



**LupusAcademy**

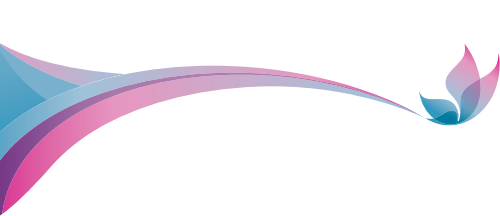
Communicate. Educate. Treat.

Lupus Academy Middle East Summit Conference

# Abstract Book

Crowne Plaza Hotel,  
Dubai, United Arab Emirates  
**9–10<sup>th</sup> December 2016**





## European Accreditation Council for Continuing Medical Education (EACCME) Accreditation



The Lupus Academy/European CME Forum is accredited by the European Accreditation Council for Continuing Medical Education (EACCME) to provide the following CME activity for medical specialists. The 'Lupus Academy Middle East Summit Conference 2016' is designated for a maximum of 6 hours of European external CME credits, event code 14844. Each medical specialist should claim only those hours of credit that he/she actually spent in the educational activity. The EACCME is an institution of the European Union of Medical Specialists (UEMS), [www.uems.net](http://www.uems.net).

### Participants from Europe

EACCME is an institution of the European Union of Medical Specialists (UEMS) and operates by reciprocal agreement for recognising CME credits across Europe. More information can be found at [www.uems.net](http://www.uems.net).

### Participants from USA

Through an agreement between the European Union of Medical Specialists and the American Medical Association, physicians may convert EACCME credits to an equivalent number of AMA PRA Category 1 Credits™. Information on the process to convert EACCME credits to AMA credits can be found at [www.ama-assn.org/go/internationalcme](http://www.ama-assn.org/go/internationalcme).

### Participants from Canada

Live educational activities, occurring outside of Canada, recognized by the UEMS-EACCME for ECMEC credits are deemed to be Accredited Group Learning Activities (Section 1) as defined by the Maintenance of Certification Program of The Royal College of Physicians and Surgeons of Canada.

### Participants from Other Countries

CME accreditation by EACCME and ECMECs are recognised internationally by many national authorities across the globe. Please check with your local authority to confirm its validity for your purposes.

### Meeting Organisation

The content for this activity has been developed under the control of the meeting Chairmen Professor Munther Khamashta London UK/Dubai UAE and Dr Jamal Al-Saleh Dubai UAE, on behalf of the Steering Committee of the Lupus Academy. No supporting companies have had any influence over the presentation of any aspects of this meeting. For information about financial and in-kind support received to assist Lupus Academy in the delivery of its educational programme, please visit the website [www.lupus-academy.org](http://www.lupus-academy.org). CME compliance, accreditation and fulfilment has been facilitated by European CME Forum, on behalf of the Lupus Academy.



# Welcome

## Dear Friends and Colleagues,

We are delighted to welcome you to the Lupus Academy<sup>†</sup> Middle East Summit Conference, the first dedicated lupus meeting to be held in the Gulf Cooperation Council region. Developed in collaboration with Dubai Hospital, this meeting will address key challenges and developments in current clinical practice through a series of presentations and interactive workshops. We sincerely hope that you will enjoy and benefit from what we believe is one of the most informative and interactive learning programmes you will participate in this year.

Now in its fifth year, the Lupus Academy is a global initiative that continues to strengthen its commitment to providing high quality, insightful and clinically relevant education both through interactive meetings and eLearning. With this, we aim to support you as you strive to provide best-in-class patient care and improve patient outcomes in lupus.

This meeting, which has been accredited by the European Accreditation Council for CME (EACCME), aims to provide latest insights into advances in global research and clinical practice in lupus and allied diseases. Delegate feedback from our previous annual meetings continues to guide us in selecting the topics and speakers you need to ensure translation of treatment advances into your clinical practice.

The scientific component of this programme, developed by our Steering Committee of 12 international experts in lupus, is designed to create a highly interactive forum through which we can all develop a logical approach to the management of lupus worldwide.

This meeting will give you the opportunity to meet like-minded clinicians and scientists and, through the sharing of clinical experience, develop your knowledge in this fast moving therapeutic area.

We sincerely hope that this meeting will provide you with new ideas for your clinical work, enriched enthusiasm for collaborative research, and fruitful discussions with your colleagues who care for patients with lupus.

We look forward to meeting and talking with you here in Dubai.

With kind regards,

**Professor Munther Khamashta**

Course Director and Meeting co-Chairman

**Dr Jamal Al-Saleh**

Meeting co-Chairman

and

The Lupus Academy Steering Committee

**Professor Zahir Amoura**

**Professor Richard Furie**

**Professor Roger Levy**

**Professor Ricard Cervera**

**Professor Bevra Hahn**

**Professor Sandra Navarra**

**Professor Andrea Doria**

**Professor David Isenberg**

**Professor Murray Urowitz**

**Professor Thomas Dörner**

**Professor Munther Khamashta**

**Professor Ronald van Vollenhoven**

<sup>†</sup>The Lupus Academy is a long-term initiative dedicated to improving patient outcomes in SLE through an interactive educational forum dedicated to sharing best clinical practice through the dissemination and discussion of clinical and basic scientific research about SLE and allied diseases.



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# Programme

Friday 9 <sup>th</sup> December 2016			Page
14:30	<b>Opening Address</b>	Jamal Al-Saleh (UAE) & Munther Khamashta (UK/UAE)	
<b>Plenary Session I</b>			<i>Moderators: Jamal Al-Saleh (UAE) &amp; Munther Khamashta (UK/UAE)</i>
14:45	Biomarkers in SLE: How useful are they?	David Isenberg (UK)	18
15:10	Guidelines: Use of steroids and hydroxychloroquine in SLE	Guillermo Ruiz-Irastorza (Spain)	20
15:35	The challenge of APS	Graham Hughes (UK)	22
16:00	Discussion		
<b>Curbside Consults</b>			<i>Moderator: Jamal Al-Saleh (UAE)</i>
16:15	The experts tackle complex cases including lupus nephritis and lupus arthritis	Munther Khamashta (UK/UAE) David Isenberg (UK) Ian Bruce (UK) Liz Lightstone (UK)	24
17:00	<b>Break</b>		
<b>Four Parallel Workshops Focusing on Best Clinical Practice for Managing Lupus Manifestations</b>			
17:30	<i>Moderator: Jamal Al-Saleh (UAE)</i> Workshop: 1 — Investigating the febrile lupus patient	 Guillermo Ruiz-Irastorza (Spain) & Zoubida Tazi Mezalek (Morocco)	36
	<i>Moderator: Ricard Cervera (Spain)</i> Workshop: 2 — Haematologic challenges: Cytopaenias	 Ahmed Al-Shaikh (Saudi Arabia) & Ala Al-Heresh (Jordan)	40
	<i>Moderator: Munther Khamashta (UK/UAE)</i> Workshop: 3 — Pregnancy and lupus	 Samar Al-Emadi (Qatar) & Humaid Al-Wahshi (Oman)	44
	<i>Moderator: Graham Hughes (UK)</i> Workshop: 4 — CNS and lupus	 Imad Uthman (Lebanon) & Khaled El-Hadidi (Egypt)	46
19:00	Close (Day 1)		
20:00	<b>Welcome Dinner</b>		
Saturday 10 <sup>th</sup> December 2016			Page
<b>Four Parallel Workshops Focusing on Best Clinical Practice for Managing Lupus Manifestations</b>			
09:00	<i>Moderator: Jamal Al-Saleh (UAE)</i> Workshop: 1 — Investigating the febrile lupus patient	 Guillermo Ruiz-Irastorza (Spain) & Zoubida Tazi Mezalek (Morocco)	36
	<i>Moderator: Ricard Cervera (Spain)</i> Workshop: 2 — Haematologic challenges: Cytopaenias	 Ahmed Al-Shaikh (Saudi Arabia) & Ala Al-Heresh (Jordan)	40
	<i>Moderator: Munther Khamashta (UK/UAE)</i> Workshop: 3 — Pregnancy and lupus	 Samar Al-Emadi (Qatar) & Humaid Al-Wahshi (Oman)	44
	<i>Moderator: Graham Hughes (UK)</i> Workshop: 4 — CNS and lupus	 Imad Uthman (Lebanon) & Khaled El-Hadidi (Egypt)	46
10:30	<b>Break</b>		
<b>Plenary Session II</b>			<i>Moderators: Ricard Cervera (Spain) &amp; Munther Khamashta (UK/UAE)</i>
11:00	Clinical manifestations and evaluation of cutaneous lupus	Annegret Kuhn (Germany)	26
11:25	Decreasing morbidity and mortality and improving outcomes in SLE	Ian Bruce (UK)	28
11:50	Lupus nephritis: Update on modern management	Liz Lightstone (UK)	30
12:15	Treat-to-target in SLE	Andrea Doria (Italy)	32
12:40	Discussion		
13:00	<b>Closing Address</b>	Jamal Al-Saleh (UAE) & Munther Khamashta (UK/UAE)	

## Biographies



### **Dr Samar Al-Emadi, MBBS, FRCPC, FACR, ABIM**

Hamad Medical Corporation, Doha, Qatar

Samar Al-Emadi has been Senior Consultant in Rheumatology at the Hamad Medical Corporation (HMC) since 2005, and is an Assistant Professor at Weill Cornell Medical School in Qatar.

Samar was certified in internal medicine and rheumatology at the Royal College of Physicians and Surgeons of Canada (2003) and the American board of Internal Medicine and Rheumatology (2004). She is currently the Chief of Rheumatology at Hamad Medical Corporation and the Programme Director of fellowship training in rheumatology.

Dr Al-Emadi established a pregnancy and rheumatic disease clinic in 2005. She is a well-known speaker having participated in local and regional conferences and is a member of multiple local and regional committees, including HMC's Governance & Strategy of Research Committee, HMC's Institutional Research Board and the Steering Committee for OPD development. Samar has several publications in peer reviewed journals and is involved in multiple randomised controlled trials a Principal Investigator and co-Principal Investigator.

#### **Disclosures**

Grants/Research

Support:

Abbvie; Pfizer

Consultant/Advisor:

Abbvie; Pfizer; Roche; UCB

Meeting Honorarium/

Expenses:

Abbvie; Pfizer; Roche; UCB



### **Dr Ala' Al-Heresh, MD, FRCP**

Dr Sulaiman Al-Habib Medical Group, Riyadh, Saudi Arabia

Ala' Al-Heresh is a Senior Consultant Rheumatologist and, since 1996, Founder and Head of the Rheumatology Unit, King Hussein Medical centre, Jordan. He is also a Clinical Associate Professor at Jordan University Medical College.

He did his training in internal medicine at the Kent and Canterbury Hospital, UK and in 1993 continued his fellowship training in rheumatology at the Royal National Hospital for Rheumatic Diseases, Bath, UK. He was awarded his MRCP (UK) in 1992 FRCP (London) in 2003. He is currently an international adviser for the Royal College of Physicians of London.

Dr Al-Heresh is a member of many professional national and international bodies, he is President of Jordanian Rheumatism Society and member of the Higher Scientific Committee of the Jordan Medical Council. He is an Examiner at the department of internal medicine at the Royal College of Surgeons of Glasgow and Examiner at the Jordanian Board of Rheumatology and Internal Medicine.

Dr Al-Heresh was the Editor in Chief of the *Medical Journal of the Royal Medical Services* (Jordan), reviewer of several medical journals, and has published many articles in peer reviewed journals. His main interests include vasculitis and connective tissue diseases. He currently works at the Dr Suleiman Al-Habib Medical Group in Riyadh, Saudi Arabia.

