence of pericardial thickening or definitive enhancement. PET scan was completed which did not show any abnormal uptake. Temporal artery biopsy showed no evidence of arteritis. His quantiferon was strongly positive. Igg4 subclass was the upper limit of normal at 0.82 (0.039 – 0.864g/l) with a low C3. While TB cultures from pericardial fluid were sterile, the infectious diseases service commenced quadruple anti-tuberculous therapy.

Method
During follow up the patient developed a recurrent pericardial effusion necessitating pericardiectomy. Histology showed an acute lymphocytic process with organising fibrinous reaction. He was re-referred to Rheumatology due to severe anterior knee pain which left him wheelchair bound. Knee Xray and MRI were normal. Repeat CT TAP demonstrated pericardial thickening, perinephric fat stranding and subtle mural enhancement of his infra-renal aorta. He was commenced on methotrexate and oral steroid therapy with modest result for presumed Igg4 disease. He did not tolerate methotrexate. Leflunomide 20mg OD was added. Symptoms progressed and rituximab was given but he again relapsed. Due to ongoing knee pain a NM bone scan was performed which showed focally intense radiotracer uptake in the proximal left fibula. CT TAP was again repeated which demonstrated an increase in size of the periaortic inflammatory change, now amenable to biopsy.

Results
A retroperitoneal soft tissue biopsy revealed a histiocytic neoplasm with BRAF-V600E mutation consistent with Erdheim-Chester disease. He was started on first line treatment with interferon, however this failed and he was started on vemurafinib.

Conclusions
In patients that fail to respond or follow an atypical clinical course re-imaging and re-evaluation is of critical importance.

22S106

The Diagnostic Utility of Cardiac MRI in Lupus Pericarditis Missed by Echocardiogram

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Introduction
C-reactive protein (CRP) elevation may reflect chronic serositis in SLE patients. Pericarditis is one of the common cardiac manifestations of SLE and the standard imaging modality is transthoracic echocardiogram (TTE); however, diagnosing pericarditis can be more challenging in some cases.

Aims/Background
A 47-year-old overweight female known to have SLE (on maintenance hydroxychloroquine) presented to the ED with substernal pleuritic chest pain associated with positional variation, dyspnoea and significant bradycardia (36 bpm). She was afebrile, and apart from bradycardia, vital signs and physical examination were unremarkable including normal heart sounds without murmur, pericardial rub or palpable synovitis.

Method
Blood analysis including renal function, haemoglobin, lymphocyte count, D-dimer, TFTs, C3/C4 and serial troponin were unremarkable except slightly elevated CRP of 15. CXR evidenced moderate cardiomegaly. Apart from significant bradycardia, serial ECG/telemetry didn't reveal any other abnormality. Septic screen including viral screen and SARS-CoV-2 was negative and urine dipstick negative for blood and protein. Based on her symptoms, signs and CXR finding, urgent TTE was sought; however, this (including pericardial assessment) surprisingly was reported normal.

Results
She remained symptomatic which prompted assessment with cardiac MRI (CMR). This revealed pericardial STIR enhancement at basal/right ventricle consistent with pericardial inflammation associated with moderate global concentric pericardial effusion without hemodynamic compromise. T2-mapping revealed focal increased signal in anterior anteroseptal wall, mid ventricle and atria. Her symptoms/CRP responded (though not fully) with high-dose prednisolone taper and colchicine but she wasn't able to be weaned them off completely. Repeat TTE (+/- CMR) is awaited with consideration of anakinra if pericarditis remains unresolved.

Conclusions
The initial normal TTE may be explained by patient's symptoms/elevated CRP preceding the evolution of pericarditis or fatty tissue obscuring the TTE results while imaging quality is operator-dependent. CMR is superior to TTE in its images and doesn't depend on patient factors (e.g., adiposity) or user-dependent factors (e.g., probe technique); however, may be limited due to its high cost and limited availability. This case highlights the importance of CMR as a valuable diagnostic tool in afebrile lupus patients without palpable synovitis who presents with elevated CRP, chest symptoms and/or cardiac rhythm abnormality, despite normal cardiac markers or TTE.

22S108

Dramatic Resolution of Diffuse Scleroderma Skin Disease Following Treatment with IV Tocilizumab for COVID-19 Infection

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Introduction
Use of Tocilizumab in systemic sclerosis (SSc) with progressive skin disease has been shown to stabilise SSc-associated interstitial lung disease but to date has not been shown to lead to a significant difference in modified Rodnan skin score (mRSS).
We present the case of a 57 year old lady with diffuse scleroderma and aggressive skin disease, with significant improvement in her modified Rodnan skin score (mRSS) after treatment with Tocilizumab for Covid-19 pneumonia.

Aims/Background
The patient presented in 2017 with sclerodactyly, Raynaud's, arthralgia and weight loss. Anti-SCL 70 antibody was negative. No evidence of pulmonary hypertension or interstitial lung disease to date. Her baseline mRSS was 23.
Initial treatment was with Cyclophosphamide 500mg/m2 4-weekly for 6 months and low dose steroid due to prominent flexor tendon involvement in the hands with marked pruritis. Unfortunately she progressed; mRSS increased to 44, with involvement extending to thighs and upper arms.

Method
The patient was subsequently treated with Rituximab and Mycophenolate Mofetil (MMF) 1.5g daily; some softening of skin was documented. Raynaud's symptoms remained problematic. Illoprost infusions were commenced 6 monthly. Her skin score was 27 after 3 cycles of Rituximab. A further cycle was planned but deferred due to the Covid-19 pandemic. The patient continued on MMF. She then received Cycle 5 and 1 dose of Cycle 6 – the patient declined 2nd dose due to concerns regarding covid. She received one further dose in July 2021 (Cycle 7).

Results
The patient tested positive for Covid-19 in September 2021. She presented with profound hypoxic respiratory failure requiring ventilation for 7 days. She was treated with dexamethasone and intravenous tocilizumab 8mg/kg.

Conclusions
Four months later at rheumatology review, her appearance had changed dramatically - softened, more flexible skin in her face, upper

arms, torso and thighs. Her mRSS score was 18 with only significant residual involvement remaining in her hands. Although not yet substantiated in scleroderma clinical trials, this case illustrates a significant improvement following IV tocilizumab in a patient with severe skin disease.

Figure

22S109

Ear Pain and Pre-Auricular swelling: An Unusual Presentation of Ultrasound Positive, Biopsy Positive Giant Cell Arteritis

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Introduction
Temporal Artery biopsy has been the gold standard for diagnosis of giant cell arteritis (GCA) for many years, with temporal artery ultrasound increasingly utilised due to high sensitivity and specificity with experienced sonographers.
We present a case of a 75 year old lady with treated hypertension and hypothyroidism, presenting with an atypical history and normal inflammatory markers but positive imaging and histology.

Aims/Background
The patient attended with a 2 week history of painful ears without hearing loss and associated bilateral pre-auricular discomfort unchanged when talking/masticating. She reported gradual onset bi-temporal/occipital headache. No history of visual symptoms, but the patient is congenitally blind in the right eye. No features of systemic upset other than fatigue; no features of polymyalgia rheumatica. Temporal arteries were pulsatile and non-tender symmetrically. Radial arteries pulsatile with no radio-radial delay. ESR 8, CRP 10.

Method
Given this presentation, no treatment was initiated. The patient was reviewed 3 weeks later, and felt better, with improved energy. She had ongoing intermittent bi-temporal/occipital headaches of reduced severity and a sensation of uncomfortable fullness in her ears; these symptoms were improving hearing was unchanged. Full blood count was normal, ESR 10 and CRP 6.

Results
Temporal artery ultrasound was requested due to the history of ongoing headaches; it was performed 8 weeks after presentation and exhibited characteristics consistent with GCA in the frontal branch of the superficial temporal artery bilaterally. Prednisolone 20mg daily was commenced.
Left temporal artery biopsy was performed 10 weeks after symptoms began and showed classical features of GCA. Prednisolone was increased to 40mg to protect vision in the functional left eye. At telephone review, her symptoms have completely resolved.

Conclusions
There are limited case reports of auricular involvement as a preceding or concurrent symptom of GCA, most commonly with sensorineural hearing loss. Treatment with glucocorticoid led to partial or full recovery. Ear pain is a rare but recognised symptom and in this case was the presenting and persistent feature of radiologically and biopsy confirmed GCA.

22S110

Overlap of Giant cell arteritis and small vessel vasculitis – A Case report

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Introduction
Giant cell arteritis and ANCA associated vasculitis (AAV) rarely present simultaneously. We present the case of a 74-year-old lady admitted following fracture of her right distal radius. Two weeks prior to admission, the patient had been commenced on prednisolone 10mg in the community for presumed polymyalgia rheumatica.

