



# Nephrotic Syndrome Patient Information Day

27<sup>th</sup> April 2015  
Education Centre Lecture Theatre, Birmingham  
Children's Hospital

Programme and Information on  
Nephrotic Syndrome





## Welcome!

Many thanks for taking the time to attend this patient information day, which has been kindly sponsored by the British Kidney Patient Association. We have designed the programme to give you a chance to listen to talks about Nephrotic Syndrome, network over coffee as well as pose your questions to the experts – we hope you find the day useful.

The Organisation Committee:

Moin Saleem – Professor of Paediatric Renal Medicine - University of Bristol, Consultant Paediatric Nephrologist – Bristol Royal Hospital for Children

Wendy Cook – Nephrotic Syndrome Trust (NeST) Co-ordinator

Liz Colby – Researcher at the University of Bristol

## Programme

12.00 - 1.30	Lunch	
1.30 – 1.35	Welcome	
1.35 – 1.50	Dr Asheeta Gupta (Paediatric Nephrology Registrar, Birmingham Children’s Hospital)	Introduction to Nephrotic Syndrome (Medical Perspective)
1.50 – 2.20	Simone Brown and Sam Davies-Abbott	Nephrotic Syndrome from a Patient and Parent Perspective, and Fundraising
2.20 – 2.50	Dr Judy Sutton (Clinical Psychologist, Birmingham Children’s Hospital).	Coping tricks for families and individuals living with the effects of Nephrotic Syndrome & its treatments – an interactive session
2.50 – 3.20	Coffee	
3.20 – 4.00	Melanie Dillon (Birmingham Children’s Hospital), Prof. Nick Webb (Manchester Children’s Hospital), Prof. Moin Saleem (University of Bristol, Bristol Children’s Hospital)	Clinical + Basic Science Research into Nephrotic Syndrome (RaDaR, PREDNOS2 and NephroS)
4.00	Question + Answer Panel	

The information supplied in this booklet, is correct to the best of the organisers’ knowledge. The reader is advised to consult a doctor regarding all aspects of individual health care.

## Information on Nephrotic Syndrome

### What is Nephrotic Syndrome?

Nephrotic Syndrome is a serious and debilitating kidney condition that affects around 10,000 people in the UK each year. It mostly affects children but can develop in teens and adults too. Nephrotic Syndrome isn't a disease in itself, it's a condition that can be caused by a range of different diseases which damage the glomeruli – the tiny filtering units of the kidney.

Healthy kidneys act like a sieve, allowing waste products and excess water to be excreted as urine. This process controls the body's water and salt balance, and regulates blood pressure. In Nephrotic Syndrome, there is a problem affecting the sieve mechanism of the kidney. The holes of the sieve enlarge, causing large amounts of protein to leak from the blood into the urine.

Nephrotic Syndrome is characterised by:

- Very high levels of protein in the urine (proteinuria)
- Low levels of protein in the blood
- Swelling (oedema) of the ankles and legs. Extra fluid may also build up in the abdomen and around the eyes.

Symptoms can include:

- Frothy urine
- General feeling of tiredness
- Raised blood cholesterol
- Increased risk of blood clots (thrombosis) as proteins which help prevent the blood from clotting can be passed out in the urine.
- Infections as antibodies (good proteins in the blood that help fight off infections) can be lost in the urine.
- Low levels of vitamin D.

### Nephrotic Syndrome Diagnosis

Nephrotic Syndrome is usually diagnosed by a simple dipstick urine test that confirms protein in the urine, followed by a blood test to determine blood protein levels. A kidney biopsy may also be taken, where a tiny piece of kidney is removed for examination under the microscope.

### What causes Nephrotic Syndrome?

Nephrotic Syndrome can be caused by any disease that causes inflammation of the glomeruli. Minimal change disease (MCD) is the most common cause of Nephrotic Syndrome in children (80%).