#### **Disclosures**

None



**Dr Jamal Al-Saleh, MD, FRCP**  
Dubai Hospital, Dubai, UAE

#### Disclosures

None

Jamal Al-Saleh is a UK-trained Rheumatologist in practice for 21 years. Currently, he is the Head of the Rheumatology Section and Director of Medical affairs at Dubai Hospital.

Dr Al-Saleh completed his undergraduate training at the Royal College of Surgeons in Ireland, graduating with honours in medicine and surgery in 1995. He obtained his membership of the Royal College of Physicians in the UK in 1999 and was awarded a Masters in Clinical Rheumatology from the University of Manchester in 2003. He was awarded his FRCP in London and Edinburgh in 2009 and 2010 respectively.

Dr Al-Saleh is an Examiner at the Royal College of Physicians and an International Advisor for the Royal College of Physicians (London) for the UAE. He is a Senior Lecturer at the Dubai Medical

School and is actively involved in undergraduate and postgraduate training. He is a Chairman and a member of several committees in the Dubai Health Authority, past-Chairman of the Emirates Society of Rheumatology and past-Chairman of Emirates Society of Osteoporosis.

Dr Al-Saleh has conducted several research projects on the prevalence and clinical and immunological features of lupus patients in Dubai including; Dubai Hospital Lupus Cohort and PRODUBAI, which researched the prevalence of rheumatic diseases and osteoporosis in Dubai. He has built the first Arthritis Registry in the Middle East, which gathers data on six diseases. His current interests include systemic lupus erythematosus, early rheumatoid arthritis, seronegative spondyloarthritis and osteoporosis.



**Dr Ahmed Al-Shaikh, MD**  
King Faisal Specialist Hospital & Research Centre, Riyadh, Saudi Arabia

#### Disclosures

None

Ahmed Al-Shaikh is a Consultant Rheumatologist and Section Head of Rheumatology in the Department of Medicine at King Faisal Specialist Hospital and Research Centre (KFSHRC), Riyadh, Saudi Arabia. He is also an active member of several departmental and hospital committees. Currently, he is the Chairman of Scientific Committee of Rheumatology Fellowship in the Saudi Commission for Health Specialties.

In 1985, Dr Al-Shaikh obtained his MBBS degree from King Saud University, Riyadh. He completed his residency in internal medicine at KFSHRC in 1991; clinical fellowship in rheumatology at the Rheumatic Disease Unit in University of Ottawa,

Canada in 1993; and research fellowship at the University of Texas, Southwestern Medical Center, Dallas, Texas, USA in 1994.

He is a regular Examiner for the Saudi Board and Arab Board of Internal Medicine, as well as the Saudi Board of Rheumatology.

He has conducted several research projects, is author and co-author of several peer-reviewed papers, and has presented numerous invited lectures on a national and international level. His main interests include systemic lupus erythematosus and vasculitis.



## Biographies



### **Dr Humaid Al-Wahshi, MD, FRCP**

Royal Hospital, Muscat, Oman

#### **Disclosures**

None

Humaid Al-Wahshi is a Senior Consultant Internist, Rheumatologist and Immunologist at the Royal Hospital, Muscat, Oman. He obtained his medical degree from Sultan Qaboos University, in Oman in 1997. He was awarded his MRCP (UK) certificate in 2004, completed his rheumatology training at the University of Toronto, Canada in 2008 and was awarded his FRCP (London) in 2011. He did his training in obstetric medicine at Guy's and St Thomas' Hospitals, UK in 2014 and immunology at the Royal London Hospital in 2015.

Dr Al-Wahshi is a Senior Clinical Lecturer at the College of Medicine, Sultan Qaboos University, Oman. He is involved in undergraduate and postgraduate training and is an official Trainer and Examiner for the Oman Medical Specialties Board. He was a member of the Internal Medicine

Scientific Committee and Assistant Programme Director. He is the President of the Oman Rheumatology Society and a member and fellow of many regional and international professional societies. Dr Al-Wahshi has authored several publications in rheumatology and immunology.

Dr Al-Wahshi established the first combined rheumatology/obstetric clinic in Oman in 2016 and is establishing a cohort for pregnant patients with rheumatic diseases. He continues to treat patients with rheumatic diseases, particularly systemic lupus erythematosus and antiphospholipid syndrome (APS). Dr Al-Wahshi is currently working on establishing a local research forum for SLE and APS as well as establishing the rheumatology fellowship programme in Oman.



### **Professor Ian Bruce, MD, FRCP**

University of Manchester, UK

#### **Disclosures**

##### **Grants/Research**

##### **Support:**

Genzyme/Sanofi; GSK; Roche; UCB

##### **Consultant/Advisor:**

BMS, GSK, Eli Lilly, Roche, Medimmune, UCB

##### **Speaker's Bureau:**

BMS, GSK, Pfizer, Roche, UCB

Ian Bruce is a National Institute of Health Research (NIHR) Senior Investigator and Professor of Rheumatology at the Arthritis Research UK Centre for Epidemiology, Centre for Musculoskeletal Research, University of Manchester. He is Director of The NIHR Manchester Musculoskeletal Biomedical Research Unit and Deputy Director of the NIHR/Wellcome Trust Clinical Research Facility.

Professor Bruce qualified in medicine from Queen's University Belfast in 1988 and gained his MRCP in 1991. He trained in medicine and rheumatology in Northern Ireland and completed his MD thesis on the pathogenesis of systemic vasculitis in 1995. He was the Geoff Carr Lupus Fellow at the University of Toronto with Murray Urowitz and Dafna Gladman (1996–1998) before moving to Manchester in 1998.

Professor Bruce is the immediate past-Chair of the Systemic Lupus International Collaborating Clinics (SLICC), is a member of the British Isles Lupus Assessment Group, and leads the BILAG Biologics Register. He is also Chief Investigator of the MASTERPLANS consortium, a Medical Research Council-funded stratified medicine consortium focusing on systemic lupus erythematosus (SLE). He also serves on Data Safety Monitoring Committees in several commercial and academic clinical trials. Professor Bruce's research is focused on the association between inflammatory rheumatic diseases and premature atherosclerosis/coronary heart disease as well as stratified medicine in SLE.





**Professor Ricard Cervera, MD, PhD, FRCP**  
Hospital Clinic, Barcelona, Catalonia, Spain

#### Disclosures

Consultant/Advisor:  
GSK

*Professor Cervera is a member of the Lupus Academy Steering Committee.*

Ricard Cervera is co-Founder and Head of the Department of Autoimmune Diseases at Hospital Clinic, Barcelona. He is also leader of the Research Team on Systemic Autoimmune Diseases at the Institut d'Investigacions Biomèdiques August Pi i Sunyer (IDIBAPS) and Professor at the University of Barcelona where he coordinates the Masters Course on Autoimmune Diseases. He qualified in medicine in 1983 from the University of Barcelona and received his PhD in 1988 for his thesis on anticardiolipin antibodies. He spent 2 years at the Lupus Research Unit at The Rayne Institute, St Thomas' Hospital, London.

Professor Cervera is an Associate Editor of the journal *Lupus Science & Medicine* and is on the Editorial Boards of 20 medical journals. He is coordinator of the European Forum on Antiphospholipid Antibodies and of the European Working Party on Systemic Lupus Erythematosus (SLE) (Euro-Lupus Group). He is Chairman of the Medical Advisory Board of the Catalan Association of Lupus Patients and Medical Advisor to Lupus

Europe. He chaired the 6<sup>th</sup> and 8<sup>th</sup> International Congresses on Autoimmunity, the 1<sup>st</sup> and 2<sup>nd</sup> Latin-American Congresses on Autoimmunity, the 5<sup>th</sup> Meeting of the European Forum on Antiphospholipid Antibodies and the 8<sup>th</sup> European Lupus Congress.

Professor Cervera's research interests include clinical and epidemiological aspects of systemic autoimmune diseases, particularly SLE and antiphospholipid syndrome, with special focus on its 'catastrophic' variant. He has presented over 300 invited lectures and published more than 800 scientific papers (H-factor, 67), including original articles in the *New England Journal of Medicine*, *The Lancet*, *Annals of Rheumatic Diseases*, *Arthritis & Rheumatism*, *American Journal of Medicine* and *Medicine* (Baltimore). He is co-Editor of 25 books, including 'The Antiphospholipid Syndrome', 'Vascular Manifestations of Systemic Autoimmune Diseases' and 'Diagnostic Criteria in Autoimmune Diseases'.

## Biographies



### Professor Andrea Doria, MD

University of Padova, Italy

#### Disclosures

Consultant/Advisor:  
GSK

*Professor Doria is a member of the Lupus Academy Steering Committee.*

Andrea Doria is Professor of Rheumatology and Head of the Unit of Connective Tissue Disease and Rare Rheumatic Diseases, Division of Rheumatology, Department of Medicine, University of Padua, Italy.

Professor Doria received his medical degree and qualification in Rheumatology from the University of Padua. He was Council member of the Italian College of Rheumatology (CRO) between 1999 and 2005 and a Council member of the Italian Society of Rheumatology (SIR) from 2007 to 2010 and from 2013 until now. He is also a member of American College of Rheumatology.

Professor Doria has organised over ten international conferences on autoimmunity and was involved as “expert” in the European League Against Rheumatism (EULAR) Standing Committee for the development of clinical and therapeutic recommendations: (1) EULAR recommendations for the management of systemic lupus erythematosus (SLE)—Assessment of the SLE patient (2008–2009); (2) EULAR recommendations for the management of SLE Part II—Neuropsychiatric disease (2008–2009); (3) Joint EULAR and European Renal Association-European Dialysis and Transplant Association (EULAR/ERA-EDTA) recommendations for the management of adult and paediatric lupus nephritis (2012). Professor Doria is a member of the Lupus Academy Steering Committee and co-Chaired the 4<sup>th</sup> Annual Meeting held in Rome 27<sup>th</sup> February to 1<sup>st</sup> March 2015. He was the chair of the 10<sup>th</sup> European Lupus Meeting, held in Venice (Italy) 5–8<sup>th</sup> October 2016.

Professor Doria is on the Editorial Boards of several rheumatology and immunology journals, including *Lupus*, *Autoimmunity*, *Clinical and Experimental Rheumatology*, *Autoimmunity Reviews*, *Journal of Autoimmunity*, *Experimental Biology and Medicine*, *Rheumatology Reports*, *Journal Autoimmunity Highlights* and *Reumatismo* (the official journal of Italian Society of Rheumatology).

He has authored over 250 ISI publications on SLE and other connective tissue diseases. These include clinical studies describing new manifestations or subgroups of autoimmune disorders, prognostic risk factors, diagnostic tests and therapeutic interventions, as well as immunochemical studies that evaluate autoantibodies, epitopes and complementary epitopes of autoantigens. In addition, he has authored and co-authored three books, over 90 book chapters and conference proceedings, and over 500 abstracts for national and international conferences.

Professor Doria has long-standing experience of the clinical management of patients with connective tissue diseases. The Unit in which he works is a tertiary referral rheumatology centre, within Italy, for the diagnosis and management of patients affected with systemic connective diseases. In addition, he has expertise in the management and follow-up of pregnant patients with systemic rheumatic diseases. Professor Doria has also trained over 30 students in rheumatology.



**Professor Khaled El-Hadidi, MD**  
Cairo University, Egypt

#### Disclosures

None

Khaled El-Hadidi is Professor of Rheumatology at Cairo University and vice-President of the Egyptian Society of Rheumatology.