Aims/Background
On the fracture ward, she developed scalp tenderness, temporal headaches and intermittent blurring of vision. CRP 94, ESR 65, haemoglobin 85g/L, white cell count 10.0 x109/L, eosinophil levels were normal/low. Temporal Artery ultrasound showed a positive compression sign in the parietal branch of the right temporal artery with a halo thickness of 0.7mm.

Method
Biopsy of the right temporal artery confirmed arterial inflammation with leukocytoclastic ducts consistent with giant cell arteritis. The prednisolone dose was increased. A few days later she developed complete right foot drop and could not mobilise. MPO- and P-ANCA were strongly positive. CT chest, abdomen and pelvis showed no malignancy or other organ involvement. Extensive viral screening was negative. MRI brain was normal.
The patient was pulsed with intravenous methylprednisolone and discharged on reducing oral corticosteroid to return the following week for her first rituximab infusion. At review 2 weeks later her ankle dorsiflexion improved to grade 3/5 and she completed the cycle of rituximab. ESR 20 and CRP 1.

Results
A new diagnosis of giant cell arteritis may make it difficult to attribute developing symptoms to a further disease process. Although giant cell arteritis is most commonly associated with headaches, scalp tenderness and polymyalgia rheumatica symptoms, it can also cause many of the symptoms described in AAV including rarely mononeuritis multiplex, contributing to diagnostic confusion.

Conclusions
Hassane et al in 2018 reviewed eight case reports of patients presenting with both granulomatosis with polyangiitis (GPA) and giant cell arteritis. They warned of symptom overlap in both conditions leading to potential delays in diagnosis and treatment. In summary, vasculitis can cause multi-organ involvement but clinicians must be cognisant that albeit unusual, both large vessel and small vessel vasculitides can co-exist; as awareness is important in aiding treatment decisions.

22S111

"DADA tell you about this case ? " A rare case of monogenic auto-inflammatory disease in childhood

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Introduction
A 3 ½ years old boy presented with an 8 week history of a nodular tender rash, abdominal distention and generalized arthralgia and myalgia. This was followed with a 3 week history of intermittent fevers. Past medical history unremarkable but there was a maternal history of mesenteric panniculitis.
On examination the patient was reported to have multiple purplish macular lesions and palpable subcutaneous nodules, and a small effusion of left knee. Investigations showed mildly raised inflammatory markers with negative auto-antibodies and infectious screen. Skin biopsy undertaken locally reported nodular vasculitis versus erythema induratum .
He was transferred to Rheumatology at CHI Crumlin where extensive investigations including ECHO, bone marrow aspirate and biopsy, urine catecholamine were undertaken and normal. Inflammatory marker and ferritin were elevated , renal function, blood pressure and urinalysis normal. A repeat skin biopsy showed panniculitis (Polyartheritis nodosa vs nodular vasculitis). Abdominal and Doppler Ultrasound for splenic infarct, DSMA, CT Angiogram and stool FOB were all normal. Oral corticosteroids for suspected Polyarteritis nodosa were commenced and with good effect. On trial of corticosteroid wean he relapsed with fever, worsening rash, and malaise.
Molecular genetic tests were positive for heterozygous c.140G>C;p. Gly47Ala which is pathological for ADA2 . Subsequent serum levels of adenosine Deamninase 2 level came back to be very low (1.4). Deficiency of adenosine deaminase 2 (DADA2) diagnosis was confirmed. He was started on subcutaneous adalimumab with significant improvement and corticosteroids were discontinued.
DADA2 is a rare monogenic auto-inflammatory disease, due to mutations in ADA2 gene, resulting in broad spectrum manifestations including livedo reticularis, cutaneous vasculitis, ischemic stroke, intracranial haemorrhage, neuropathy, recurrent fevers, anaemia, neutropenia as well as Primary immune deficiency. Laboratory findings are non-specific but functional protein assay can detects low or absent ADA2 enzymatic activity and provide a rapid diagnosis while waiting for confirmatory genetic test. It can mimic PAN and should be considered in all forms of suspected systemic vasculitis. Currently main treatment are TNF inhibitors while HSCT is reserved for patients with immunodeficiency or cytopaenia .

22S112

A NEXUS of striking ocular and systemic inflammatory features. Clinical manifestations of a newly reported haemato-immune disease

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Introduction
We herein report a rare case of VEXAS in a 69 year old male presenting with orbital apex syndrome, chondritis, macrocytic anaemia, venous thrombosis, pyrexia of unknown origin and inflammatory rash. This was on a background of a fluctuating two year history of unexplained autoinflammatory symptoms.

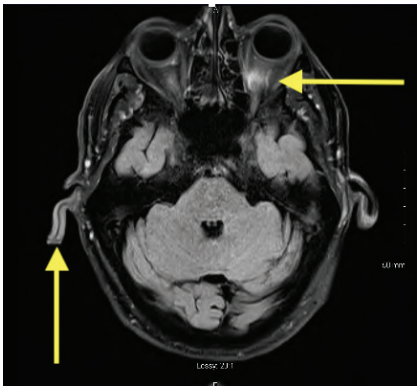
Aims/Background
VEXAS syndrome is a novel haemato-immune condition first reported in 2020. It is the result of an acquired somatic mutation affecting methionine-41 of the X-linked gene UBA1, responsible for ubiquitylation. It is characterised by vacuoles in myeloid and erythroid precursors, autoinflammatory features, almost exclusive male predominance and a median age of onset of sixty-four. Treatment options are limited, requiring high dose steroids, with early evidence suggesting the benefit of Tocilizumab and Janus-kinase inhibition. Severe cases require autologous haemopoetic stem cell transplantation. Mortality is as high as 25%. The objective of our case report is to highlight this recently documented condition and its unusual constellation of symptoms, including a novel ocular presentation, avoiding incorrect diagnosis.

Method
Investigations revealed raised ESR, CRP, Ferritin and D-dimer. Longstanding macrocytic anaemia was noted as was the presence of neutropenia, lymphopenia and decreased monocytes. Ultrasound of the right lower limb revealed superficial thrombophlebitis. The patient was noted to have left hypertropia and proptosis accompanied by pain on extraocular movement and facial pain. MRI brain confirmed left orbital apex inflammation and also chondritis of the ear. Punch biopsy of the diffuse popular rash was consistent with vasculitic process. Bone marrow demonstrated the presence of vacuoles and genetic testing has confirmed the presence of a mutation in UBA1. The patient was treated with pulsed IV methylprednisolone 500mg for three days, followed by oral prednisolone 40mg.

Results
Following steroid treatment the patient showed almost complete resolution of ocular symptoms, chondritis and rash within two weeks. He was discharged on oral prednisolone and despite initial improvement, suffered a relapse of symptoms within two months. The patient has since been commenced on tofacitinib.

Conclusions
This is a newly described condition with prevalence estimated between 1:20,000 and 1:30,000. VEXAS should now be considered as a differential in elderly male patients with macrocytic anaemia and systemic inflammation, including ocular presentations.

Figure



22S113

Multi-Organ IgG4-RD:a diagnostic dilemma for the Rheumatologists

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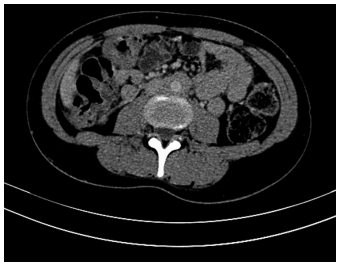
Introduction
IgG4 related disease is a rare systemic fibro inflammatory disease with or without elevated plasma IgG4 levels.While there are a few classic presentations documented,cardiac and CNS involvement are less often encountered in clinical practice.Ideally the diagnosis should rely on histopathological findings but not always possible in day-to-day practice.

Method
A previously healthy,37y old woman was admitted to the Hospital with 8 weeks history of abdominal pain and constitutional symptoms. A CT abdomen completed in the community showed a retroperitoneal mass near the aorta, suspicious of Lymphoma. Furthermore,a bulky pancreas and dilatation of the biliary tree was suspected.On examination,she had only mild proptosis and tender epigastrium. Laboratory findings included elevated CRP,ESR and predominantly cholestatic LFTs. Auto immune workup showed mildly positive ANA(1:80) and total IgG levels of 13.38. An MRCP revealed a non obstructive intra and extra hepatic duct dilatation and a PET-CT ruled out any other lymphadenopathy.She went on to develop headache and right partial third nerve palsy.MRI brain reported a dural thickening and hyperenhancement at the right temporal lobe inclining the differentials towards Tolosa Hunt Syndrome,lymphoma or hypertrophic meningitis secondary to IgG4 RD.Unfortunately none of the sites involved were amenable to biopsy.Clinically she met the 2019 ACR/EULAR classification criteria for IgG4 RD,so was trialled with steroids.After an initial positive clinical response,she was readmitted with an acute anterolateral myocardial infarction. Cardiac catheterization revealed 70 percent luminal stenosis in the first diagonal and a Cardiac MRI findings suggested infarction in the LAD territory but with a significant burden of myocardial edema and ischemia.This is most likely in keeping with cardiac manifestation of igG4-RD.