In other cases, Nephrotic Syndrome can occur as a result of a malfunction in the immune system, which could be the result of a virus, or autoimmune disorder, such as Systemic Lupus Erythematosus. Other secondary causes of Nephrotic Syndrome include certain cancers, diabetes and infections, including hepatitis B. Approximately 30% of adults with Nephrotic Syndrome will have an underlying medical problem.

Nephrotic Syndrome may also have a genetic cause. Our bodies are made from around 21,000 genes, which each have a function. You can think of each gene as an instructional code and sometimes there are errors in the code, which scientists call 'mutations'. In approximately 20% of childhood Nephrotic Syndrome cases there is a genetic fault identified. Genetics is likely to play less of a contribution in cases of Nephrotic Syndrome that develop over the age of 2, however research into genetic errors across patients of all ages will help identify how common these errors are.

## How is Nephrotic Syndrome treated?

Treatment for Nephrotic Syndrome depends upon the underlying cause.

When Nephrotic Syndrome is caused by a disease, treating that disease usually relieves the kidney symptoms.

Autoimmune disorders are treated with drugs that suppress the immune system's production of antibodies. Drugs called steroids, which reduce inflammation, may also be used to treat the damaged glomeruli. These steroids, often prescribed for several months, can prevent proteins from moving from the bloodstream into the urine. In some cases, this will be a one-off treatment and Nephrotic Syndrome may not develop again. In other more complex cases, the condition may return so you may need another course.

If the disease can't be controlled by steroids alone, other medications may be used alongside or instead, to help control symptoms. These include levamisole, cyclophosphamide, ciclosporin, tacrolimus, mycophenolate mofetil (MMF) and rituximab. The kidneys may gradually lose their ability to filter waste and excess water from the body. Kidney failure may occur that progresses to end-stage renal disease needing dialysis, or a kidney transplant. Transplants are normally successful but there is a small chance the condition could return.

Drugs may also be used to treat other side effects. Including:

- Reducing high cholesterol
- Reducing high blood pressure
- Reducing swelling - diuretics or 'water tablets' may be given to help reduce fluid-build up. They work by acting on the cells in your kidneys to make them pass out more water rather than reabsorbing water back into the bloodstream.

Most of the protein that is lost in Nephrotic Syndrome is albumin. If symptoms are severe, you may be admitted to hospital to receive albumin infusions. Albumin is slowly added to the blood over a few hours through a tube into a vein in the arm.

Immunosuppressive drugs will also be given after a transplant. These medicines lower the body's ability to reject a transplanted organ by reducing the strength of the body's immune system.

## Diet

To help prevent further water retention during a relapse, a no-added-salt diet is recommended. This means avoiding processed foods and not adding salt in cooking or at the table. The emphasis is on a healthy, balanced diet.

## Vaccines

When taking immunosuppressive drugs the immune system is weakened. Live vaccines (including MMR, chickenpox and BCG) are therefore unsuitable for anyone taking or likely to start taking immunosuppressive medication (including steroids (prednisolone), tacrolimus, mycophenolate mofetil, cyclophosphamide or regular rituximab infusions) in the near future. This also includes those who have received high dose steroids within the last 3 months.

## Chicken Pox

Chicken pox contact whilst immunosuppressed can be very serious. It is therefore important that the varicella zoster (chicken pox) immunity is known for each patient with Nephrotic Syndrome. If a patient is non-immune then a post-exposure prophylaxis may be required. Please contact your local team for advice on local policies. A chicken pox vaccination should be given at the first opportunity of the patient being non-immunosuppressed. Thought should also be given to immunising non-immune siblings.