In the early 1980s he worked on his MD thesis titled “Anti-cardiolipin antibodies in 100 Egyptian lupus patients”. He took blood samples from lupus patients to the UK, where he performed tests at St Thomas’ Hospital under the supervision of Professor Graham Hughes and the team who discovered antiphospholipid syndrome. In 1990, after graduating from Kasr Al Ainy, Khaled spent 2 years training in Manchester, UK.

Professor El-Hadidi’s private clinic in Cairo includes a very dedicated and talented group of junior rheumatologists and he is blessed with the presence of Professor Tahsin El Hadidi, the former President of the International League Against Rheumatism and the godfather of rheumatologists in Egypt.

Professor El-Hadidi is currently finalising a large retrospective study, describing the clinical manifestations of around 1500 Egyptian lupus patients.



**Professor Graham Hughes, MD, FRCP**  
The London Lupus Centre, London, UK

#### Disclosures

None

Graham Hughes, often known as “the father of lupus” in the UK, is Head of the London Lupus Centre at London Bridge Hospital, UK. He trained at The London Hospital, after which he spent 2 years in New York working on the introduction of the DNA-binding test, under the leadership of Dr Charles Christian. In 1971, he opened the Lupus Clinic at Hammersmith Hospital, moving to St Thomas’ Hospital where he set up the Louise Coote Lupus Unit, a specialist unit dealing uniquely with lupus and related diseases. He instituted an annual postgraduate meeting “Ten Topics in Rheumatology”, now in its 28<sup>th</sup> year, with satellite “Ten Topics” meetings in six countries. In 1983,

he described the antiphospholipid syndrome, now known as Hughes Syndrome, and in 1991 was awarded the International League Against Rheumatism (world research) prize for this work. Other honours include Doctor Honoris Causa in the Universities of Marseille and Barcelona, and ‘Master’ of the American College of Rheumatology.

Professor Hughes is Founder and Editor of the international journal *Lupus*. He is mostly known for his work with patients, and is Life President of the patients’ charity Lupus UK.

## Biographies



**Professor David Isenberg, MD, FRCP, FAMS**  
University College London, UK

### Disclosures

*Professor Isenberg does not accept personal honoraria but asks that an equivalent sum is given to an arthritis charity of his choosing.*

*Professor Isenberg is a member of the Lupus Academy Steering Committee.*

David Isenberg is the Arthritis Research UK Diamond Jubilee Professor of Rheumatology at University College London (UCL). He graduated from St. Bartholomew's Hospital, London, in 1973, and trained in general medicine, rheumatology, neurology, psychiatry and gastroenterology, becoming a Research Fellow at UCL/The Middlesex Hospital in 1979. He was awarded his MD in 1984, based on his studies of myositis. During a year of research at Tufts University, Boston, he became interested in autoantibody structure/function and origin. He was appointed Consultant Rheumatologist in 1984, Professor in 1991 and became the arc Diamond Jubilee Chair of Rheumatology at UCL in 1996. He has fellowships of both the Royal College of Physicians and the Academy of Medical Sciences.

Professor Isenberg is on the Editorial Boards of five journals, including the *Journal of Rheumatology*. He is Chair of the British Isles Lupus Assessment Group and Lupus UK's Research Committee and was Chair of the Systemic Lupus International

Collaborating Clinics group (1998–2003). During the past 20 years, Professor Isenberg has undertaken many roles at Arthritis Research UK and currently sits on the Executive Board. He is past-President of the British Society for Rheumatology (2004–2006) and he chaired the Society's Biologics Register Committee for 5 years (2006–2011). Professor Isenberg was the 2010 recipient of the Evelyn V. Hess Prize from the Lupus Foundation of America for his contribution to lupus research and treatment. He has authored over 550 original articles, 275 reviews/chapters and 18 books, many on topics related to lupus.

Professor Isenberg's principal clinical interests are the development of disease activity and damage assessment tools in patients with lupus. His specialist interest is autoimmune rheumatic diseases, notably systemic lupus erythematosus, Sjogren's syndrome, myositis and antiphospholipid antibody syndrome. In 2016 he became a Master of the American College of Rheumatology.



**Professor Munther Khamashta, MD, PhD, FRCP**  
St. Thomas' Hospital, London, UK & Dubai Hospital, Dubai, UAE

### Disclosures

#### Consultant/Advisor:

INOVA Diagnostics;  
Medimmune/AstraZeneca;  
NewBridge

*Professor Khamashta is a member of the Lupus Academy Steering Committee and has been involved in the planning and development of the Lupus Academy Middle East Summit Conference programme and materials.*

Munther Khamashta is Professor/Consultant Physician and Director of the Lupus Research Laboratory at St Thomas Hospital, London. He studied medicine in Barcelona and internal medicine in Madrid, Spain, where he developed an interest in connective tissue diseases and received his PhD. He was awarded the MRCP in 1999 and FRCP in 2002. He joined the Lupus Unit in London 30 years ago and has been instrumental in developing it into an internationally recognised tertiary centre receiving referrals from all over the UK. He is currently on sabbatical setting up lupus services at Dubai Hospital, United Arab Emirates.

Professor Khamashta has served on the Editorial Boards of many journals, including Clinical & Experimental Rheumatology, Lupus, and Current Rheumatology Reviews. He is a member of several professional societies, including the International

Society of Internal Medicine, the American College of Rheumatology, and the Spanish Society of Rheumatology. He is a member of the Steering Committee of the International Board on the Study of Antiphospholipid Antibodies and of the Steering Committee of the International Advisory Board for Systemic Lupus Erythematosus. He is a founding member of the Lupus Academy and APS-ACTION. He has received several international awards for his work, including the European League Against Rheumatism and International League Against Rheumatism prizes.

Professor Khamashta has a strong research interest in lupus and connective tissue diseases, with a special interest in pregnancy and antiphospholipid syndrome. He has published extensively in Lupus, Hughes Syndrome and related areas, with more than 700 original papers.

Photo: Peter Pulkowski



**Professor Annegret Kuhn, MD, MBA**  
University Medical Center Mainz, Germany

Professor Kuhn is Head of the Interdisciplinary Study Center for Clinical Trials (IZKS) at the University Medical Center of the Johannes Gutenberg University of Mainz, Germany.

She acquired her Doctor of Medicine at the University of Düsseldorf, Germany, where she also received her German Board Certification for Dermatology and Venereology and her Qualification as University Lecturer. In 1999, Professor Kuhn was awarded a Lise-Meitner Fellowship by the Ministry for School and Education of North Rhine-Westphalia and in 2004 she was awarded a Heisenberg Fellowship by the German Research Foundation (DFG). She has worked in several University Departments and Scientific Institutions, such as the Max-Planck-Institute for Molecular Biomedicine, Münster, and the German Cancer Research Center, Heidelberg, Germany.

Her major clinical and scientific interests include translational immunology and human autoimmune diseases. Professor Kuhn is particularly interested in the pathogenesis, diagnosis, and classification

and treatment of cutaneous lupus erythematosus. The IZKS performs translational research and transfers technologies from basic science to pharmaceutical application by conducting investigator-initiated and sponsor-initiated trials according to the guidelines for Good Clinical Practice. Professor Kuhn is Chair of the European Society of Cutaneous Lupus Erythematosus (EUSCLE) and member of several German and international medical societies.

Professor Kuhn is interested in Health Care and Science Management. In 2013, she completed the MBA Health Care Management Program at the University of Bayreuth, Germany. She is also member of various editorial and clinical advisory boards and reviewer of several clinical and scientific peer reviewed journals. She has published widely in high impact journals, such as *Nature Medicine*, *Nature Reviews Rheumatology*, *Annals of the Rheumatic Diseases*, and the *Journal of Investigative Dermatology*, including more than 150 original and review articles.

## Disclosures

### Grants/Research

#### Support:

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## Biographies



**Professor Liz Lightstone, PhD, FRCP**  
Imperial College London, UK

### Disclosures

Grants/Research

Support:

Roche

Consultant/Advisor:

GSK; Medimmune

Meeting Honorarium/

Expenses:

GSK; UCB

Liz Lightstone is Professor of Renal Medicine in the Division of Immunology and Inflammation, Department of Medicine, Imperial College London, and an Honorary Consultant Renal Physician in the Imperial College Healthcare NHS Trust Renal and Transplant Centre (ICHNT RTC). After an undergraduate degree at Cambridge, she graduated in medicine from the University of London in 1983 and trained in nephrology at the Royal Postgraduate Medical School. She won a Medical Research Council (MRC) Training Fellowship in 1988 and undertook a PhD in immunology at University College London. This was followed by a MRC Clinician Scientist Fellowship at the Royal Postgraduate Medical School. She was appointed Senior Lecturer and Honorary Consultant Physician in 1995.

Professor Lightstone has held major roles in undergraduate and postgraduate medicine at Imperial College, in particular as Director of the Academic Foundation programme within the North West Thames Foundation School from 2009 to 2015. She is a member of the LUPUS UK Peer Review Panel. She was an elected member of the UK Renal Association Executive and remains active in the Renal Association Programme Planning group and Equal Opportunities Committee. She is on the EU executive of the Lupus Nephritis Trials Network, was an author on the 2012 EULAR recommendations for the treatment of lupus nephritis and regularly advises on the design of trials in lupus nephritis. She is co-Chair of the UK Renal Association Glomerulonephritis Clinical Study Group.

Professor Lightstone's research is now focused on Lupus Nephritis as well as Pregnancy in Women with Kidney Disease. Together with colleagues in the ICHNT RTC, she pioneered the use of steroid-minimising regimens in lupus nephritis. She is Chief Investigator on the international multicentre randomised RITUXILUP trial funded by Arthritis Research UK. She is particularly interested in developing better ways of predicting outcomes, not least by improving adherence to therapy. To this end she has established a new assay of hydroxychloroquine levels as a marker of adherence to therapy. She is also working on identifying tissue and urine biomarkers and histological features that better predict the outcome of lupus nephritis. She was the inaugural National Coordinator of the Pregnancy and Chronic Kidney Disease Rare Disease group and has pioneered the use of tacrolimus in the treatment of lupus nephritis in pregnancy.

Her main clinical interests are in lupus nephritis (she jointly manages a combined renal/rheumatology lupus clinic following over 400 patients with lupus nephritis) and the management of women with kidney disease in pregnancy—she established and runs a renal obstetric clinic and a preconception counselling clinic.





### **Professor Guillermo Ruiz-Irastorza, MD, PhD**

Autoimmune Research Unit, Cruces University Hospital, Bizkaia, Spain

#### **Disclosures**

None

Guillermo Ruiz-Irastorza is Head of the Autoimmune Research Unit at Cruces University Hospital, Bizkaia, Spain, where he has been since 2001.

Professor Ruiz-Irastorza received his MD from the Universidad Autónoma de Madrid, Spain in 1990 and became a specialist in internal medicine in 1996. Following his PhD from the University of the Basque Country, Spain in 1999, he spent a year as a Research Fellow at the Lupus Research Unit, St Thomas' Hospital, London, UK, before returning to the Hospital Universitario Cruces as Consultant Physician in Internal Medicine. He became Professor of Medicine at the University of the Basque Country, Spain in 2004.

Professor Ruiz-Irastorza is a member of the Editorial Board of the journal *Lupus*, and a reviewer of several other journals in the fields

of rheumatology and autoimmune diseases, including *Annals of Rheumatic Diseases*, *Arthritis & Rheumatology*, *Rheumatology*, *Journal of Rheumatology* and *Lupus Science & Medicine*.

