

# 10 years of UK JSLE Cohort Study Away Day

Institute in the Park, Alder Hey Children's Hospital, Liverpool, 25<sup>th</sup>/26<sup>th</sup> January

2016 is a very important year for the UK Juvenile Systemic Lupus Erythematosus (JSLE) Cohort Study as it is now coming into its 10<sup>th</sup> year! Since 2006, we have been recruiting young people to take part in the UK JSLE Cohort Study. We have come a long way, and have recruited more than 550 young people from 25 centres throughout the UK.



The UK JSLE Study Group, led by Michael Professor Beresford in Liverpool, brings together multi-disciplinary experts from across the country and representatives from all centres and includes different paediatric sub-specialists, adult rheumatologists, scientists, allied health professionals and above all patient and parent representatives.



In this time we have undertaken a wide range of research looking at the immunology of JSLE, genetics, treatments and improving the monitoring of the disease. We publish our research in various medical journals and discuss our results with other researchers around the world at conferences to help generate new ideas on how to treat Lupus.

The aim of the UK JSLE Study Group 'away day' in Liverpool was to bring together patients, doctors, nurses, scientists and researchers involved in the study to reflect upon and celebrate what the study has achieved so far. We gathered in the new "Institute in the Park" next door to the brand new £280m state-of-the-art "Alder Hey Children's Hospital"! We were delighted to have three senior representatives from LUPUS UK at the meeting: Kevin Weston (Chair), Chris Maker (Director) and Jane Dunnage (ex-Chair) to help with the discussions. Importantly, three young people with lupus and parents took part in the Away Day – their insights and experience as essential to helping us make maximum use of the meeting. We used this meeting to brainstorm what we would like to achieve over the next 10 years!



In the lead up to the meeting we were keen to get a better understanding on what research topics patients find important and to hear their views on what we are already

doing. We virtually (through Skype) attended a meeting of the LUPUS UK young persons group in London and also sent out a Survey Monkey survey to the young people who had been unable to attend that meeting.

We were struck by the fact that 11% of the individuals surveyed knew about the work of the UK JSLE Cohort Study. Starting with this report, going forward we are **very committed to letting LUPUS UK members and patients know about our research and work**. I will now take you through the four workshops we ran during the meeting, brainstorming key areas. We hope you find our discussions important and exciting plans we have for the next 10 years!



## Workshop 1 – Follow-up of patients into adult care

As part of studying lupus in children and young people, it is essential that we learn more about the long-term outcomes of their lupus, the safety and effectiveness of treatments, and what can be done in childhood to prevent complications later on. The group therefore discussed how patients in the UK JSLE Cohort Study as children/young people can be best followed up as they transition into adult care. It was agreed that this remains an extremely challenging yet crucial area. It was felt to be important, as we work to improve clinical care for patients moving from paediatric to adult care, that continued involvement in research is also considered alongside. It was acknowledged that paediatric rheumatologists aren't always familiar with adult care pathways and that it would be important to work with adult rheumatologists to define these in order to move forward. Senior LUPUS UK representatives agreed to work with the membership and the UK JSLE Study Group in defining these pathways.

The group agreed that empowering patients is important to helping research continue through transition to adult care. Education of patients about the research they are involved in could really improve continued participation. We need to engage better with adult rheumatology colleagues about the Study and produce resources which can help new adult centres get involved in and understand the importance of the Study.

## Workshop 2 – Data quality

Over the last 10 years we have collected a very very large amount of important data! To get the best, most accurate and useful results in studies it is important that the data is collected carefully and completely. Currently the study collects all data on paper forms but this has increasingly become difficult with some hospitals becoming paper free! Many studies have moved to on-line databases for data collection.

We agreed this was a priority going forward and practical ways to make it useful for everyone in our Study were considered.



Those using the forms need to understand them and we agreed the need to make educational resources, available on-line, to help those completing them.