## Useful Links

- The British Kidney Patient Association (BKPA) is the main charitable body supporting kidney services in the UK: [www.britishkidney-pa.co.uk](http://www.britishkidney-pa.co.uk)
- The National Kidney Federation (NKF) is the largest kidney patient support group in the UK: <http://www.kidney.org.uk>
- There is information about rare renal diseases available on the Renal Association Rare Renal website: <http://rarerenal.org/>
- NHS Information:  
<http://www.nhs.uk/livewell/kidneyhealth/Pages/Kidneyhealthhome.aspx>

### Additional Information for Paediatric Patients:

- infoKID is a UK-based website for children's kidney information which has information on various conditions, tests and procedures: <http://www.infokid.org.uk/>
- Great Ormond Street Hospital also have a large range of patient information on their website:  
<http://www.gosh.nhs.uk/medical-information/clinical-specialties/nephrology-information-for-parents-and-visitors/conditions-we-treat/>
- East Midlands, East of England and South Yorkshire (EMEESY children's kidney network). They have a number of patient information booklets for parents in various conditions and procedures as well as a number of story booklets to help prepare children for commonly performed procedures: <http://www.emeesykidney.nhs.uk/>

## Research into Nephrotic Syndrome

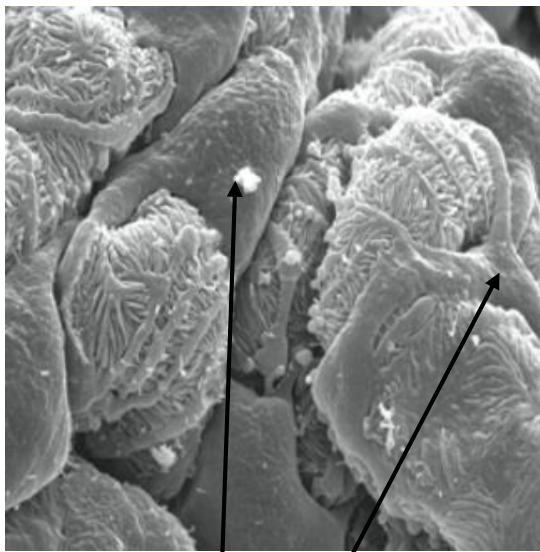
During the afternoon you will hear about research that is being carried out into Nephrotic Syndrome. If you are interested in participating in any of this research – please talk to your specialist at the hospital who can provide you with further information.

The University of Bristol has a strong team of researchers who are carrying out research to improve the management, treatment and prevention of renal disease. They are doing this by integrating laboratory research with clinical research.

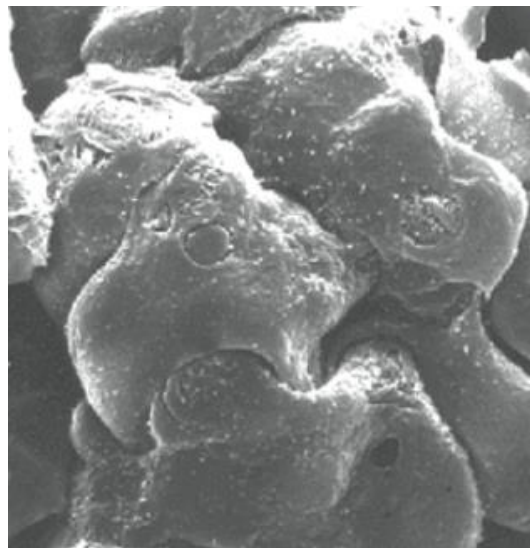
### In the Lab:

In Nephrotic Syndrome there is a breakdown of the kidney ‘filter’, which normally functions to stop blood proteins entering the urine but allows toxins and waste products to be filtered out. The cell type at the centre of this filter mechanism is called a podocyte, a highly specialised cell which is unique to the kidney.

Podocytes can be seen using a high powered microscope. They have branch-like extensions that wrap around blood vessels in the kidney, stopping the escape of blood proteins. You can see how this filter is disrupted in Nephrotic Syndrome.



**Normal**



**Nephrotic Syndrome**

(pictures from BKPA)

**Podocyte** - a highly specialised cell in the kidney. You can see the branch like structures of the cell, wrapping themselves around a blood vessel to form the filter – preventing proteins from entering the urine.