He is a member of the Grupo de Estudio de las Enfermedades Autoinmunes Sistémicas (GEAS), and coordinator of the first Spanish national lupus inception cohort study. He has been member of the Systemic Lupus International Collaborating Clinics since 2008.

Professor Ruiz-Irastorza's clinical and research interests focus on systemic lupus erythematosus, antiphospholipid syndrome, and pregnancy and autoimmune diseases. He is author of 151 peer-reviewed publications and 20 book chapters.



### **Professor Zoubida Tazi Mezalek, MD**

Ibn Sina Hospital, Rabat, Morocco

#### **Disclosures**

None

Zoubida Tazi Mezalek is a Moroccan internist with 21 years of medical practice. She is currently, the Head of the Internal Medicine and Hematology Unit at Ibn Sina Hospital, Rabat, Morocco.

Professor Tazi Mezalek studied in Morocco, graduated in medicine from Rabat University in 1992 and completed her residency in internal medicine at Ibn Sina Hospital in 1996. She completed her fellowship in internal medicine at French National Reference Centre for Systemic Lupus Erythematosus at Pitie-Salpetriere Hospital of Paris (France) between 1993 and 1995.

Professor Tazi Mezalek became an Associate Professor in 1996, and joined the Department of Internal Medicine at Ibn Sina Hospital as a Senior Lecturer, Senior Practitioner and Professor of Internal Medicine at the University of Rabat's Faculty of Medicine in 2000. She is actively involved in undergraduate and postgraduate training in haematology and internal medicine.

She is an International Advisor of the French Medical Review of Internal Medicine, and member of the group of the international and multidisciplinary expert recommendations for the use of biologics in systemic lupus erythematosus: the FLEUR, CRI-IMIDIATE and FAI2R task forces. She is also Chair and a member of several committees in the Moroccan Health Authority, Treasurer of the Moroccan Society of Internal Medicine, and President of the Moroccan Society of Vascular Diseases.

Professor Tazi Mezalek has conducted some research projects on clinical and immunological features of lupus patients in Morocco. Her main interests are systemic lupus erythematosus, Behcet's disease, Takayasu's disease and systemic connective tissue diseases.

In the last 15 years, she has published over 100 peer-reviewed papers, of which 30 focused on the clinical features of lupus in Morocco.



## Biographies



### **Professor Imad Uthman, MD, MPH, FRCP**

Medical Centre of the American University of Beirut, Lebanon

#### **Disclosures**

**Grants/Research**

**Support:**

Newbridge; Pfizer; Roche

**Meeting Honorarium/**

**Expenses:**

Abbvie; Newbridge; Pfizer;

Roche

Imad Uthman is Professor of Clinical Medicine and Head of the division of Rheumatology at the Faculty of Medicine and Medical Centre of the American University of Beirut, Lebanon. He received his MD from the American University of Beirut in 1988, before qualifying in internal medicine and then specialising in rheumatology. He also spent time as a Rheumatology Fellow at Notre Dame Hospital, Montreal, Canada, returning to his *alma mater* in 1995. He is past-President of the Lebanese Society of Rheumatology, and sits on his institution's Biomedical Review Board. He is the international advisor in Lebanon for the Royal College of Physicians of London.

Professor Uthman's major research interests include the study of antiphospholipid syndrome, systemic lupus erythematosus (SLE), biologic therapies in rheumatic diseases, vasculitis, paediatric rheumatology, and the clinical characteristics of rheumatic diseases in Lebanon. He is the author of more than 100 publications on various aspects of SLE, lupus and the rheumatic diseases in leading international rheumatology journals.

## Notes

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**Professor David Isenberg, MD, FRCP, FAMS**  
University College London, UK

## Biomarkers in SLE: How useful are they?

### References

1. Murphy G, Lisnevskaja L, Isenberg D. Systemic lupus erythematosus and other autoimmune rheumatic diseases: challenges to treatment. *Lancet*. 2013;382(9894):809–18.
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4. Budde P, Zucht HD, Vordenbaumen S, et al. Multiparametric detection of autoantibodies in systemic lupus erythematosus. *Lupus*. 2016;25(8):812–22.
5. Biesen R, Rose T, Hoyer BF, Alexander T, Hiepe F. Autoantibodies, complement and type I interferon as biomarkers for personalized medicine in SLE. *Lupus*. 2016;25(8):823–9.

Biomarkers play a diversity of roles in the diagnosis and management of patients with systemic lupus erythematosus (SLE). For example, some antibodies to double-stranded DNA (ds-DNA) and antiphospholipid antibodies are used in classification and diagnosis – both the ACR and SLICC criteria sets use them for instance. Serial measurements of certain biomarkers, such as serum C3 and anti-dsDNA antibodies are helpful to the clinician as they may reflect clinical activity or, on occasion, with rising DNA-binding and falling C3 levels predict a pending lupus flare.

Biomarkers are of value in identifying disease subsets, for example anti-Ro antibodies are linked to photosensitivity and neonatal lupus syndrome, and anti-phospholipid antibodies are associated with increased risk of clotting and miscarriages.

Organ-specific biomarkers such as the serum creatinine in renal disease, lung function tests in pulmonary disease and the creatinine kinase enzyme levels in muscle disease will help to capture end effects.

It is notable that autoantibodies may be detected in the serum of some healthy individuals destined to develop lupus many years later. Thus anti-ENA and anti-dsDNA antibodies can occasionally be found in patients whose disease will not become clinically overt for 10 years.

There is an ongoing attempt to develop a wide variety of newer and better biomarkers. It is believed by some that complement breakdown products (e.g., C3A, C4A) may be more useful than measuring the parent molecule for example. Numerous attempts have been made to identify biomarkers in the serum (e.g., anti-C1q) or urine (e.g., TWEAK) of patients with lupus nephritis in order to help distinguish between activity and damage in the kidney. Numerous cytokines, chemokines and adhesion molecules have been measured in patients with lupus and varying claims are made for their utility.

### Learning Objectives

- Understand the range of functions that biomarkers can be used for in SLE.
- Appreciate the modest benefits and deficiencies of the currently available biomarkers.
- Understand the need for developing new biomarkers.





## Professor Guillermo Ruiz-Irastorza, MD, PhD

Autoimmune Research Unit, Cruces University Hospital, Bizkaia, Spain

## Guidelines: Use of steroids and hydroxychloroquine in SLE

### References

1. Ruiz-Irastorza G, Ramos-Casals M, Brito-Zeron P, Khamashta MA. Clinical efficacy and side effects of antimalarials in systemic lupus erythematosus: a systematic review. *Ann Rheum Dis*. 2010;69(1):20–8.
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Antimalarials and glucocorticoids have long been the basis of the treatment of systemic lupus erythematosus (SLE). Both treatments share a high level of efficacy, however, their safety profiles are radically different.

Several studies have demonstrated the effect of antimalarials in controlling lupus activity, reducing damage accrual and improving survival of patients with SLE.<sup>1</sup> Side effects are usually mild, with retinal toxicity being rare and easy to detect at early stages with regular eye monitoring, which should include high-sensitivity techniques such as optical coherence tomography.<sup>2</sup>

The use of high doses of oral prednisone/prednisolone has been recommended by most guidelines in cases of moderate to high lupus activity. However, such recommendations are based more on custom than on true evidence. Results from clinical studies have consistently shown that glucocorticoid-related damage is dose-dependent, with doses below 5–7.5 mg/d having a good safety profile whereas doses >30 mg/d are associated with a sharp increase in the frequency

of side effects.<sup>3</sup> The use of methylprednisolone pulses has been free from secondary damage in most studies.<sup>3</sup> In terms of efficacy, recent data from observational studies and a small clinical trial support the fact that low doses of oral prednisone are as effective as high doses for treating active lupus including mainly, but not only, renal disease.<sup>4,5</sup>

Thus, antimalarials should be considered the main treatment for SLE, and are therefore recommended for all patients with lupus without contraindications. Glucocorticoids should be used to treat specific manifestations of the disease. Mild flares can be managed with doses up to 7.5 mg/d and, when more potency is needed, we advocate three consecutive methylprednisolone pulses of 125 mg. Severe organic manifestations can be initially managed with pulses of 250 or 500 mg, followed by a medium dose of prednisone, always <30 mg/d with very rapid tapering to maintenance doses ≤5 mg/d, usually in combination with immunosuppressive drugs in order to help control lupus activity and spare steroids.

### Learning Objectives

- Understand the excellent efficacy and safety profiles of hydroxychloroquine.
- Understand the current recommendations for monitoring antimalarial retinal toxicity.
- Discuss published data in SLE patients showing the association of different doses of glucocorticoids with side effects including damage.
- Recall the results from recent studies on the efficacy and toxicity of therapeutic schemes using lower doses of prednisone in severe lupus.
- Implement practical guidelines for using antimalarials and lower doses of glucocorticoids in the setting of active lupus.

## Notes

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**Professor Graham Hughes, MD, FRCP**  
The London Lupus Centre, London, UK

## The challenge of APS

### References

1. Hughes Syndrome: Highways & Byways  
Hughes GRV & Khamashta MA 2013. Springer. ISBN 978-1-4471-5160-9.
2. Hughes GR. Hughes syndrome/APS. 30 years on, what have we learnt? Opening talk at the 14th International Congress on antiphospholipid antibodies Rio de Janeiro, October 2013. *Lupus*. 2014;23(4):400–6.

Where will Hughes Syndrome be in 2050? To attempt a forecast for such a rapidly developing subject is hard. For what it is worth, here are my forecasts:

1. Antiphospholipid (aPL) testing will become standardised in kit form.
2. aPL testing kits will be available 'over the counter' in pharmacies and drug stores world-wide.
3. Antiphospholipid Syndrome (APS)/Hughes Syndrome will become recognised as a major (and treatable) cause of migraine.
4. The incidence of stroke and heart attack, especially in younger (<45 years) women will be reduced by a half, thanks to widespread aPL testing.
5. aPL positivity will be recognised in general practice as an even more important health check than cholesterol.
6. aPL testing will be included in the routine 'early pregnancy' screen.
7. Stillbirths will be reduced by 50% ("the Stillbirth Scandal").
8. aPL testing allow for the successful treatment for some cases of "multiple sclerosis".
9. Not all cases of memory loss are caused by "Alzheimer's disease" – some are due to Hughes Syndrome ....., which is treatable.

### Learning Objectives

- Understand the importance of high international normalised ratio in many APS cases.
- Understand whether two tests 12 weeks apart is really necessary.
- Recognise why pregnancy testing for APS is currently poor.
- Appreciate the difficulties of diagnosing seronegative Hughes Syndrome.
- Recognise "fellow travellers": Hughes Syndrome, Sjogren's Syndrome and thyroid problems.



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## Curbside Consults



**Moderator:** Jamal Al-Saleh (UAE)

**Dr Jamal Al-Saleh, MD, FRCP**

Dubai Hospital, Dubai, UAE

**Panellists:** Munther Khamashta (UK/UAE), David Isenberg (UK), Ian Bruce (UK), Liz Lightstone (UK)

### Curbside consults: The experts tackle lupus nephritis and lupus arthritis



Curbside Consults may be a new feature for the Lupus Academy in 2016, but it is an old practice that occurs in every country and every hospital. Confronted with a difficult clinical problem, the physician “curbside” another physician for what is an informal opinion about the case. The answer may be based in evidence or it may be purely opinion. Regardless, this type of transfer of information occurs all the time. Nothing is written and money is not exchanged. In fact, most of the time, the “consultant” doesn’t even see the patient. These educational interactions are very efficient with a rapid transfer of knowledge or experience in a short period of time.