Results
Immunosuppression with Rituximab was started and showed excellent results with substantial reduction of para-aortic fibrosis and the dural thickening of the temporal lobe on follow-up scans at six weeks. At a recent followup,after two cycles of Rituximab,the patient is symptom free with complete resolution of the LFTs.

Conclusions
The diagnosis of IgG4 RD can be challenging.Coronary and CNS involvement is exceedingly rare and can cause serious complications. Keeping an open mind and looking for a unifying diagnosis will help institute early treatment with immunosuppressive agents and a favourable outcome.

Figure



22S114

Spondyloarthropathy And Sarcoid : Watch Out For This Rare Mix

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Introduction
Co-existence of Sarcoid and Spondyloarthropathy (SpA) is rare has been more reported as an adverse effect of anti-TNF alpha therapy than an association.We report a case of newly diagnosed pulmonary sarcoidosis discovered while investigating for an axial spondyloarthropathy naïve of anti-TNF α therapy.

Aims/Background
case report

Method
A 31 years old gentleman was referred to Rheumatology service with a history of inflammatory lower back pain and an hour long early morning stiffness for five years. He had no respiratory symptoms. There were no other medical comorbidities and the family history was significant for his father having Rheumatoid Arthritis. The examination revealed slightly limited lumber flexion.Laboratory assessment confirmed normal serum calcium,sedimentation rate and C-reactive protein. As a routine workup ,a dedicated MRI of the sacroiliac joints was performed which showed bilateral sacroiliitis but also raised a suspicion of abnormal architecture of the left iliac bone,querying an insufficiency fracture.He subsequently had an isotope bone scan which not only revealed abnormal tracer uptake in the left iliac crest but also reported multifocal uptake in the thoracolumbar spine. Meanwhile he was found to be HLA-B27 positive favouring the primary diagnosis of SpA.We proceeded with an ultrasound guided left iliac bone biopsy which reported the presence of non-caseating granulomas.Further workup showed an elevated serum ACE levels and a chest xray showed bilateral hilar lymphadenopathy. He fulfilled the ASAS criteria for the diagnosis of axial spondyloarthropathy and CT thorax confirmed Stage II pulmonary sarcoidosis.He had a normal echocardiogram,CT brain and pulmonary function tests . A co-occurrence of axial SpA and sarcoidosis was established.

Results
His musculoskeletal symptoms improved with combination of exercise,weight reduction and non-steroidal anti-inflammatory drugs. From a pulmonary perspective,he remains asymptomatic without any immunosuppression and is under care of specialist Respiratory Service in our Hospital.

Conclusions
Sarcoid and SpA may co-exist besides the paradoxical drug effect. With the increasing use of anti-TNF alpha drugs as the main stay of treatment for SpA,accurate diagnosis is very critical.Since our patient was asymptomatic from a pulmonary perspective,we recommend that all SpA patients undergo baseline chest xray and thorough investigation of their respiratory functions.

22S115

Fundus Fluorescein Angiography – A Valuable Adjunct to Diagnosis and Monitoring in Systemic Autoimmune Disease

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Introduction
Systemic autoimmune disease (SAD) is often associated with ophthalmic signs and symptoms. Ophthalmic symptoms of retinal vasculitis are often nonspecific. Fundus fluorescein angiography (FFA) can be a useful adjunct in objective assessment and aid in reaching a diagnosis or monitoring response to therapy. FFA features of retinal vasculitis and anterior ischemic optic neuropathy (AION) include retinal arterial leakage and sheathing, peripheral capillary drop out, reduced venous perfusion, patchy choroidal filling and increased arm to eye time. Here we present a case series illustrating the utility of FFA in monitoring and diagnosing a variety of SAD.

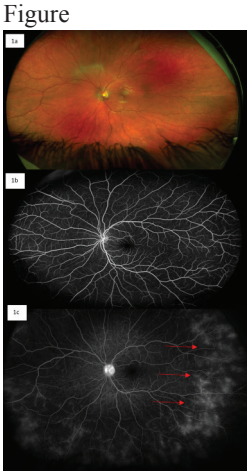
Aims/Background
We retrospectively describe a case series in which FFA was used in the diagnosis and / or monitoring of SAD with visual involvement. All patients were attending the Rheumatology and Ophthalmology department at the Mater Misericordiae University Hospital (MMUH), Dublin between May-October 2021. All data was anonymised, and patient consent was granted for their information to be presented and published as an abstract.

Method
We identified 4 patients with a variety of SAD in whom defining the nature and extent of visual symptoms and retinal vasculitis proved challenging or in whom FFA provided additional diagnostic utility.

Results
Case1
75-year-old female with biopsy proven giant cell arteritis (GCA) diagnosed in 2016 re-presented with recurrence of headache but no other signs or symptoms of active GCA. Inflammatory markers were normal. FFA showed an enlarged foveal avascular zone and patchy choroidal filling suggestive of imminent AION. Given the FFA findings, the patient was treated as a GCA relapse. Her corticosteroid dose was increased with subsequent resolution of her FFA finding.
Case 2
43-year-old female with SLE who presented with acute painless monocular visual loss. MRI brain showed non-specific white matter changes and lumbar puncture was bland. FFA demonstrated loss of capillary bed in the temporal periphery with adjacent abnormal vasculature. These findings were consistent with AION with bilateral posterior segment peripheral ischemia at the level of the capillary beds. FFA was repeated at 3 months following initiation of immunomodulatory therapy and showed no interval change, indicating stable disease.
Case 3
19-year-old male presented with a 4-day history of headache, bloodshot eyes, photophobia, and reduced vision in both eyes. CRP was 105mg/L and ESR 77mm/hr. He had bilateral acute anterior uveitis and was prescribed topical steroids. He complained of bilateral flashing field defects. His retinal exam was normal. FFA showed bilateral occlusion of his capillary beds in the peripheral temporal retina. The FFA confirmed a panuveitis. He subsequently developed inflammatory lower back pain and was diagnosed with axial spondyloarthropathy and commenced on anti-TNF therapy.
Case 4
49-year-old female presented with gradual onset inferior quadrantanopia in the setting of a weak positive MPO-ANCA and no other clinical or laboratory features of systemic vasculitis. Retinal exam revealed inferiorly swollen optic discs. The working differential diagnosis was AION secondary to MPO-ANCA vasculitis vs non-arteritic ischaemic optic neuropathy (NAION) secondary to anatomical anomaly of the optic discs. Monitoring via FFA has shown resolution of disc leakage following steroid treatment inferring an AION and commencement of systemic immunosuppression

Conclusions
These cases illustrate the potential utility of FFA in the diagnosis and monitoring of patients with SAD and ocular involvement. Ophthalmic symptoms of SAD are often mild, and patients find them difficult to describe. FFA allows detection and monitoring of subclinical or overt manifestations of SAD. FFA also allows delineation of the type and level of vascular bed affected. Further research is needed to validate the utility of FFA in diagnosing and monitoring SAD with eye involvement.

Figure



22S116

Dilemma of Polymyalgia or Polymyositis

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Introduction
The idiopathic inflammatory myopathies are a heterogeneous group of conditions characterized by varied patterns of inflammation within striated muscles and evidence of autoimmune mediated muscle breakdown. Its characteristic presenting features are symmetrical proximal muscle weakness developing over weeks or months associated with elevated muscle enzymes, typical muscle biopsy findings, presence of autoantibodies and radiological changes. However, yet the dilemma continues with diagnosing and management.