Scientists can look at the biology of these cells to understand the filtration barrier and the effects of disease states. Researchers at Bristol were the first in the world to grow these specialised kidney cells (podocytes) that make up the filtration barrier in a dish. Using this resource, scientists have been making ground breaking discoveries using a range of scientific techniques. The ultimate aim of their research is to design and study new therapies in patients with kidney disease.

### In the Clinic:

The **National Registry of Rare Kidney Diseases (RaDaR)** is an initiative by UK kidney specialists (the Renal Association). It is designed to pull together information from patients who have certain rare kidney diseases. This will give a much better understanding of how these illnesses affect people.

This work is done in partnership with patients. Where the research leads to practical benefits, such as better diagnosis, treatments or general advice this will be publicised on the project website [rarerenal.org](http://rarerenal.org). The National Registry for Rare Kidney Diseases (RaDaR) allows information to be collected from patients with rare diseases, upon their consent.

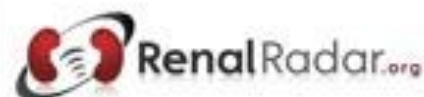
This information is used by expert groups appointed by the Renal Association. These groups are called Rare Disease Groups (RDGs). They consist of clinicians, scientists and patient representatives.

The Nephrotic Syndrome RDG is led by Prof. Moin Saleem, who heads the research group at the University of Bristol and is the lead clinician on the **National Study of Nephrotic Syndrome (NephroS)**. This study aims to understand why some patients get Nephrotic Syndrome and how the disease causes the changes in the kidney that result in the Nephrotic Syndrome. The team will look at genes that may cause Nephrotic Syndrome and see how common they are in patients in the UK with the disease as well as study blood plasma for biomarkers of disease (a factor within the blood that differs in times of relapse and remission).

Further information can be found at [www.rarerenal.org](http://www.rarerenal.org).



# *DO YOU SUFFER FROM NEPHROTIC SYNDROME?*



**If you think you suffer from Nephrotic Syndrome, we would like to invite you to take part in the NephroS study.**

**Nephrotic Syndrome is a rare disease and research is essential to improve the understanding and treatment of the disease.**

## **HOW YOU CAN HELP:**

**If you want to know more regarding this research project please contact your local Renal Research Team.**

## **WHAT WE WILL NEED YOUR HELP WITH:**

- 1. We will ask you for an extra blood sample at the same time as your routine blood samples.**
- 2. We will need to collect some clinical information.**

## **WHAT WE WON'T BE DOING:**

- 1. Ask you to come in for additional hospital visits.**
- 2. Ask you to complete forms or questionnaires.**

**PREDNOS2** is being run by Prof. Nick Webb who is a renal consultant at Manchester Children's Hospital.

### **An invitation to join the PREDNOS 2 study**

#### **Short course daily prednisolone therapy at the time of upper respiratory tract infection (URTI) in children with relapsing steroid sensitive nephrotic syndrome; the PREDNOS 2 study.**

70-80% of children and young people with nephrotic syndrome will experience relapses (i.e. the nephrotic syndrome coming back after initial treatment). There is known to be a strong link between upper respiratory tract infection (URTI – the common cold) and relapse. The PREDNOS 2 study is going to test whether taking 6 day course of steroid treatment at the time of URTI can reduce relapses occurring in children and young people with nephrotic syndrome.

The study will recruit participants for 2 years and each participant will have study visits occurring once every three months; at baseline, 3, 6, 9 and 12 months. To enter the study, children and young adults must be aged over 1 year and under 19 years of age and need to have suffered two disease relapses in the preceding 12 months. Patients who have participated in the PREDNOS study will be able to enter PREDNOS 2. The study is supported by the NIHR Clinical Research Network: Children, has been funded by the NIHR Health Technology Assessment Programme and is sponsored the University of Birmingham and the Central Manchester University Hospitals NHS Foundation Trust.