The Lupus Academy Middle East Summit Conference Curbside Consults feature a panel of rheumatologists and a renal physician who will render their views about challenging clinical scenarios. The first area chosen for this session deals with the lupus nephritis patient who has had a partial response to a therapeutic intervention. We all have such patients, but we all have different views about how to proceed. The second topic deals with the lupus patient who has an erosive form of arthritis, the “rhupus” patient. Should this type of complication be treated differently than conventional lupus arthritis? The discussions will focus on these themes in this Curbside Consults session.



#### Learning Objectives

- Recognise therapeutic options for the treatment of patients with lupus nephritis who partially respond to induction therapy.
- Describe therapeutic approaches to the patient with erosive arthritis.



## Notes

### References

- 1.** Dall'Era M, Cisternas MG, Smilek DE, et al. Predictors of long-term renal outcome in lupus nephritis trials: lessons learned from the Euro-Lupus Nephritis cohort. *Arthritis Rheumatol.* 2015;67(5):1305–13.
- 2.** Li J, Wu H, Huang X, et al. Clinical analysis of 56 patients with lupus syndrome: manifestations and comparisons with systemic lupus erythematosus: a retrospective case-control study. *Medicine (Baltimore).* 2014;93(10):e49.





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# Case Study Workshops

## Friday 9<sup>th</sup> December

### Parallel Case Study Workshops (Friday PM)





#### Four Parallel Workshops Focusing on Best Clinical Practice for Managing Lupus Manifestations

17:30	<i>Moderator: Jamal Al-Saleh (UAE)</i> Workshop: 1 — Investigating the febrile lupus patient	 Guillermo Ruiz-Irastorza (Spain) & Zoubida Tazi Mezalek (Morocco)
	<i>Moderator: Ricard Cervera (Spain)</i> Workshop: 2 — Haematologic challenges: Cytopenias	 Ahmed Al-Shaikh (Saudi Arabia) & Ala Al-Heresh (Jordan)
	<i>Moderator: Munther Khamashta (UK/UAE)</i> Workshop: 3 — Pregnancy and lupus	 Samar Al-Emadi (Qatar) & Humaid Al-Wahshi (Oman)
	<i>Moderator: Graham Hughes (UK)</i> Workshop: 4 — CNS and lupus	 Imad Uthman (Lebanon) & Khaled El-Hadidi (Egypt)
19:00	Close (Day 1)	
20:00	Welcome Dinner	

## Saturday 10<sup>th</sup> December

### Parallel Case Study Workshops (Saturday AM)

#### Four Parallel Workshops Focusing on Best Clinical Practice for Managing Lupus Manifestations

09:00	<i>Moderator: Jamal Al-Saleh (UAE)</i> Workshop: 1 — Investigating the febrile lupus patient	 Guillermo Ruiz-Irastorza (Spain) & Zoubida Tazi Mezalek (Morocco)
	<i>Moderator: Ricard Cervera (Spain)</i> Workshop: 2 — Haematologic challenges: Cytopenias	 Ahmed Al-Shaikh (Saudi Arabia) & Ala Al-Heresh (Jordan)
	<i>Moderator: Munther Khamashta (UK/UAE)</i> Workshop: 3 — Pregnancy and lupus	 Samar Al-Emadi (Qatar) & Humaid Al-Wahshi (Oman)
	<i>Moderator: Graham Hughes (UK)</i> Workshop: 4 — CNS and lupus	 Imad Uthman (Lebanon) & Khaled El-Hadidi (Egypt)
10:30	Break	

### Please Note

These workshops will be held in breakout rooms. Please follow the appropriate signs/symbols corresponding to the workshops you are registered to attend.



**Moderator:** Dr Jamal Al-Saleh (UAE)

**Presenters:** Guillermo Ruiz-Irastorza (Spain) & Professor Zoubida Tazi Mezalek (Morocco)

## Investigating the febrile lupus patient

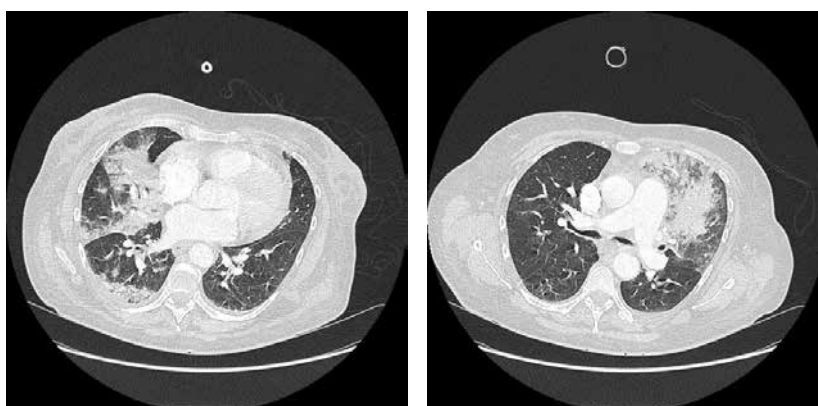


**Professor Guillermo Ruiz-Irastorza, MD, PhD**

### Case 1: 52-year-old female with bilateral pulmonary infiltrates and bilateral pleural effusion

A 52-year-old female was admitted with fever, cough with bloody sputum and bilateral lung infiltrates. She had a previous diagnosis of systemic lupus erythematosus (SLE) 7 years ago, with Class IV lupus nephritis diagnosed one month before admission. She was being treated with hydroxychloroquine 200 mg/d, prednisone 10 mg/d, cyclophosphamide 500 mg/2w, with a total of two pulses received, calcium plus vitamin D and losartan.

On admission, she was extremely unwell, febrile, with tachypnoea. Inspiratory crackles could be heard on both pulmonary bases. Blood tests showed hypoxaemia (pressure of oxygen 61 mmHg with oxygen at 4 lpm), Hb 7.6 g/dL, leukocytes 3700/mm<sup>3</sup>, creatinine 1.2 mg/dL, albumin 2.8 g/L, ferritin 646 mg/dL, complement C3 48 mg/dL, C4 8 mg/dL. Her chest X-ray showed bilateral pulmonary infiltrates and bilateral pleural effusion, confirmed by CT scan.



The patient was admitted to the intensive care unit.

#### Discussion points:

*Diagnostic and therapeutic workup.*



**Professor Zoubida Tazi Mezalek, MD**

### Case 2: Macrophage activation syndrome as a life-threatening complication of systemic lupus erythematosus

A 23-year-old female presented to the emergency unit with fever, abdominal pain and vomiting. The fever had lasted one month and was not associated with chills. She was being treated empirically with oral antibiotics by a local physician but had no relief.

She had asymmetrical polyarthritis involving larger joints more than smaller joints, with axial sparing and no early morning stiffness. On admission, her temperature was 40.0°C, heart rate 100 bpm and blood pressure 100/60 mmHg. She had oral ulcers and malar rash, and also complained of polyarthralgia and myalgia. General physical examination revealed presence of pallor, facial puffiness and moderate hepatosplenomegaly on abdominal examination. Cardiovascular and CNS examinations were unremarkable.

Her laboratory examinations showed ESR 68 mm/h, WBC  $10.1 \times 10^9/L$ , haemoglobin 95 g/L, platelet count  $122 \times 10^9/L$ , elevated liver transaminases (AST 454 IU/L and AGT 306 IU/L) and LDH 1893 IU/L (normal range 200–400 IU/L). Lipase levels were 980 IU/L. Abdominal CT scan was consistent with grade B pancreatitis. Her triglycerides (458 mg/dL) and ferritin (6580 ng/mL) were markedly elevated. Her 24-hour urinary protein was 1210 mg/d, but her other renal parameters like serum urea and creatinine were normal. Her infectious disease work-up was negative. She was initially treated with intravenous broad-spectrum antibiotics.

Over one week, her symptoms did not improve and laboratory studies showed worsening pancytopenia and rising liver transaminases. Immunological screening was positive for ANA (1:160) with a homogenous pattern, anti-dsDNA, and decreased complement C3. Screening tests included Widal, human immunodeficiency virus (HIV), anti-hepatitis B surface antibody, hepatitis C antibody, *brucella*, Epstein–Barr virus (EBV), and all were found to be negative. Echocardiogram showed mild pericardial effusion with left ventricular ejection fraction of 60%. Bone marrow aspirate was cellular reactive marrow with no evidence of haemophagocytosis.

Hence, our patient was diagnosed with systemic lupus erythematosus (SLE) with pancreatitis and renal disease, complicated with concurrent macrophage activation syndrome (MAS). She was treated first with IV pulse methylprednisolone without improvement. After one week etoposide and mycophenolate mofetil was prescribed. The patient started feeling better 2 days after the start of therapy. Renal histology showed Class III lupus nephritis. Cytopenias, normalised by Day 12 and serum ferritin came down in about 3 weeks. She is currently in remission and on regular follow-up.

#### Discussion points:

*Early recognition of life-threatening complications in SLE.*

*Treatment of MAS in patient with SLE*

### Professor Zoubida Tazi Mezalek, MD

#### Case 3: Infectious complications of SLE

A 53-year-old Moroccan female with SLE presented with fever of 2–3 months duration. She complained of gradual worsening fatigue, malaise, night sweats, unintentional weight loss of 18 kg and intermittent headaches. She was diagnosed with SLE at the age of 38 years and was undergoing treatment with oral low dose of prednisone (5 mg) and hydroxychloroquine (400 mg). Initial clinical presentation of SLE was mainly mucocutaneous (malar rash and oral ulcerations), arthralgia, pleuritis, with positive DNA antibodies.

The patient had been treated for presumptive non-specific infectious disease, having shown no signs of improvement. Due to prolonged fever and worsening of weight loss, the patient was admitted to the Department of Internal Medicine at Rabat Medical Teaching Hospital to undergo further tests. She denied having chills, loss of appetite, cough, shortness of breath, diarrhoea or urinary symptoms. The patient had lived most of her life in the Morocco, with no “exotic” trips. Her SLE had been deemed controlled, until these symptoms started, and she had no history of tuberculosis or any chronic febrile infection. Physical examination on admission revealed fever (38.5°C), marked pallor with normal vital signs. There were no abnormalities on cardiac or lung auscultation, no rash, and her neurologic and articular examination were unremarkable. Abdominal examination was consistent with hepatomegaly and mild splenomegaly. Preliminary laboratory examinations showed an increased ESR (96 mm/h) and CRP (256 mg/L) and decreased WBC ( $2.4 \times 10^9/L$ ), lymphocytes ( $0.75 \times 10^9/L$ ) and haemoglobin (79 g/L), a low serum albumin (18 g/L) with increased total proteins (90 g/L). A 24 hour urine protein was performed and found to be 2.54 mg/24 h. Blood was also collected for culture of bacteria and for serologic study. ANAs were positive with high titer of 1:1,280. A CT scan of abdomen and chest revealed hepatomegaly (17.3 cm), mild splenomegaly (15.7 cm), with no enlargement of lymph nodes. A urine sample tested positive for *Escherichia coli*. Anti-infective treatment (ceftriaxone 2 g/day and gentamycin 160 mg/day) was administered, with an incomplete improvement of symptoms.