Aims/Background
To highlight an unusual presentation which puzzled making a final diagnosis.
Sixty-eight years old gentleman, presented to Emergency department with 3 days history of sudden onset of pain in his bilateral shoulders and legs. He had history of myocardial infarction, hypothyroidism, right carpel tunnel decompression. Few weeks ago, he had an

uneventful robotic prostate surgery. Before hospital presentation, his general practitioner commenced him on oral steroid keeping in mind PMR, but 2 days later his symptoms got worse. His examination was unremarkable with no evidence of lymphadenopathy or rash, power in upper limbs was 5/5 and lower limbs 4/5 (limited due to pain). His initial bloods showed CRP of 312, and ESR 69, and negative blood cultures. Primary team started him on broad spectrum antibiotics with no response in inflammatory markers. Extensive investigations with, muscle enzymes, CT-TAP, MRI whole spine and thighs, PET CT scan, muscle biopsy to rule out any inflammatory, infection or malignancy were inconclusive. He had excellent clinical and serological response to high dose of steroids. However, he had recurrent flares with tapering steroids. Lately his extensive myositis panel, processed at St James's hospital, showed presence of PL-7 antibodies. He was treated with pulsed intravenous methylprednisolone and commenced on methotrexate with excellent response. He was also started on Rituximab infusion as maintence therapy. Currently he is in Clinical remission.

Results
Myositis can have atypical presentation with symptoms overlapping with polymyalgia.

Conclusions
Inflammatory myositis can present in variable ways with normal result of some of specific investigations characteristic for this condition. However, presence of raised inflammatory markers along with myositis specific antibodies as in our case, and response to targeted treatment can be helpful in establishing a final diagnosis.

22S117

Sore hands are a pain in the neck

Author(s)
Dr. Patrick Mulkerrin, Professor Paul O'Connell

Department(s)/Institutions
Beaumont Hospital

Introduction
MMcN is a 67 year old lady who was admitted with progressive widespread pain and weakness.
She had a background of scoliosis, addison's secondary to exogenous steroid use. She had received a diagnosis of rheumatoid arthritis based on hand deformities and pain.
Her regular medications included butran's patch, pregabalin, paracetamol, hydrocortisone.
She lived alone and was finding it progressively difficult to transfer, mobilize and follow out daily activities.
One of her primary complaints was hand pain and loss of function.
A rheumatology consult was sought regarding poorly controlled rheumatoid arthritis.
On examination there were very obvious deformities(PIC 1&2). Note the fixed flexion deformities at 3rd digit. There is also a pronounced thenar and hypothenar eminence muscle wasting. However when the individual joints were examined there was no swelling or tenderness that would be associated with rheumatoid arthritis.
Routine bloods including FBC, U&E, LFT, CRP, ESR were normal. Rheumatoid factor and anti-CCP were negative as was the connective tissue disease screen.
She had previously been diagnosed with spinal deformities and an MRI C-Spine was sought. The rationale was that any signal deficit in that region would subsequently lead to deficits in power, sensation and function in the hand region.
MRI C-Spine was revealing in that there was marked kyphotic abnormality with marked displacement of C4 posteriorly on C3 and C5 posteriorly on C6 with compression of the spinal canal and cord compression.
A neurosurgical opinion was sought and corrective cervical surgery was performed at a later date.
The deformities and wasting in hands could be explained by the cervical spine deformities.
She is currently being transferred to neuro rehab for further therapy. She also has ongoing pain issues. There is no role for rheumatology medications in her care.
The learning point from this case would be that wasting and deformities had led to doctor's presuming that she had poorly controlled rheumatoid arthritis when in fact it was due to neuromuscular disruption which then corresponded to deformities.

Conclusions
Images to follow I have to return to my laptop apologies

22S118

Pause on the steroids and go back to the history

Author(s)
Sharon Cowley, Robert McEvoy, Sinead Harney, Arthur Jackson

Department(s)/Institutions
Cork University Hospital, Cork

Introduction
A 68 year old gentleman presented to the emergency department of our institution with progressive blurring of vision.

Aims/Background
One month earlier he had attended an audiologist as he noticed his hearing had decreased. Subsequent audiogram revealed bilateral sensorineural hearing loss. His GP had completed bloodwork including ESR and CRP which were elevated at 99mm/hr and 160mg/l respectively. Vasculitis was suspected and high dose IV methylprednisolone was commenced.

Method
On further questioning, the visual symptoms were ongoing for 2 months, but he had seen an ophthalmologist elsewhere who had reassured him. Subsequently a detailed history revealed he endorsed MSM relationships.

Results
Lumbar puncture performed to rule out infectious causes. Syphilis antibodies came back positive with elevated rapid plasma regain (RPR) titres and elevated Treponema pallidum antibodies (TPA) and neurosyphillis was confirmed. NVRL Syphilis IgM was also positive. MRI brain was unremarkable apart from chronic left sided mastoiditis. A second ophthalmologist at CUH confirmed mild uveitis and vitritis and commenced topical steroids.

Conclusions
He started IV benzylpenicillin however his liver function tests became deranged and he transitioned to IV ceftriaxone to compete the treatment course. He has since fully recovered. A full STI screen and contact tracing was done.
The key learning point is that a detailed history including sexual history is critically important if there is an unexplained syndrome.

22S119

First do no harm and always dip the urine

Author(s)
Sharon Cowley, Eoghan Whyte, Siun O 'Flynn, Eva Long, Sinead Harney

Department(s)/Institutions
Cork University Hospital, Cork

Introduction
A 77 year old gentleman was referred to our outpatients with an ESR of 99mm/hr to investigate for possible underlying rheumatological disease.

Aims/Background
He had a past medical history of hypertension, osteoarthritis, right hip replacement, renal calculi, atrophic left kidney, COPD, abdominal aortic aneurysm of 5cm, empyema with VATS 2020, carpal tunnel syndrome and diastolic dysfunction on echocardiogram.
Medications included bisoprolol 2.5mg OD, Pantoprazole 40mg OD, Aspirin 75mg OD, Folic acid 5mg OD, Viagra 100mg PRN, Olmesartan/amlodipine 40/10mg, trelegy inhaler 92/55/22mcg OD and atorvastatin 20mg.

Method
The patient reported feeling well and denied early morning stiffness, arthralgia, rash, fever, weight loss, headaches, visual disturbance, jaw claudication, chest pain, shortness of breath, nosebleeds, sinusitis or loss of hearing. He also examined normally.
Extensive work up was completed which showed ESR 90-140 over the last 18 months, normochromic normocytic anaemia with Hb of 10.9g/dl. Eosinophils were marginally elevated at 0.43x 10^9/l. Creatinine was 128umol/l, urea of 12.2mmol/l, albumin was low at 28g/l, cholesterol elevated at 9.3mmol/l and c-reactive protein of 6.9mg/l. Autoimmune screen was negative, IgG4 subclasses were normal, SPEP showed raised alpha 2 globulin with a monoclonal IgG kappa band, bence jones were negative, as was quantiferon.
Skeletal survey was negative for lytic lesions. CT TAP showed no evidence of malignancy with known 5cm abdominal aortic aneurysm. PET scan revealed no source for elevated inflammatory markers. He was not treated with steroids empirically.

Results
Urine dipstick was 3+ positive for protein. Urinary protein creatinine ratio was elevated at 745.8mg/mmol (0-45). He was referred to the renal service and kidney biopsy was performed. This showed numerous sub-epithelial and intramembranous electron dense, immune complex-type deposits consistent with membranous glomerulonephritis. Anti-PLA2R levels were checked and were very elevated at >800. Primary membranous nephropathy was diagnosed and patient was commenced on treatment with rituximab.

Conclusions
This case highlights the importance checking a urine dipstick. This was on of the most important findings in a patient with extensive work up.

22S120

Upper airways destruction with ANCA positivity – don't forget the drug history!

Author(s)
Salamat Ullah; Dr Elizabeth Ball, Dr Claire Riddell

Department(s)/Institutions
Rheumatology Department ,Musgrave Park Hospital, Belfast.

Introduction
A 39 year old gentleman was referred to ENT with a 'hole in his nose', 6 months after a reported local skin injury while welding. On ENT assessment, he had a 1cm defect from skin to nasal cavity just above the left alar crease with extensive crusting and inflammatory granulation throughout. On nasal scoping he was noted to have total destruction of nasal septum.

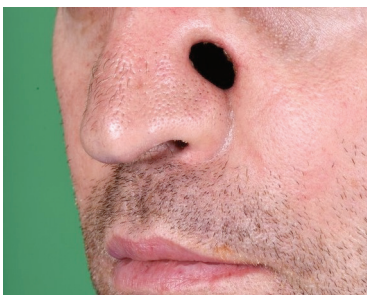
Aims/Background
Histopathology was non-diagnostic. Crusting improved over 2 weeks with local antibiotic therapy. Screening bloods revealed a strongly positive cANCA (160) and PR3 (117 iu/mL (NR 0-4.9). At ENT review he was noted to have an oroantral fistula into the maxillary sinus at the site of a previous dental extraction of his left upper molar. At this stage he was referred to rheumatology for assessment of a possible ANCA vasculitis and was assessed at our Rapid Access Service. On questioning, he admitted regular cocaine use over the past year. No other features of systemic vasculitis were evident. Inflammatory markers were modestly raised – CRP 28 mg/L (NR <5) and ESR 26 mm/hr (NR 1-12)
Repeat nasal biopsy was performed and no definite vasculitis features were seen. It was reported as in keeping with cocaine use.
MRI scanning (Fig.2) revealed a large mid-face cavity with extensive nasal septum, turbinate and medial wall of maxillary sinus destruction. There was a naso-cutaneous and oro-antral fistula with mild diffuse mucosal thickening and enhancement.