We do very much hope that you/your child would be interested in participating in PREDNOS 2 if your child meets the entry criteria. We require a total of 300 patients nationally and over 100 centres across the UK are taking part. Further information and a list of participating UK centres is available on the PREDNOS 2 website ([www.birmingham.ac.uk/prednos2](http://www.birmingham.ac.uk/prednos2)). If you are interested in taking part in the study please speak to your doctor when you next attend a clinic appointment.

Yours faithfully

Prof Nick Webb

PREDNOS 2 Chief Investigator, Royal Manchester Children's Hospital, Oxford Rd, Manchester M13 9WL UK Email: [nicholas.webb@cmft.nhs.uk](mailto:nicholas.webb@cmft.nhs.uk), Tel: +44 161 701 2961, Fax: +44 161 701 2630



# PREDNOS 2

## Do you, or does your child have relapsing childhood nephrotic syndrome?

**Children and young people aged over 1 year and less than 19 years who have relapsing nephrotic syndrome may be eligible to take part in the PREDNOS 2 study.**

70-80% of children and young people with nephrotic syndrome will experience relapses. There is known to be a strong link between upper respiratory tract infection (URTI – the common cold) and relapse. The PREDNOS 2 study is going to test whether taking 6 day course of steroid treatment at the time of URTI can reduce relapses occurring.

The PREDNOS 2 study is recruiting 300 children and young people with relapsing nephrotic syndrome from over 100 hospitals across the UK.

Could you or your child be one of them?

If you are interested in taking part in the study please speak to your doctor when you next attend a clinic appointment

Or for more details please visit [www.birmingham.ac.uk/prednos2](http://www.birmingham.ac.uk/prednos2)

Or for more details email Prof Nicholas Webb at [nicholas.webb@cmft.nhs.uk](mailto:nicholas.webb@cmft.nhs.uk)

PREDNOS 2 is funded by the NIHR HTA Programme

## **A Message from our Sponsors: The British Kidney Patient Association**

[www.britishkidney-pa.co.uk](http://www.britishkidney-pa.co.uk)



***The British Kidney Patient Association are absolutely delighted to play our small part in supporting this Nephrotic Syndrome Patient Information Day. We hope that you have an informative, helpful but also an enjoyable day.***

***Paddy Tabor***

***CEO, British Kidney Patient Association***

The British Kidney Patient Association is the leading UK charity, which for nearly 40 years has been providing support to kidney patients and working to improve health and social care services for patients and their families. With over 40 patients supported every week, and around £3 million invested every year in research, funding health and social care professionals and improving hospital services, we are committed to improving the quality of life for everyone affected by kidney disease.

We do this by:

- Providing grants to help patients in times of financial hardship
- Campaigning to inform policy and practice to improve the lives and choices of kidney patients
- Funding kidney units to improve services and patient care
- Providing information, advice, counselling and support services
- Commissioning research to improve health and care services.

## The Nephrotic Syndrome Trust (NeST)

<http://www.nstrust.co.uk>



You may have heard this charity mentioned by Simone Brown and Sam Davies-Abbot who gave talks on a patient/parent perspective of Nephrotic Syndrome. NeST also supports the Bristol-based team researching into Nephrotic Syndrome. Its work includes researching causes of Nephrotic Syndrome, and ultimately, to find a cure. Money raised by NeST is used to fund this vital research

“The Nephrotic Syndrome Trust (NeST) was set up in 2005 by trustee David Yearsley, whose son James suffers from the disease. Its main aims are to:

- Raise awareness of the disease
- Raise funds for research to find a cure
- Offer a website resource and forum for sufferers and their carers/families who often feel isolated. Please check out our Facebook page – which is a buzzing place full of people sharing their stories and experiences, building up a network of support that you can access any time.”

Notes...