After 10 days, the symptoms worsened with high fever (39.2°C) and upper abdominal pain. New pulmonary X-ray revealed slight pleurisy and laboratory examinations showed a high CRP (156 mg/l) and elevated procalcitonine of 0.6 ng/mL (0.02–0.06 ng/mL). Complement (C3) 0.39 g/l was slightly decreased. She was treated on tazocilline and amikacin, and pulse methylprednisolone 750 mg IV was





## Case Study Workshop

started for 3 days then switched to oral prednisone 60 mg/d, without success. She also had been treated for presumptive tuberculosis for 3 weeks, having shown no signs of improvement. Serial blood cultures for bacteria, mycobacteria were persistently negative. The serologic study for EBV, cytomegalovirus and parvovirus B-19, HIV, *Coxiella burnetti*, *Mycoplasma pneumoniae*, *Salmonella*, *Brucella* and for *Rickettsia conorii* were negative. Bone marrow cultures for bacteria and fungi were negative. The patient had progressively worsening of cytopenias, with Hb 6.2 mg/dL, WBC 3020/mm<sup>3</sup>, and platelet count of 98,000/mm<sup>3</sup>, and had also marked polyclonal hypergammaglobulinemia and elevation of lactic dehydrogenase of 2331 U/L.

One hypothesis was finally considered: the patient had prolonged fever, hepatosplenomegaly, pancytopenia, and increased protein with hypergammaglobulinemia.

### Discussion points:

*Early recognition of life-threatening infectious complications in SLE.*

*Distinguishing infections and flares in patients with SLE.*

### Learning Objectives

- Discuss the differential diagnosis of the febrile lupus patient with lung infiltrates.
- Understand diagnostic and therapeutic approach in the context of a life-threatening situation.
- Better identify life-threatening complications in SLE.
- Recognise of life-threatening infectious complications in SLE early.
- Differentiate infections and flares in patients with SLE.
- More effectively treat MAS in patients with SLE.

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**Moderator:** Professor Ricard Cervera (Spain)

**Presenters:** Dr Ahmed Al-Shaikh (Saudi Arabia) & Dr Ala' Al-Heresh (Jordan)

## Haematologic challenges: Cytopaenias



### Dr Ahmed Al-Shaikh, MD

#### Case 1: 36-year-old female-Mezna

Mezna, a 36-year-old housewife, presented with 8-year history of systemic lupus erythematosus (SLE). Her lupus manifestations included polyarthritis, malar rash, alopecia, pleuritis, positive ANA and dsDNA antibodies and low complement. In addition she had proteinuria (1.5 g/24 h), with a kidney biopsy showing Class II glomerulonephritis according to SN classification. She had three 2<sup>nd</sup> trimester foetal losses, one living child, and no history of thrombosis. Her laboratory profile revealed high IgA ACA on several occasions, but negative ACA, IgG, IgM, IgA, anti-β2 GP1 and lupus anticoagulant. She was treated with oral prednisone, initially 1 mg/kg/d + hydroxychloroquine 400 mg/d + mycophenolate mofetil 2 g/d plus angiotensin converting enzyme inhibitors.

She was admitted in June 2014 with severe pancytopenia; Hb 7.2 (12.0–16.0 g/dL), WBC 1.8 ( $4.5\text{--}10.5 \times 10^9/\text{L}$ ), neutrophils 1.1 ( $1.35\text{--}7.5 \times 10^9/\text{L}$ ), reticulocytes 18 ( $25\text{--}85 \times 10^9/\text{L}$ ), platelets 6 ( $150\text{--}400 \times 10^9/\text{L}$ ). She presented with bruises, oral ulceration and alopecia. Peripheral blood smear showed thrombocytopenia, but no schistocyte. Her vitamin B12 was low 120 (140–637 pmol/L), but the haemolytic workup was negative. Her anti-dsDNA was high and complements C3 and C4 were low, but matched her baseline. Bone marrow biopsy confirmed severe aplastic anaemia with cellularity of less than 10%, and abnormal infiltration. She had no evidence for viral infections. She received three pulses of methylprednisolone (1000 mg each) followed by oral prednisone 1 mg/kg/d; a course of two doses of rituximab 1000 mg each; azathioprine 100 mg/day was discontinued and low vitamin B12 was replaced. The patient didn't respond and continued to have severe pancytopenia. Eventually, the patient was given an allogeneic stem cell transplant.

#### Discussion points:

*Causes of aplastic anaemia in SLE.*

*SLE as a cause of aplastic anaemia.*

*Management options for lupus patients with aplastic anaemia.*

*Role of bone marrow transplantation in such cases.*



### Dr Ahmed Al-Shaikh, MD

#### Case 2: 45-year-old male-Khaled

Khalid was referred to the haematology service following presentation with thrombocytopenia for one-month duration. His platelet count was 26 ( $150\text{--}400 \times 10^9/\text{L}$ ), rest of his CBC revealed: Hb 10.2 (12.0–16.0 g/dL), WBC 11.1 ( $4.5\text{--}10.5 \times 10^9/\text{L}$ ). He had history of epistaxis and bruises. His bone marrow was consistent with peripheral platelet destruction.

The patient was referred to rheumatology for further evaluation. He has a history of polyarthritis and fatigue, but denied mucocutaneous manifestations and his clinical examination was unremarkable. He was found to have positive ANA, anti-dsDNA and anti-sm and low complement (C3, C4). His platelet count dropped to  $2 \times 10^9/\text{L}$ . Despite dexamethasone 40 mg daily for 4 days, there was no significant improvement in the platelet count ( $7\text{--}9 \times 10^9$ ). Therefore he was given one dose of IVIG 1 g/kg, and started on prednisone 1 mg/kg/d, danazol 400 mg b.i.d., and hydroxychloroquine 400 mg/d. Ten days later his platelets increased to  $250 \times 10^9/\text{L}$ . With tapering the prednisone dose, the platelet count started to fall gradually to  $120 \times 10^9/\text{L}$ .

#### Discussion points:

*Recognising and diagnosing thrombocytopenia in SLE.*

*Management options for patients with lupus and thrombocytopenia.*

**Dr Ala' Al-Heresh, MD, FRCP****Case 3: 22-year-old female**

A 22-year-old female university student presented for the first time with sudden onset skin rash in both lower limbs 2 weeks prior to her presentation. She had no history of drug use.

Clinical examination showed she was afebrile, normotensive, with mild pallor and icterus. She did not have lymphadenopathy, and had intact peripheral pulses. Chest, heart and abdominal examinations were unremarkable. She had symmetrical small joint arthritis, malar rash and palpable purpuric skin rash in both lower limbs.

Initial laboratory investigation revealed: Hb 7.3 g/dL normochromic anaemia, high reticulocyte count, platelet count 30,000/mm<sup>3</sup>, ESR 85 mm/1<sup>st</sup> hr, direct bilirubin 2.2 mg/dL, positive ANA 1/640 and dsDNA, and low complement (C3, C4). She had positive platelets antibody and a positive direct Coombs test. Urine analysis showed no proteinuria or active casts. Chest X-ray was normal.

The working diagnosis was Evans syndrome, SLE complicated with autoimmune thrombocytopenia and Coombs positive autoimmune haemolytic anaemia. Her platelet count dropped further to critical levels (<50,000/ $\mu$ L) in spite of medication. She developed diffuse ecchymosis.

During hospitalisation, she received the following medications: IV pulse methylprednisolone, oral prednisolone, hydroxychloroquine, IVIG for 5 days and two doses of rituximab. She did not respond to these medications during her 3 weeks of hospitalisation. Splenic embolisation was performed, with rapid correction of her thrombocytopenia.

**Discussion points:**

*How to manage refractory cases of thrombocytopenia in patients with SLE.*

**Dr Ala' Al-Heresh, MD, FRCP****Case 4: 16-year-old female**

A 16-year-old female with SLE, diagnosed 10 months ago, presented with a history of polyarthritis, malar rash and recurrent oral ulcers. Her CBC was normal, ESR 39 mm/1hr, CRP negative, ANA 1/640 (speckled pattern), negative dsDNA, ENA and ACL antibodies. Urine analysis was negative for protein.

She was treated with, prednisolone and hydroxychloroquine, with good response and improvements in ESR (20 mm/1hr) and normal CBC. Prednisolone was reduced accordingly.

Six months later the patient presented with a progressive alteration in her level of consciousness and fatigue, which had been progressive for 2 weeks. Physical examination revealed the patient was very pale, confused, and afebrile, with no lymphadenopathy or focal neurological deficit. The rest of her physical examination was unremarkable.

Laboratory results revealed: Hb 8 g/dL, fragmented RBC, WBC 4,200/mm<sup>3</sup>, platelets 46,000/mm<sup>3</sup>, prothrombin time, partial thromboplastin time normal, ESR 56mm/1<sup>st</sup> hour; CRP high, urea 35 mg/dL, creatinine 1.0 mg/dL, total bilirubin 2.1 mg/dL, LDH 680 U/L (normal <250), protein: 8.1 g/dL, albumin 4 g/dL, urine nil for protein and active casts.

This young lady had thrombocytopenia and elevated lactate dehydrogenase, negative direct Coombs test, and peripheral blood smear showed fragmented RBCs. Working diagnosis was thrombotic thrombocytopenic purpura (TTP). The patient was successfully treated with pulse methylprednisolone, cyclophosphamide and plasma exchange.

**Discussion points:**

*Diagnoses, differential diagnosis and management plan for TTP.*



# Case Study Workshop

**Dr Ala' Al-Heresh, MD, FRCP**

**Case 5: 18-year-old female**

An 18-year-old female student, presented with history of general weakness, fatigue, and pain and swelling in the small joints of both hands for one year. She had history of photosensitivity, Raynaud's and alopecia and no history of relevant drug intake.

Examination showed she had pallor, was afebrile, with no lymphadenopathy and no skin rash. Joint examination, revealed signs of symmetrical active synovitis in the metacarpophalangeal and proximal interphalangeal joints. The rest of her physical examination was unremarkable.

She had been followed up by her local general practitioner and given NSAIDs, without proper relevant laboratory work up. Laboratory results revealed: Hb 8.5 g/dL, red cell indices suggestive of microcytic hypochromic cell morphology, WBC count 4,800/mm<sup>3</sup>, neutrophil count 1,400/mm<sup>3</sup>, platelets 110,000/mm<sup>3</sup>, reticulocyte count 0.04% and LDH 230 (normal <250). Coombs test was negative, ANA 1/640, anti-dsDNA was positive and ESR 88 mm/h. Working diagnosis was SLE with anaemia.

**Discussion points:**

*Diagnosis, differential diagnosis and management of anaemia in SLE.*

## Learning Objectives

- Recognise and describe the haematological manifestations of SLE:
  - anaemia
  - thrombocytopaenia
  - haemolytic anaemia
  - leucopaenia.
- Recognise unusual haematological manifestations of SLE.
- Discuss the therapeutic approaches to the haematological manifestations of SLE.

[illegible]



**Moderator:** Professor Munther Khamashta (UK/UAE)

**Presenters:** Dr Samar Al-Emadi (Qatar) & Dr Humaid Al-Wahshi (Oman)

## Pregnancy and lupus



**Dr Samar Al-Emadi, MBBS, FRCPC, FACR, ABIM**

### Case 1: 26-year-old female with Class IV lupus nephritis

A 26-year-old female, diagnosed with systemic lupus erythematosus (SLE) in 2013 presented with lupus nephritis Class IV, positive ANA, anti-dsDNA, low complement and haemolytic anaemia. She had previously been treated with pulse steroids, mycophenolate mofetil (MMF), hydroxychloroquine, angiotensin converting enzyme inhibitor (ACEi) and tapered dose prednisone. She had experienced no flares since first diagnosis.