Method
This patient was advised to avoid further cocaine and was given a short course of prednisolone due to persistent inflammation. He responded well and was referred to plastic surgery for consideration of reconstruction.

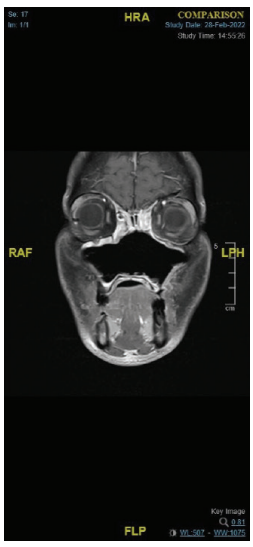
Results
Cocaine use has been associated with:
1. Cocaine induced midline destructive lesions (CIMDL) mimicking granulomatosis polyangiittis (GPA) findings in the upper airways and 2. an ANCA+ vasculitis attributed to levamisole (a contaminant in >80% cocaine) which may present with cutaneous findings, arthralgia, nasopharyngeal involvement and agranulocytosis(1,2) Antibodies to human neutrophil elastase (HNE) ANCA may help distinguish.(3). Cutaneous lesions remit after discontinuation of cocaine, but recur with rechallenge(2).

Conclusions
Key points:
- GPA may be difficult to distinguish from CIMDL
- Full history taking should include recreational drug use.

Figure

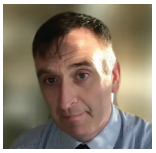


Figure



SpR Training Day - Virtual Meeting

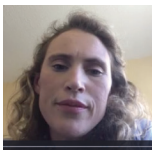
Thursday, 21 October 2021
(Sponsored by Novartis Ireland Ltd)



09.00-09.15 **Introduction -**
Dr John Ryan NSD,
Consultant Rheumatologist, CUH.

09.15-10.00 **Myositis**
Dr Lorraine O'Neill,
Consultant Rheumatologist, SVUH.

10.00-10.45 **Difficult to Treat R.A.**
Dr Claire Sheehy,
Consultant Rheumatologist, UHW.



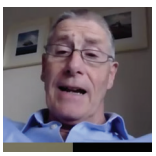
11.00- 11.45 **Lupus**
Dr Grainne Murphy,
Consultant Rheumatologist, CUH.

11.45-12.30 **Bone Health**
Dr Bryan Whelan,
Consultant Rheumatologist, Manorhamilton

12.30-13.15 **Ankylosing Spondylitis**
Dr Barry O'Shea,
Consultant Rheumatologist, SJH.

14.00-14.45 **Rare and Wonderful**
Prof Gaye Cunnane,
Consultant Rheumatologist, SJH.

14.45-15.30 **Crystal Arthritis**
Dr John Stack,
Consultant Rheumatologist, MMUH.



15.30-16.15 **Psoriatic Arthritis**
Prof Doug Veale,
Consultant Rheumatologist, SVUH.

16.15 **Final Comments - Dr John Ryan**

ISR Meeting
Spring, 2022 Sponsors

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22S121

Gardner Diamond Syndrome in a young woman with Complex Regional Pain Syndrome

Author(s)
Cillian Hurst Claire Sheehy

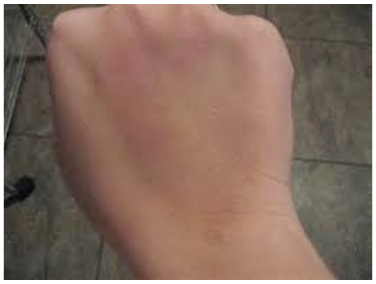
Department(s)/Institutions
Rheumatology Dept, University Hospital Waterford

Method
Case Report:
GON is a 16 year old female referred by the orthopaedic team, for assessment of a painful bruised right hand and wrist. She had suffered a direct blow from a hurley one year earlier. No fracture was seen on Xray at the time.
Upon review she reported pain and swelling in her right hand, with no history of new trauma. There was abnormal tingling and hyperaesthesia. Given the history of prior trauma and these symptoms she was diagnosed with complex regional pain syndrome by orthopaedics. MRI and ultrasound of the area showed only soft tissue oedema. The area was splinted and she was referred to OT and rheumatology.
When seen in rheumatology, she was currently an in-patient in another facility for management of anorexia nervosa. She reported episodes of spontaneous bruising on the dorsum of her right hand every 6-8 weeks and associated with swelling (see image 1). Episodes lasted a few days at a time.
Clinical examination was otherwise unremarkable. The patient and her mother were educated about CRPS and she was advised to continue sertraline which she was taking for her eating disorder. Three months after this initial visit the patient was still experiencing the episodes. A diagnosis of Gardner Diamond Syndrome was given. At follow up nine months after her initial visit to rheumatology the patient was back in school. She continued to take Sertraline 150mg daily with no further episodes of bruising, swelling or pain of her right hand in the preceding 4 months.

Results
Discussion:
Gardner Diamond Syndrome (GDS) is also known as “psychogenic purpura”, and “auto-erythrocyte sensitisation”.
The young patient initially presented with almost classical CRPS to the orthopaedic clinic. Her recurrent episodes of self limiting, spontaneous bruising, swelling and pain are atypical in this.
GDS occurs in patients with a variety of psychiatric disorders.

Conclusions
Learning Points:
Gardner Diamond syndrome is a rare, unrecognised disorder, which should be considered in the presence of unexplained bruising.
Early recognition of this disorder enables initiation of appropriate management, involving multiple specialities and limits unnecessary investigations.
Treatment of underlying psychiatric conditions is paramount.

Figure



22S122

Atypical Kawasaki Disease in an Adult following the SARS-CoV-2 Vaccine

Author(s)
Clodagh Flanagan, Elizabeth Keeling, Genevieve Kelly, Catherine Hughes

Department(s)/Institutions
Rheumatology Department, Dermatology Department, Tallaght University Hospital

Introduction
We report the first case of atypical KD following the Pfizer-BioNTech mRNA vaccine. There has been one case of atypical KD following the SARS-CoV-2 vaccine, and this was with the AstraZeneca Vaccine, Vaxzevria. (1.)

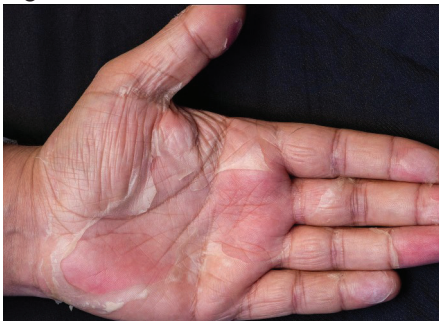
Aims/Background
Kawasaki Disease (KD) is an acute small to medium vessel vasculitis that characteristically presents in children between the ages of 6 months to 5 years. Less than 60 adult patients have been described worldwide who met the the American College of Rheumatology’s diagnostic criteria. (1.,2.,3.) There has been one case of atypical KD following the SARS-CoV-2 vaccine, and this was with the AstraZeneca Vaccine, Vaxzevria. (1.)

Method
We describe a further case of Kawasaki disease presenting in an otherwise well 43-year-old man. The gentleman presented to an Irish hospital with a 3-day history of fever, rash, myalgia, headache, vomiting and diarrhoea. Past medical history was non-contributory. Three weeks prior to this acute presentation he had received the second Pfizer-BioNTech mRNA vaccine as part of the national immunization programme.