To improve the strength of our scientific discoveries into lupus it is important that we obtain patient samples (e.g. of blood and urine) from as many different lupus patients as possible. Lupus can affect people in many different ways and it's severity can be influenced by many things. Currently we collect most of our samples from just a few centres around the UK. It was recognized as important that this area of the Study grows over the next few years. Centres that offered help included Manchester, Leicester, Birmingham, Scotland and Sheffield. To help set this up, members of the lupus research team from the LUPUS UK "Centre of Excellence for Childhood Lupus" at Alder Hey and the University of Liverpool will visit these centres to discuss directly with the relevant clinical and laboratory teams.



### Workshop 3 – Delivering ‘add on studies’

The overarching aim of the UK JSLE Study Group is to “*Improve our care and understanding of Childhood Lupus*”. The UK JSLE Cohort Study collects real world clinical data and is not set-up to do very close surveillance or treatment studies. It does however integrate very closely with other lupus studies. We therefore discussed three emerging studies and how the UK JSLE Cohort study can help to make them a success!



**Study 1 – ‘British Paediatric Surveillance Unit / LUPUS UK study of the Incidence of JSLE in children and young people in the UK and Republic of Ireland’.** This study plans to look at the incidence (number of new cases) of JSLE. Data will be collected from the medical records of all new JSLE patients over a one year period. There was a useful workshop discussing which key questions should be asked as part of this study. There was also an important discussion about how this study can be used to improve participation in and awareness of the UK JSLE Cohort Study. The

outcomes of these discussions are now being taken forward with the next stage of protocol development for this study.

### Study 2 – ‘Validation of a urine biomarker panel for lupus nephritis’

When the kidney is affected by lupus the condition is called Lupus Nephritis. Lupus Nephritis may wax and wane, but can lead to long term problems such as kidney failure. Currently, the most accurate way of diagnosing and monitoring Lupus Nephritis is by taking a kidney biopsy, and examining the kidney tissue under the microscope. However, this procedure is invasive, and has associated risks.



Over the past 7 years, as part of the UK JSLE Cohort Study we have been investigating whether biological molecules found in the urine (biomarkers) can be used to monitor LN, replacing the need for repeated



biopsies. We have identified a panel of six urine biomarkers which together accurately tell apart patients with active lupus nephritis versus those without kidney disease.

This work has been undertaken on samples from patients attending Alder Hey Children's Hospital and Great Ormond Street Hospital. The next step is to look at frequent serial samples from a larger number of patients over time to see if different combinations of this panel predict Lupus Nephritis flare-ups and whether a patient will respond to treatment. To do this we need more UK centres to come on-board with this work and hope that patients will be able to send us urine samples from home through the post! This is an exciting area which we ultimately hope to move to a urine test that can be used in clinical practice.



### Study – 3 'MASTERPLANS'

This is a national lupus study, which is in the process of being set up and led by Professor Ian Bruce, University of Manchester. The idea behind it is that lupus patients have a unique set of features; each patient differs in the severity of their disease and how well they respond to different drugs. Overall, only 40 – 60% of patients respond well to each drug. Often an even smaller proportion achieve the excellent responses we want so that they no longer have any symptoms or signs of SLE (which we call remission). Patients who do not respond well to their first treatment often need to try several others until their SLE is controlled. This study seeks to understand better which patients will respond well to particular drugs, by looking at their genetic features and chemicals (biomarkers) in the blood, urine and tissues.



To support this important study, we discussed the specific challenges



for young people taking part. This is important as if we can identify groups of patients who respond well to different drugs, we could develop tests to help doctors treat individual patients with the drug most likely to work best for them first time. The study group