Six months following presentation, she sought advice on becoming pregnant. She was newly married, her menstrual cycle was regular and her lupus was being managed with MMF 2 g/d, hydroxychloroquine 400 mg/d, prednisone 5 mg/d, calcium, vitamin D and ACEi. Disease activity (SLEDAI) and damage (SLICC) were measured, BP was 130/70 mmHg and laboratory tests revealed positive anti-dsDNA, lupus anticoagulant, and negative SS-A/Ro or SS-B/La antibodies.

#### Discussion points:

*Risk of flare during pregnancy.*

*Treatment recommendations during pregnancy.*



**Dr Humaid Al-Wahshi, MD, FRCP**

### Case 2: 27-year-old female with tiredness, polyarthralgia, hair loss and photosensitivity

A recently married 27-year-old female presented with tiredness, polyarthralgia, hair loss and photosensitivity. Her physical examination was stable, BP 110/80 mmHg, she was pale with facial rash, alopecia and active synovitis.

Investigations revealed: Hb 10 g/dL, platelets  $130 \times 10^9/L$ , WBCs  $3.6 \times 10^9/L$ , ESR 70 mm/h, CRP 8 U/mL, and normal LFT, U/E and G6PD.

Immunology revealed positive ANA, anti-ds DNA, anti-SM and Anti-Ro. Her complement C3 was normal and C4 was low. 24 hr urinary protein was 0.2 g/d and she tested negative for APLs.

#### Discussion points:

*Diagnosis, treatment and contraceptive advice.*

### Case 2: 27-year-old female with tiredness, polyarthralgia, hair loss and photosensitivity (continued)

One year later, the patient came for one of her regular follow up appointments. She expressed her desire to be pregnant, but had some questions that needed to be answered.

#### Discussion points:

*Will I have a successful pregnancy?*

*Will pregnancy cause disease flares?*

*Will my disease affect my baby?*

*What medications should I use?*

*What medications should I stop?*

*Should I become pregnant or not? (tests to be done and predictors of successful pregnancy).*



**Dr Humaid Al-Wahshi, MD, FRCP**

### **Case 3: 28-year-old female with positive ANA**

A 28-year-old asymptomatic female was referred to Rheumatology because of incidental findings of positive ANA revealed as part of premarital screening at private clinic (although not indicated).

#### **Discussion points:**

*What advice or action is appropriate?*

### **Case 3: 28-year-old female with positive ANA (continued)**

Two years later, she was referred by her Obstetrician as she had positive anti-Ro following delivery of her first baby with congenital heart block (CHB).

#### **Discussion points:**

*What risk does anti-Ro positivity carry for the mother and foetus?*

*The pathogenesis of neonatal lupus and CHB.*

*What advice would you give this patient?*

### **Case 3: 28-year-old female with positive ANA (continued)**

Three years later, she visited clinic with her husband as she wants to become pregnant, however her husband states he does not want her to become pregnant as he does not want to have a second child with CHB.

#### **Discussion points:**

*The risk of CHB in a pregnancy for a mother of a baby already having CHB.*

*Management advice during pregnancy and post-partum.*

### **Learning Objectives**

- Understand pregnancy and immunological changes in relation to lupus.
- Understand the influence of SLE on pregnancy and vice versa.
- Improve understanding of the pregnancy planning and pre-pregnancy work-up for patients with lupus.
- Describe the main predictors of pregnancy complications in women with SLE.
- Understand the importance of counselling during pregnancy in women with SLE.
- Ensure drug safety during pregnancy.
- Provide the most appropriate contraceptive advice/scheme for individual SLE patients.
- Understand how to manage a pregnant woman with positive anti-Ro/SS-A antibodies.
- Diagnose and manage antiphospholipid syndrome during pregnancy.



**Moderator:** Professor Graham Hughes (UK)

**Presenters:** Professor Imad Uthman (Lebanon) & Professor Khaled El-Hadidi (Egypt)

## CNS and lupus



### Professor Imad Uthman, MD, MPH, FRCP

#### Case 1: 25-year-old female with severe headache and a tonic-clonic seizure

A 25-year-old female was diagnosed with systemic lupus erythematosus (SLE) 3 years ago after she presented with polyarthrititis, malar rash, photosensitivity, leucopenia (WBC count:  $3.0 \times 10^9/L$ ) positive anti-nuclear antibody test (1/320), anti-ds DNA ( $>200$  IU/mL; positive  $\geq 20$ ), anti-SSB (La) (150.6 U/mL; positive  $\geq 25$ ), and anti-SSA (Ro) ( $>200$  U/mL; positive  $\geq 25$ ) antibodies. Her antiphospholipid antibody profile revealed positive lupus anticoagulant, and positive IgM anti-cardiolipin antibodies (95 MPL; positive). No renal involvement was documented during her follow-up.

She was managed with hydroxychloroquine 400 mg/d, prednisone 5 mg/d and low dose aspirin. Prednisone was progressively tapered and discontinued 6 months after her initial presentation. In January 2016 she presented with a 2 week history of severe throbbing headache, and a tonic-clonic seizure. An MRI of her brain revealed multiple white matter lesions (periventricular hyperintensities).

#### Discussion points:

*Diagnosis and management of CNS manifestations of SLE.*



### Professor Khaled El-Hadidi, MD

#### Case 2: Schizophrenia as an initial presentation of SLE

A 31-year-old female with a family history of SLE and receiving treatment for schizophrenia since 2005 presented 6 months ago with generalised body aches and joint pains. No other symptoms of rheumatic disease were noted in her medical history. Laboratory examination revealed elevated ESR (102 mm/h), Hb (9.8 gm/L), MCV (75 fl), TLC ( $3.5 \times 10^3$  Cmm), platelets ( $235 \times 10^3$  Cmm), SGPT (35 U/L), creatinine (1.2 mg/dL), urine analysis was unremarkable, 24 hour urine protein (54 mg) and TSH (2.5 mU/ml). She tested positive for ANA, anti-ds DNA, LAC, ACL IgG, ACL IgM and complement C3 and C4. Her brain MRI was normal.

#### Discussion points:

*Differentiate between neuropsychiatric manifestations that represent active disease versus others that are representing a longstanding disease.*

*Briefly discuss the underlying possible pathogenesis, whether inflammatory or thrombotic, to try and determine the best therapeutic approach.*

### Professor Khaled El-Hadidi, MD

#### Case 3: 26-year-old female with recurrent abortions and stroke

A 26-year-old female presented with a history of recurrent abortions and stroke. In 2008 she experienced a 1<sup>st</sup> trimester abortion, followed by generalised weakness due to a stroke. In March 2013 she had a 2<sup>nd</sup> trimester abortion leading to assessment for autoimmune disease. Examination revealed BP 150/110 mmHg, malar rash, leg oedema and hyper-reflexia of both upper and lower limbs, with positive babinski bilaterally. Laboratory tests revealed ESR (132 mm/h), Hb (11.5 g/dL), WBC ( $6.7 \times 10^9/L$ ), platelets ( $254 \times 10^3$  Cmm), normal liver function, cholesterol (285 mg/dL) and LDLc (150 mg/dL), 24 hour urine protein (3.7 g/d). Antibody tests revealed ANA 1/320 homogeneous pattern, anti-ds-DNA 1/40, complement C3 70 (90–180), C4 5 (10–40), positive ACA IgG, negative IgM and positive LAC. ENA's were negative. Brain MRI revealed multiple T2 and flair hyperintensities deeply seated in the parietal and the periventricular white matter and in the basal ganglia, bilaterally. Diagnosis of SLE and secondary antiphospholipid syndrome was made, along with active nephritis.





Photo: Peter Pulkowski

**Professor Annegret Kuhn, MD, MBA**  
University Medical Center Mainz, Germany

## Clinical manifestations and evaluation of cutaneous lupus

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1. Gilliam JN, Sontheimer RD. Distinctive cutaneous subsets in the spectrum of lupus erythematosus. *J Am Acad Dermatol.* 1981;4:471–5.
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Lupus erythematosus (LE) is a complex autoimmune disease that is characterised by a heterogeneous clinical presentation. Cutaneous manifestations of LE are one of the most frequent symptoms with a high burden of the disease; however, the development of a unified classification of skin lesions has proven difficult.

The classification system developed by Gilliam and Sontheimer divided cutaneous manifestations into LE-specific and LE-non-specific cutaneous manifestations.<sup>1</sup> Skin lesions such as urticarial vasculitis and livedo reticularis are some of the most common LE-nonspecific cutaneous manifestations and are primarily associated with active systemic lupus erythematosus (SLE), reflecting potentially serious complications.<sup>2</sup> The LE-specific cutaneous manifestations encompass the subtypes of cutaneous lupus erythematosus (CLE): acute CLE (ACLE), subacute CLE (SCLE), chronic CLE (CCLE), and intermittent CLE (ICLE).<sup>3</sup>

The different cutaneous manifestations of the various subtypes require a clinical score to define activity and damage of CLE. Several scores have been developed to evaluate clinical and serological manifestations of SLE, such as BILAG, ECLAM,

or SELENA-SLEDAI. Although these disease activity scores include dermatological criteria, such as butterfly rash, generalised erythema, and oral ulcers, they are not suitable for evaluating the activity of the various cutaneous manifestations of the disease.

In 2005, the first validated score with specific evaluation of skin lesions was published as “Cutaneous Lupus Erythematosus Disease Area and Severity Index” (CLASI).<sup>4</sup> This score evaluates “activity” and “damage” of the heterogeneous skin lesions of CLE by taking into account both anatomical regions and morphological aspects. In 2010, the score was revised and modified by including additional aspects of the mucocutaneous spectrum of the disease. Reliability analysis supported the validity and applicability of the “revised CLASI” (RCLASI).<sup>5</sup> Due to its detailed and comprehensive structure, the RCLASI may be applied to support the diagnosis of the various CLE subtypes and to evaluate the efficacy of treatment. Moreover, it is a valuable instrument for monitoring the disease on different sites of involvement, not only in routine clinical practice but also in long-term clinical trials.

### Learning Objectives

- Learn the classification of skin manifestations in LE.
- Score and evaluate the different subtypes of CLE.
- Understand the RCLASI activity and damage score.

[illegible]



**Professor Ian Bruce, MD, FRCP**  
University of Manchester, UK

## Decreasing morbidity and mortality and improving outcomes in SLE

### References

1. Urowitz MB, Gladman DD, Tom BD, Ibanez D, Farewell VT. Changing patterns in mortality and disease outcomes for patients with systemic lupus erythematosus. *J Rheumatol*. 2008;35(11):2152–8.
2. Bernatsky S, Boivin JF, Joseph L, et al. Mortality in systemic lupus erythematosus. *Arthritis Rheum*. 2006;54(8):2550–7.
3. Gladman DD, Urowitz MB, Goldsmith CH, et al. The reliability of the Systemic Lupus International Collaborating Clinics/American College of Rheumatology Damage Index in patients with systemic lupus erythematosus. *Arthritis Rheum*. 1997;40(5):809–13.
4. Petri M, Purvey S, Fang H, Magder LS. Predictors of organ damage in systemic lupus erythematosus: the Hopkins Lupus Cohort. *Arthritis Rheum*. 2012;64(12):4021–8.
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Mortality associated with systemic lupus erythematosus (SLE) has improved dramatically over the past 60 years. In the 1950s a 50% 5-year mortality was reported. More recently, 10-year survival is reported to be >90%. Mortality is also improving relative to the general population and the standardised mortality ratio associated with SLE has fallen from >10.0 in the 1970s to 2.5–3.5 in the new millennium.<sup>1,2</sup> As survival improves, there is increasing interest in assessing irreversible damage in SLE. The ACR/SLICC Damage Index is an excellent and pragmatic tool to assess overall long-term morbidity associated with SLE.<sup>3</sup> Damage tends to increase over time in SLE cohorts and damage predicts both further damage and also mortality.