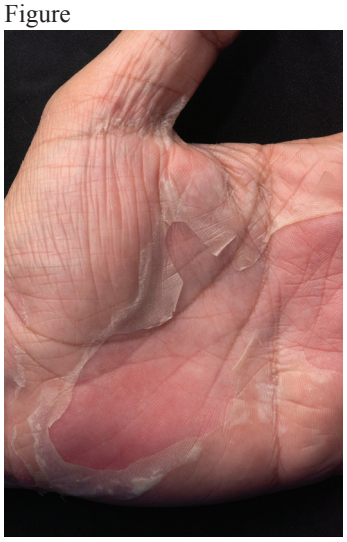
Results
On Examination, bilateral conjunctival injection alongside dry, swollen, cracked lips, and oedematous hands and feet were noted, meeting the criteria for Kawasaki Disease. (2.) Laboratory investigations revealed: C reactive protein 183 mg/L, white cell count 15.3×109/L (1.8–7.5) and Lactate 2.1 mmol/L. The rash commenced on his upper trunk before becoming erythrodermic and desquamated within 24 hours. His symptoms resolved after treatment with Intravenous Immunoglobulin (IVIG), Aspirin and Methylprednisolone

Conclusions
The diagnosis of KD is rare in adults, and atypical symptoms are uncommonly observed. KD following vaccination is a diagnosis of exclusion and only a temporal relationship is observed in this case. A single dose of IVIG 2g/kg, Aspirin, and high dose intravenous corticosteroids for 3days prior to transitioning to tapering steroids helped the rapid recovery of our patient. However, further research is required to understand the treatment options for KD following the SARS-CoV-2 vaccination.

Figure



Figure



22S123

Large Vessel Vasculitis Presenting as Fever after Chemotherapy

Author(s)
Osama Alalwan(1), Emma Carroll(2), William Mullally(2), Gregory Leonard(2), Bernadette Lynch(1)

Department(s)/Institutions
Department of Rheumatology, Galway University Hospital, Ireland(1) Department of Oncology, Galway University Hospital, Ireland(2)

There are many reported cases in the literature of vasculitis as a paraneoplastic phenomenon. Granulocyte-Colony Stimulating Factor (G-CSF) is widely used for the recovery of chemotherapy associated neutropenia. We describe a challenging case of (G-CSF) induced large vessel vasculitis (LVV). Identifying the correct underlying cause of vasculitis can have a huge impact on the management of such cases.

A 79-year-old gentleman, who was diagnosed with metastatic colorectal cancer, received his first cycle of chemotherapy FOLFOX (Oxaliplatin, leucovorin and fluorouracil) together with G-CSF on 18th of July 2021. After 10 days, he presented to our hospital again with a history of high-grade fever for one week. Initial workup was negative for an infective cause.

He was admitted with a presumptive diagnosis of infection, given his recent immune suppression. He continued to have a fever and high inflammatory markers despite multiple antibiotic courses. All cultures were negative. A wide differential diagnosis was considered including chemotherapy induced, infection, autoimmune disease and progression of his known underlying cancer. A Pan-CT was done, and it showed circumferential smooth thickening of the aortic arch wall, which was not present on his recent staging CT scans. Following review of his clinical course, pharmacological history, blood results and CT scans, the patient was diagnosed with G-CSF associated LVV. The patient had an excellent response to steroids with direct resolution of fever and subsequent normalization of inflammatory markers and CT findings.

Method

There are several cases in the literature with a similar characteristic presentation. The key feature is the development of rapid progressive thickening of the aortic wall within days after receiving G-CSF. Stopping the offending agent (G-CSF) can be sufficient for resolution of the condition. Many cases, including our case, required short-term steroids to facilitate rapid resolution.

Conclusion

G-CSF induced LVV should be kept in the differential diagnosis of patients presenting with fever without infectious focus after chemotherapy. Identifying the correct cause of LVV might avoid unnecessary antibiotics and long-term medications.

22S124

Granulomatosis with Polyangiitis presenting with cervico-thoracic myelopathy.

Author(s)
Patricia Harkins; Robert Field; Richard Conway

Department(s)/Institutions
St. James Hospital, Dublin

Introduction

Case

We present the case of a 64 year old male presenting with a 6 week history of upper back pain, gradual deterioration in his gait, with lower limb paraesthesia and weakness. Past Medical Hx was significant for recurrent sinusitis, epistaxis requiring multiple cauterisations and prior right sided mastoidectomy.

Neurological examination revealed a spastic paraparesis with sustained clonus, hyperreflexia and MRC grade 3/5 weakness in lower limbs. Sensory examination was consistent with a mid thoracic sensory level.

Aims/Background

Investigations

C-Reactive protein (43mg/L) and erythrocyte sedimentation rate (80mm/hr) were elevated. Full blood count, renal and liver indices were normal.

Gadolinium enhanced MRI spine revealed a circumferential mass like lesion extending from C5-T9 with compression of the spinal cord at T2-T3. MRI brain was normal. CT – thorax abdomen and pelvis was also normal.

Urinalysis and urinary protein: creatine ratio were also normal.

ANCA revealed a strongly positive perinuclear pattern, with a markedly elevated myeloperoxidase (MPO) antibody 117 (0-10 units) Proteinase 3 (PR3) was negative.

Method

Management

With evidence of spinal cord compromise, the patient went for urgent neurosurgical evaluation and intervention. Direct visualisation of the lesion revealed a solid, mass like structure, of which samples of tissue were excised and sent for pathological analysis. This revealed a dense lymphocytic infiltrate, with fibrinoid necrosis of the blood vessel wall, consistent with vasculitis. Of note, there was no evidence of granuloma formation.

A diagnosis of MPO positive, Granulomatosis with polyangiitis, causing compressive myelopathy was made.

Results

High dose glucocorticoids, tapered as per the PEXIVAS reduced dose regimen was commenced in tandem with pulsed intravenous cyclophosphamide. Within days of treatment initiation his neurological status improved, and he is currently engaging in rehabilitation. At his most recent review he was walking independently with crutches.

Conclusion

Our case highlights a rare cause of myelopathy but one that should enter the differential diagnosis when assessing a patient presenting with myelopathy, particularly with a mass-like lesion on neuroimaging. It should be treated aggressively and as part of the life threatening organ involvement algorithm.

3 cases of Antiphospholipid Syndrome (APLS) in systemic sclerosis (SSc)

AProf Muriel Soden, Townsville University Hospital, Qld 4814, Australia

Introduction
APLS is well recognized in association with Systemic Lupus Erythematosus (SLE). The laboratory markers include Lupus Anticoagulant (LAC), Anticardiolipin antibody (aCL) and Beta-2 glycoprotein-1 (B2GP1).
I present here 3 cases of APLS in association with SSc. This is the first report of this association.

CASE 1
SS 35 yo female, Polish Australian, Coal grader.
Presents with severe Raynaud's phenomenon and digital infarcts in 2013.
Smokes 25 cigarettes daily
2 teenage children. No pregnancy issues.
Managed with Calcium channel blockers; advised to stop smoking.
Needed to give up her work due to cold exposure
In 2015 presents with severe obtundation.
Admitted through A&E:
BP 70/40, ashen, GCS 10

LABS: Hb 3 g/dL, Plt 60; Creat 400, Urea 45
LAC +; aCL IgM and IgG pos; anti-B2GP1 pos

DIAGNOSIS: Catastrophic APLS
Admitted to ICU; fluid rescuscitation, dialysis/hemofiltration, IV heparin infusion, plasmaphoresis, cyclophosphamide pulses, transfusion.
Excellent response and was discharged 1 month later
Continue oral warfarin, azathioprine, calcium channel blockers, doxycycline for calcinosis
Has continued to smoke!

CASE 2
JK, 28 YO female, restaurant worker, German descent
24/40 miscarriage on 2nd pregnancy.
Screening for APLS showed triple positive LAC, aCL and B2GP1
Commenced on SC heparin. 3 further successful pregnancies.
Referred Rheum OPD in 2014 with severe Raynaud's phenomenon.
Clinical exam: sclerodactyly, calcinosis cutis, oral telangiectasia.

LABS: ANA 1/2560, Centromere +; U1RNP +
Diagnosis: Limited systemic sclerosis with APLS.
2016 presents with GM seizures.
MRI brain NAD; LP lymphocytosis 50/HPF and raised protein levels.

DIAGNOSIS: APLS cerebritis. Rx Azathioprine and low dose prednisolone (weaned).
Excellent outcome with no neurological relapse.

CASE 3.
MG, 46 yo female, Phillipina; home maker
Raynaud's phenomenon for many years.
Clinically sclerodactyly and cakcinosis.
Chronic mitral regurgitation with secondary pulmonary hypertension.
Chronic thrombocytopenia on bloods without ecchymoses or bleeds.

LABS: ANA 1/2560 H/S; ENA U1RNP+, Sm+, dsDNA +

DIAGNOSIS: MCTD with SSC phenotype

APLS screen as planned MVR surgery: triple positive LAC, ACLA and B2GP1
Commenced on warfarin with view to do surgery at 3 month mark.
6 weeks later presents with brainstem stroke.

LABS: Plts 60, APTT twice normal, Creat 350, Urea 35

DIAGNOSIS: Catastrophic APLS
Admitted ICU
Dialysis, plasmaphoresis, IV heparin, Cyclophopshamide, Rituximab
Despite all these measured patient succumbed.