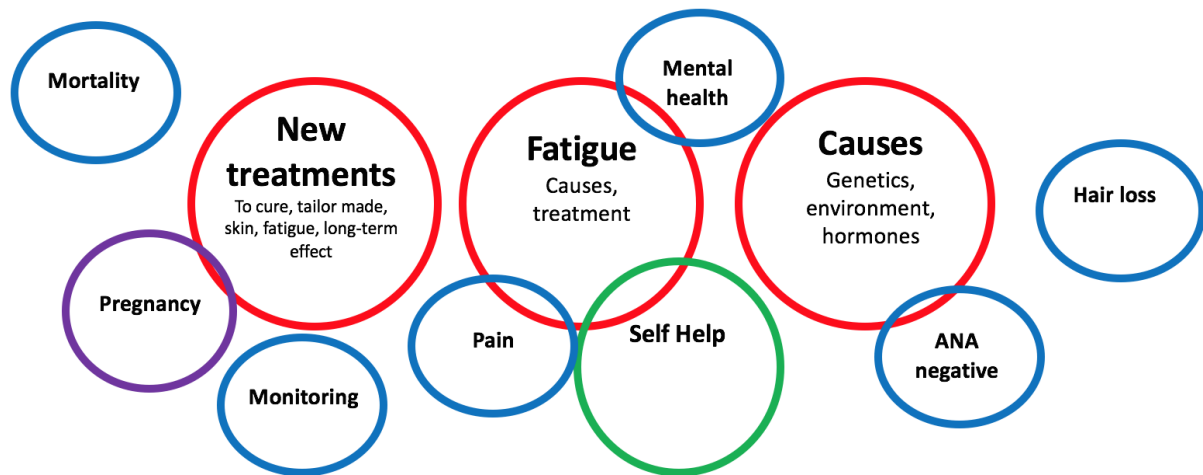
discussed the practicalities of this study involving children and young people, considering things like – how much blood is it fair to take from a young person? Would a child or young person understand this? Specific requirements for patient information sheets and consent forms. These suggestions will be fed back to Professor Bruce and the study team.

### Workshop 4 – The direction of the UK JSLE Study Group and responding to patient's research priorities!

In the lead up to the meeting we undertook a Survey Monkey survey with young people to help identify their top research priorities. In the diagram below you can see the topics that they felt to be important, with the size of the circle corresponding to the number of



people who felt that topic was a priority. Clearly the three main priorities for research highlighted the ‘causes of JSLE’, ‘new treatments’ and ‘fatigue’. We are pleased to say that over the past 10 years the UK JSLE Study Group has had a strong focus on understanding the causes of JSLE, undertaking many research studies into why the immune system attacks the body itself. By developing a better understanding of this we have also been looking for new Lupus treatment targets. There is still a lot to learn and research into causes and new treatments will remain among our top treatment priorities.



Going forward, we also plan to include a strong focus on fatigue research. The strong need for this was highlighted by young people and echoed by parents and clinicians present at the away day. We therefore held an impromptu workshop on how we can move this forward. It was agreed that it is important to think about how we can best measure fatigue in JSLE patients. Members of the lab team were keen to think about how we can investigate the biological reasons for fatigue in lupus patients, to help to understand whether it can be specifically treated by medicines. There is a local meeting for Alder Hey Children’s Hospital JSLE patients in March, and fatigue will be the focus of the day so we expect that further suggestions and ideas will emerge from this day.



We are very keen to actively involve young people in developing and carrying out some of this research and would be interested to hear from people who are keen to be involved in helping to design future studies in this area. In the first instance, our patient and public involvement co-ordinator (Mrs Jenny Preston, [Jennifer.Preston@liverpool.ac.uk](mailto:Jennifer.Preston@liverpool.ac.uk)) would be keen to hear from anyone who is interested in being involved in developing this work.



At the end of the meeting, all agreed it had been extremely productive and important meeting. Professor Beresford, Chief Investigator of the UK JSLE Cohort Study concluded: “An enormous thank you for everyone’s input and contribution to this Away

Day. Along with colleagues around the country, and very many patients and their families, we have made enormous progress over the last 10 years. The next 10 years promises to be even more important and exciting as we aim to improve still further our care and understanding of lupus amongst children and young people across the UK”



**Thanks to all who have been involved in the UK JSLE Cohort Study over the last 10 years. We are looking forward to the next 10 years and are keen to hear from anyone who would like to be involved!**

### **You can find out more about our activities at:**

- On the UK JSLE Cohort Study website
  - <https://www.liverpool.ac.uk/translational-medicine/research/ukjsle/about/>
- Or by following us on twitter
  - @EATC4Kids
  - @GenrYPAGs
  - #JSLECohortstudyawayday
- Contacts
  - Mrs Carla Roberts (study co-ordinator, [robertsc@liverpool.ac.uk](mailto:robertsc@liverpool.ac.uk))
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