Leading causes of death in SLE include cardiovascular disease, infections and SLE complications.<sup>1,2</sup> With regard to damage, the most common systems that accrue damage include musculoskeletal, CNS and renal systems. Factors associated with a poorer prognosis in SLE include a number of demographic characteristics

such as older age, race/ethnicity (including African ancestry) and male gender.<sup>4,5</sup> Lower socioeconomic status also adversely affects survival and risk of damage. Recent data have demonstrated that SLE factors such as persistently active disease, flares and renal involvement are all risk factors for damage and mortality. In addition, glucocorticoid use has been noted to increase the risk of overall damage and may also predispose to specific complications such as osteonecrosis, cataracts, cardiovascular events and infections.<sup>4,5</sup> In contrast, antimalarial drugs tend to mitigate the risk of a range of comorbidities and overall damage.<sup>4</sup>

In managing patients with SLE, preventative strategies should therefore include addressing better disease control and managing individual risk factors such as hypertension and hyperlipidaemia. In addition, minimising steroid use, optimising antimalarial drugs and recommending vaccinations, cancer screening, and smoking cessation programmes are all important to improve long-term outcomes in SLE.

### Learning Objectives

- Understand the increased risk of mortality in SLE.
- Describe the leading causes of death in SLE.
- Understand how the ACR/SLICC Damage Index can be used to identify key comorbidities in SLE populations.
- Describe the key 'fixed' and potentially modifiable risk factors for major comorbidities in SLE
- Develop an approach to address preventative strategies in SLE.

[illegible]



**Professor Liz Lightstone, PhD, FRCP**  
Imperial College London, UK

## Lupus nephritis: Update on modern management

### References

1. Wilhelmus S, Bajema IM, Bertsias GK, et al. Lupus nephritis management guidelines compared. *Nephrol Dial Transplant*. 2016;31(6):904–13.
2. Condon MB, Ashby D, Pepper RJ, et al. Prospective observational single-centre cohort study to evaluate the effectiveness of treating lupus nephritis with rituximab and mycophenolate mofetil but no oral steroids. *Ann Rheum Dis*. 2013;72(8):1280–6.
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Lupus nephritis is one of the most serious complications of systemic lupus erythematosus (SLE). It is common, affects those from non-Caucasian backgrounds more severely than those from Caucasian backgrounds, and remains a challenge to treat. While prognosis was much improved in the 1970s and 1980s, patients still experience high rates of renal failure. Treatment should aim to induce sustained remission, minimise treatment related toxicity and preserve fertility.

Currently the only licenced medications for the treatment of lupus nephritis are aspirin, steroids and hydroxychloroquine. None of the other treatments (cyclophosphamide [CyP], mycophenolate mofetil [MMF], azathioprine, tacrolimus, rituximab) we use regularly are licensed

for treating SLE. However, clinical trial data support the use of MMF or CyP as first-line agents for induction of remission. This talk will review the evidence underlying these treatments along with recent long term data. This talk will also explore the data supporting the use of tacrolimus for lupus nephritis and very recent work with voclosporin.

Challenges to the current approach come from the use of rituximab and other B-cell depleting agents. However, there are currently no positive trials for the use of any biologic for lupus nephritis. A key driver for change is the need to avoid the use of steroids and data supporting this will be presented along with some more novel approaches that hold promise for the future.

### Learning Objectives

- Understand why we treat lupus nephritis and the best use of renal biopsies.
- Understand the rationale and evidence underlying current treatment strategies.
- Explore how recent trials and novel therapies/biomarkers may change the treatment paradigm for lupus nephritis.



[illegible]



**Professor Andrea Doria, MD**  
University of Padova, Italy

## Treat-to-target in SLE

### References

1. van Vollenhoven RF, Mosca M, Bertsias G, et al. Treat-to-target in systemic lupus erythematosus: recommendations from an international task force. *Ann Rheum Dis*. 2014;73(6):958–67.
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3. Zen M, Iaccarino L, Gatto M, et al. Prolonged remission in Caucasian patients with SLE: prevalence and outcomes. *Ann Rheum Dis*. 2015;74(12):2117–22.
4. Franklyn K, Lau CS, Navarra SV, et al. Definition and initial validation of a Lupus Low Disease Activity State (LLDAS). *Ann Rheum Dis*. 2016;75(9):1615–21.
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Treat-to-target is a therapeutic strategy aimed at improving disease outcomes through the achievement of treatment goals, which has dramatically improved the prognosis of widespread disorders, such as hypertension or diabetes. The treat-to-target strategy has recently been introduced in rheumatoid arthritis (RA), outlining complete remission as the primary goal and low levels of disease activity as an acceptable benchmark when complete remission is not achievable.

Recommendations for treat-to-target in systemic lupus erythematosus (SLE) have recently been published and they suggest to look at clinical remission or at low disease activity (LDA), as in RA, due the detrimental effects of persistent disease activity and protracted corticosteroid therapy on the patient's outcome.

Unfortunately, the concepts of remission and LDA are far less clear in SLE than in RA; indeed, the efforts to delineate common goals have failed probably because of disease heterogeneity and lack of measurable biomarkers predicting disease course. An International Task Force has recently suggested some principles, which should guide the development of a definition of remission in SLE.

Fortunately, clinical remission is currently realistic for a greater number of patients than it was in the past, yet careful monitoring is required in order for patients to benefit from disease- and corticosteroid-free intervals, while minimising the risk of disease flares.

In everyday practice, patients should be brought to the lowest level of disease activity, ensuring a significant benefit over a persistently active disease, this being either clinical remission or LDA. However, complete remission, clinical remission or LDA should not be seen as different treatment goals, rather they should be read as different scores of the same lupus target, aiming to the highest degree of disease quiescence that can be applied to any patient.

### Learning Objectives

- Understand the impact of persistent disease activity, despite the standard therapy and corticosteroid intake, on lupus outcomes.
- Describe the concept of complete and clinical remission and LDA in SLE.
- Appreciate the prevalence of remission and LDA in lupus cohorts.
- Recognise the importance of achieving disease remission or LDA.

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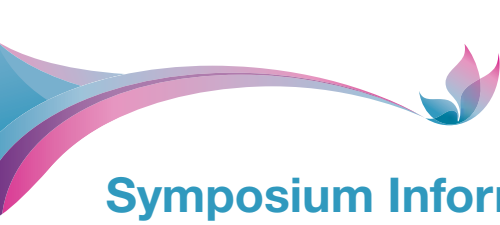
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## Symposium Information

**Date:**

Friday 9th December 2016.

**Time:**

10.00 am – 12.00 pm – GSK symposium (by invitation only)

7.15 pm – 8.15 pm – Amgen symposium followed by networking conference dinner

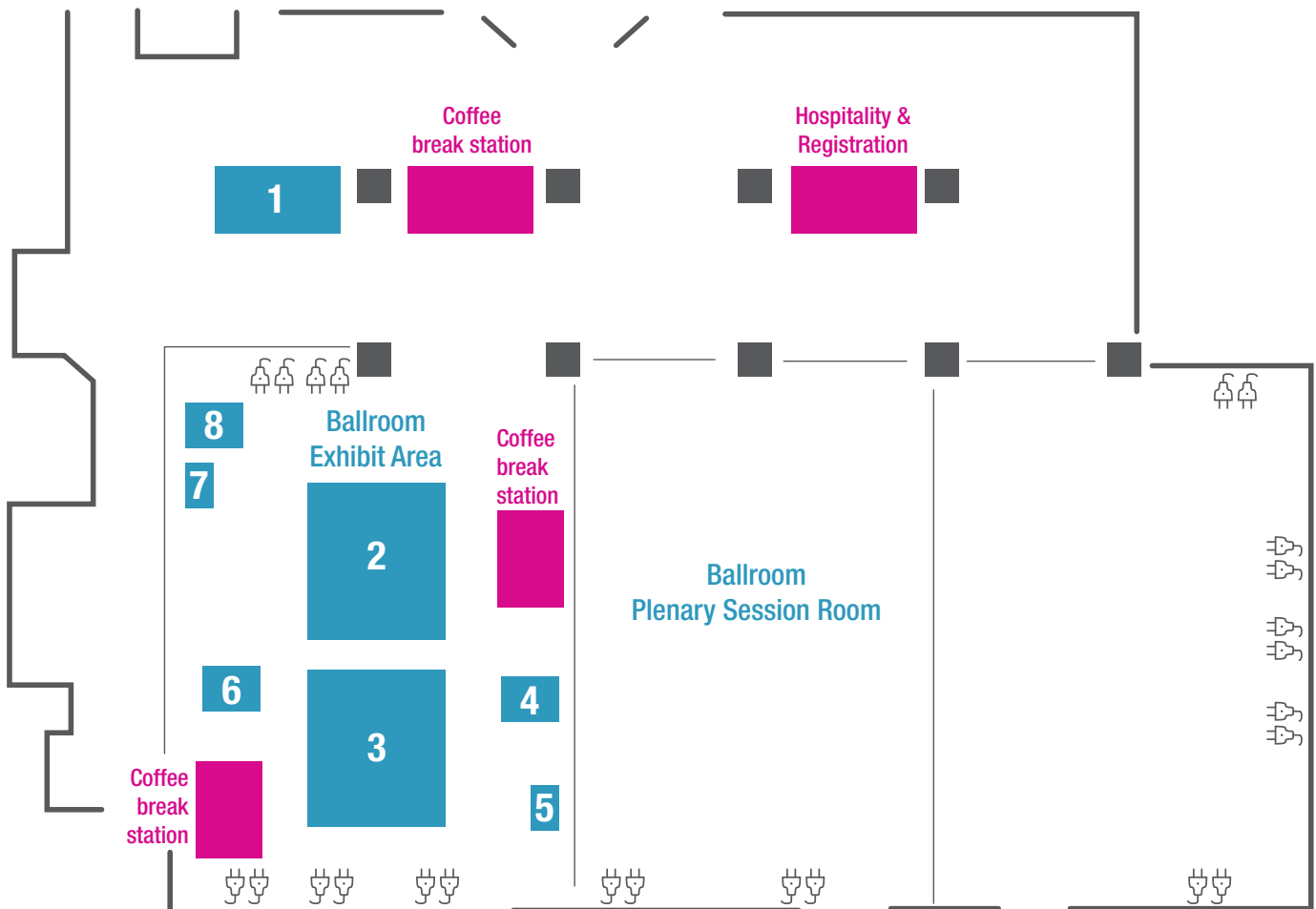
**Location:**

Crowne Plaza Dubai

Al Dhiyafah Rooms 5+6, Third Floor

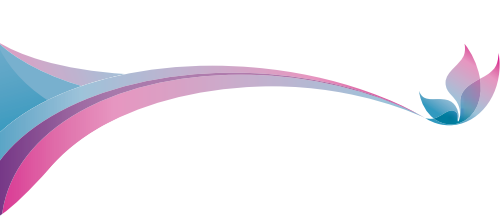
## Exhibition and Floor Plan

### Jumairah Ballroom



- |                  |           |
|------------------|-----------|
| 1. GSK (medical) | 5. BMS    |
| 2. GSK           | 6. Amgen  |
| 3. Novartis      | 7. AbbVie |
| 4. Newbridge     | 8. Pfizer |

 Power points



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