CONCLUSION:
It is generally held that APLS is NOT a clinical entity in association with SSC. While LAC, aCL and B2GP1 have been detected in patients with SSC there are no literature reports of clinical cases such as those I present here.
Clinicians need to be aware of the association – albeit rare – so that timely interventions can be implemented.

ACR - SpR Training Day - Virtual Meeting

Monday, 13 December 2021
(Sponsored by AbbVie)

A Virtual ACR Highlights Meeting, attended by 46 Trainees and Consultants took place on Monday, 13 December 2021, generously sponsored by AbbVie Limited.

The Meeting was chaired by Dr John Ryan, Consultant Rheumatologist, CUH and the following Agenda was presented.

18.55	**Introduction -** Dr John Ryan, Consultant Rheumatologist, CUH.
19.00-19.20	**Connective Tissue Disease related Interstitial Lung Disease** **Dr Wan Lin Ng.** Rheumatology SpR, Beaumont Hospital, Dublin.
	Moderated Q&A
19.25-19.45	**New ACR/EULAR classification criteria for Vasculitis** **Dr Imran Ali,** Rheumatology SpR, Beaumont Hospital, Dublin.
	Moderated Q&A
19.50-20.10	**8M118 curbside consults: controversies in Bahcet's syndrome management** **Dr Fatemah Al Faraj,** Rheumatology Fellow, CUH.
	Moderated Q&A
20.15-20.35	**Clinical Breakthroughs from ACR** **Dr Conall MacGearailt,** Rheumatology SpR, SVUH.
	Moderated Q&A
20.40-21.10	Guest Speaker **Navigating Research Opportunities in 2021** **Dr Richard Conway,** Consultant Rheumatologist, UCD.
21.15	**Final Comments - Dr John Ryan.**

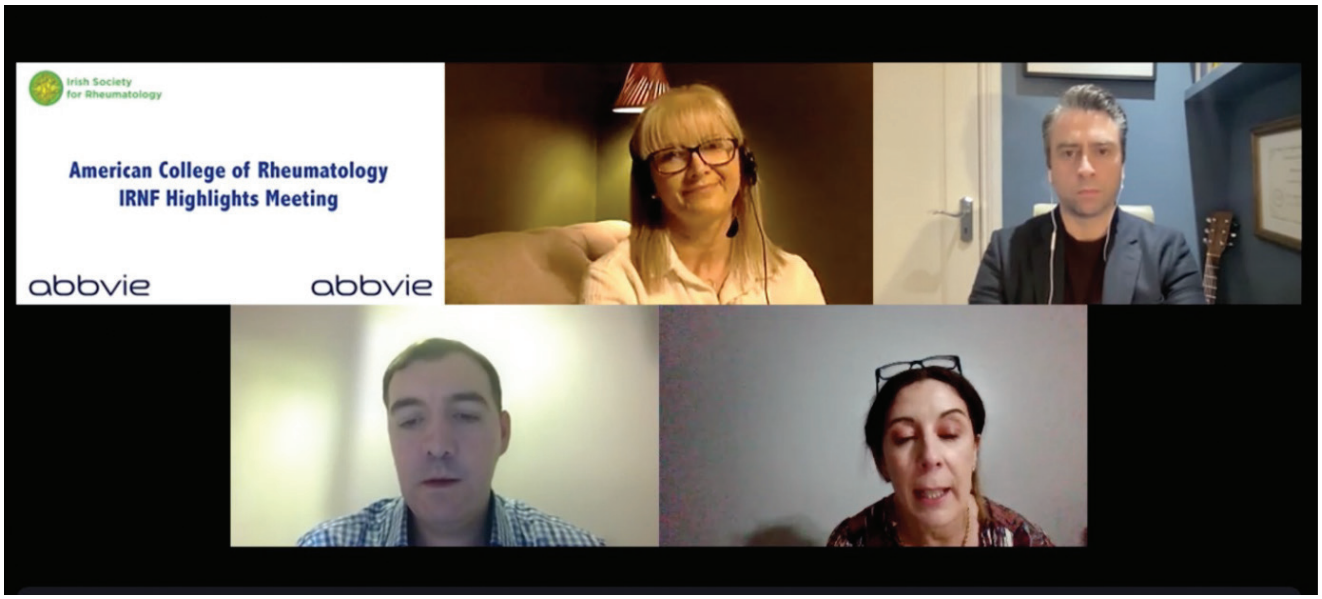
American College of Rheumatology IRNF Highlights Meeting

The ISR organised American College of Rheumatology (ACR) Highlights Meeting will take place this year on 03 February 2022 and will be chaired by Ms Ann Maria Curran. The objective of this meeting is to educate and update attending nurses regarding current issues in Rheumatology. The presentations will be prepared by attending nurses listed below:

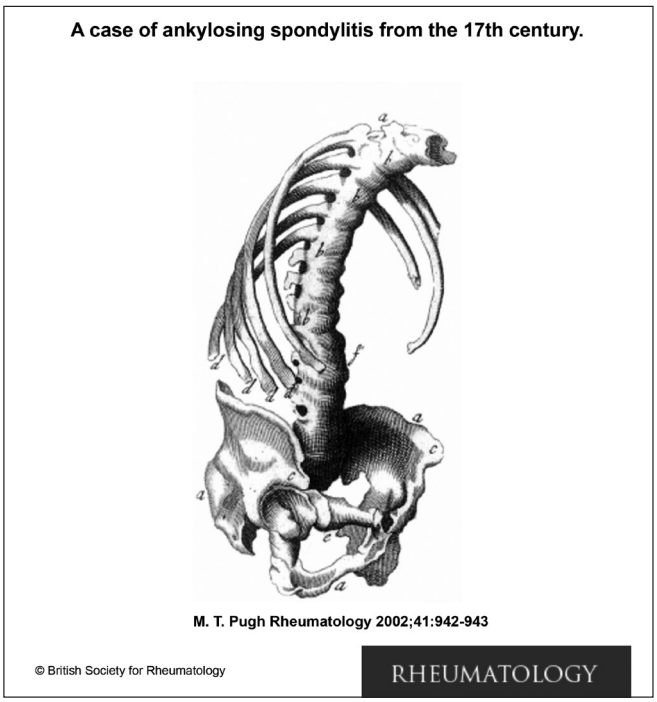
Date: Thursday, 03 February 2022
Venue: Virtual – Commencing at 19.00
Sponsored by: AbbVie Ltd

Presentations on the night will include:

19.00–19.05	**Ann Maria Curran,** RANP **Welcome and Introduction**
19.05-19.25	**Mary Gillespie,** RANP, UL Hospitals Group, Nenagh, Co Tipperary ***"A glance at topics looking at patient reported outcomes"***
19.25-19.45	**Derek Deely,** RANP (Rheumatology), OLCHC ***"Social Media for Paediatric Rheumatology Care and Research"***
19.45-20.05	**Ann Maria Curran,** RANP, Mayo University Hospital ***"The Balancing Act of Managing Spondyloarthropathies"***
20.05-20.25	**Dr Richard Hogan,** Clinical Director, Therapy Institute ***"21 Day Challenge"***
20.25-20.30	**Ann Maria Curran – *Round up, Thanks and Conclusion***



ISR Bernard Connor Medal


A case of ankylosing spondylitis from the 17th century.

M. T. Pugh Rheumatology 2002;41:942-943

RHEUMATOLOGY

The Irish Society for Rheumatology (ISR) has established the Bernard Connor Medal to promote interest in the field of rheumatology among BST trainees in medicine, *and to support engagement with the Irish Society for Rheumatology.*

The award is open to BST trainees in medicine who *fulfil the eligibility criteria below*. In addition to receiving the Connor Medal, the winner will be invited to attend the annual scientific meeting of the ISR to present their work to the membership, as a guest of the society. *Additionally, and at the discretion of the judging panel, up to two runners-up may be awarded full registration to attend the ISR annual scientific meeting.*

Bernard Connor

The Connor Medal is named in honour of Bernard Connor, an Irish physician who observed and described the characteristic skeletal and clinical features of ankylosing spondylitis in 1693, while a medical student in Paris. This award will be made annually on the basis of competitive submission.

Theme

The theme chosen by the ISR is a **Case Report with relevance to rheumatology**
The submitted work should highlight your observations/insight in relation to rheumatology as a BST trainee.

Submission Only one submission per trainee will be accepted.

Case Report

The case report should be submitted in full form and present the details of an interesting case followed by a discussion on your observations of the key points of interest, and relevant literature review. These should be submitted in full [**max 800 words**, concluding with summary key message and references (up to 5 in total)].

Bernard Connor Medal contd…

Eligibility

- Applicants must be registered RCPI BST trainees by 1 July 2022.
- Applicants must submit completed entries to the ISR by the 12 August 2022.
- It is hoped that the re-launch of this competition in its new format will not only honor a great clinician but also draw the specialty of Rheumatology to the attention of BST trainees. The ultimate aim is to attract more trainees to HST in Rheumatology.

How to Apply

Application forms for the Connor Medal will be available for download from the ISR website www.isr.ie. These must be completed in full and returned together with your submission to info@isr.ie

Closing Date: 12 August 2022.

Judging Criteria

The Medal will be awarded according to the criteria below which will be applied to all submissions

- Relevance of the submitted work to rheumatology
- Originality and Merit of the work

The Board of the Irish Society for Rheumatology shall convene the judging panel. The decision of the panel shall be final and no correspondence on submissions or final outcome can be entered into.

RABRI PROJECT

The Irish Society for Rheumatology (ISR) / Rheumatoid Arthritis Biologics Register of Ireland (RABRI) is seeking to recruit a Clinical Research Coordinator (CRO) on a fixed term contract for one year with a possible extension for a second year.

Reporting to ISR Management and nominated study leads

The CRO will be responsible for coordinating research activities for the registries/ biobanking in Rheumatology in the participating centres across Ireland. This post would suit individuals who have the ability to work independently and possess a high level of organizational skills. Previous experience in Rheumatology is not essential but would be an advantage, however experience in clinical research is essential. The successful candidate will be expected to adhere to the current standard operating procedures for clinical trials and academic research and have current ICH-GCP certification.

The successful candidate will be responsible for the overall running of the registries to include the following duties:

1. Assist sites in ethics submissions and arranging study contracts, and submissions to the Data Protection Office (DPO).
2. Act as liaison between the research site personnel and the chief investigators to ensure smooth running of the registries.
3. Create site files and provide biosample plasticware and other supplies as indicated by the relevant project.
4. Provide training to site staff on protocol and data entry to electronic data base.
5. Perform monitoring and support to sites to ensure research is performed to the highest standard.
6. Prepare and submit reimbursement invoices with sites within the agreed timelines.
7. To play an active role in encouraging sites to achieve recruitment targets across the clinical research registries.
8. Assist and support researchers in the day-to-day collection of data for written /oral publications.
9. Maintain accurate records of performance metrics for the ISR, prepare reports and make presentations as agreed by steering committee (6 monthly/annual reports).
10. Support the general activities including arranging steering committee meetings, updating and reporting to steering committees.
11. Monitor database activity and ensure data entry /queries are kept up to date.
12. Prepare 6 monthly reports on reported adverse events specific to individual pharmaceuticals supporting the individual registries.

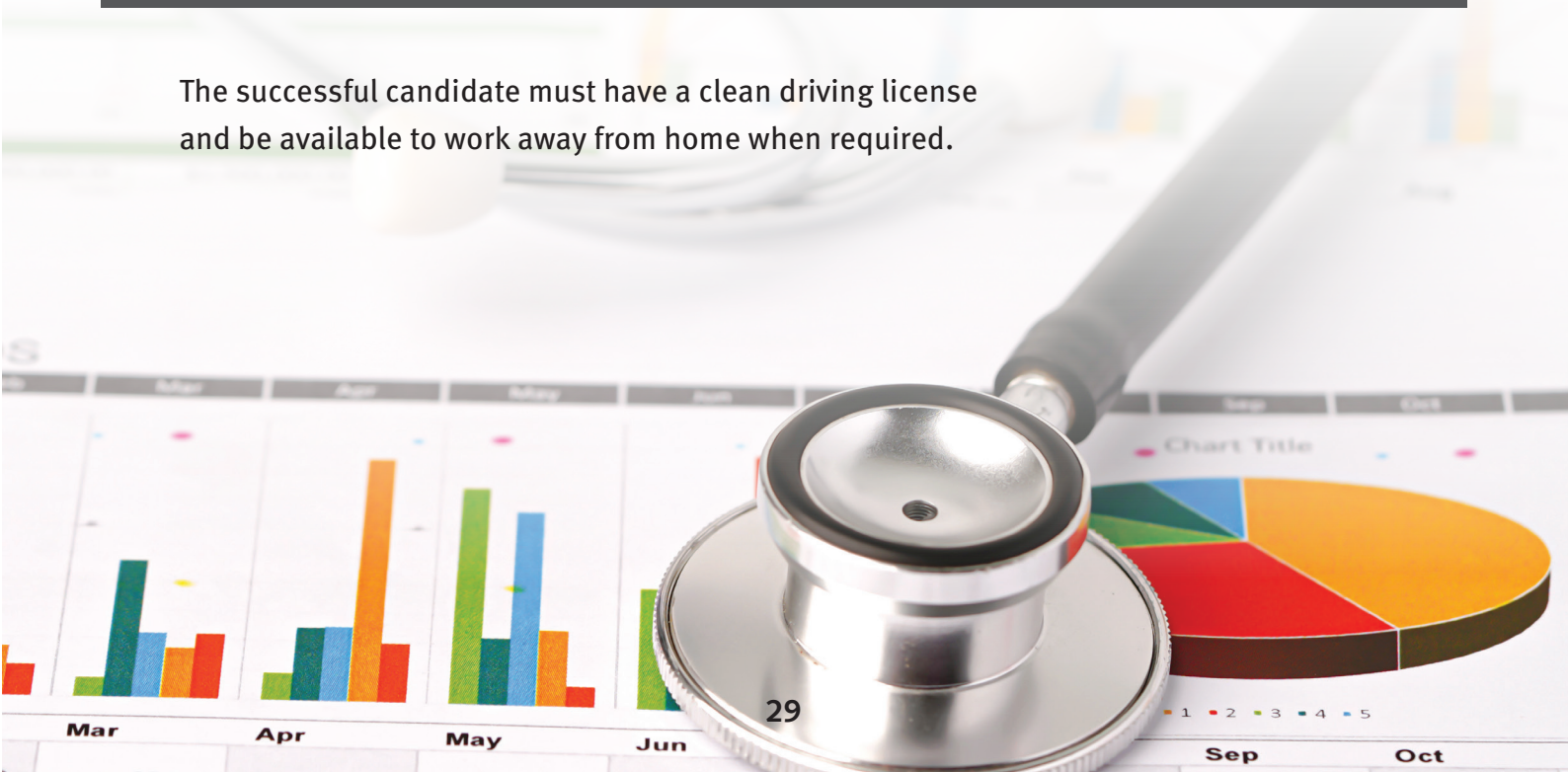
WORKER VACANCY

Desirable skills /Essential Skills

- BSc or equivalent third level qualification (E)
- Knowledge of medical/scientific terminology (E)
- Excellent attention to detail (E)
- Excellent oral and written communication skills. (E)
- Excellent IT skills and knowledge of electronic data capture systems (E)
- Ability to work on own initiative (E)
- Understanding of Good Clinical Practice (GCP) and SOP's. (E)
- Project manager experience (D)
- Familiarity with the conduct of clinical trials (D)
- Previous involvement in clinical/translational studies e.g. 3 yrs (D)
- Candidates should demonstrate an awareness of equality, diversity, and inclusion agenda (D)

Apply with cv to info@isr.ie
by Tuesday, 31 May 2022

The successful candidate must have a clean driving license and be available to work away from home when required.

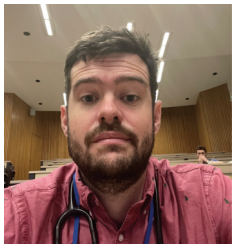


SpR Training Day
New Therapeutics in Psoriatic Arthritis

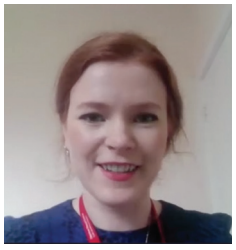
Wednesday, 6 October 2021 (Virtual Meeting)
– Chaired by Dr John Ryan, Consultant Rheumatologist, CUH
(Sponsored by AbbVie)

14.30 **Introduction and Emphasis on the increase of the disease.**
Dr John Ryan, Consultant Rheumatologist, CUH.

14.45 **New Therapeutics in Psoriatic Arthritis**
Dr Dennis McGonagle, Prof of Investigative Rheumatology, University of Leeds, UK.



15.30 **Psoriatic Arthritis Significant Case 1**
Dr Patrick Mulkerrin
Rheumatology Sp R.



15.50 **Psoriatic Arthritis Significant Case 2**
Dr Sharon Cowley
Rheumatology Sp R.

16.10 Q & A

16.30 Wrap up and thanks Dr John Ryan.