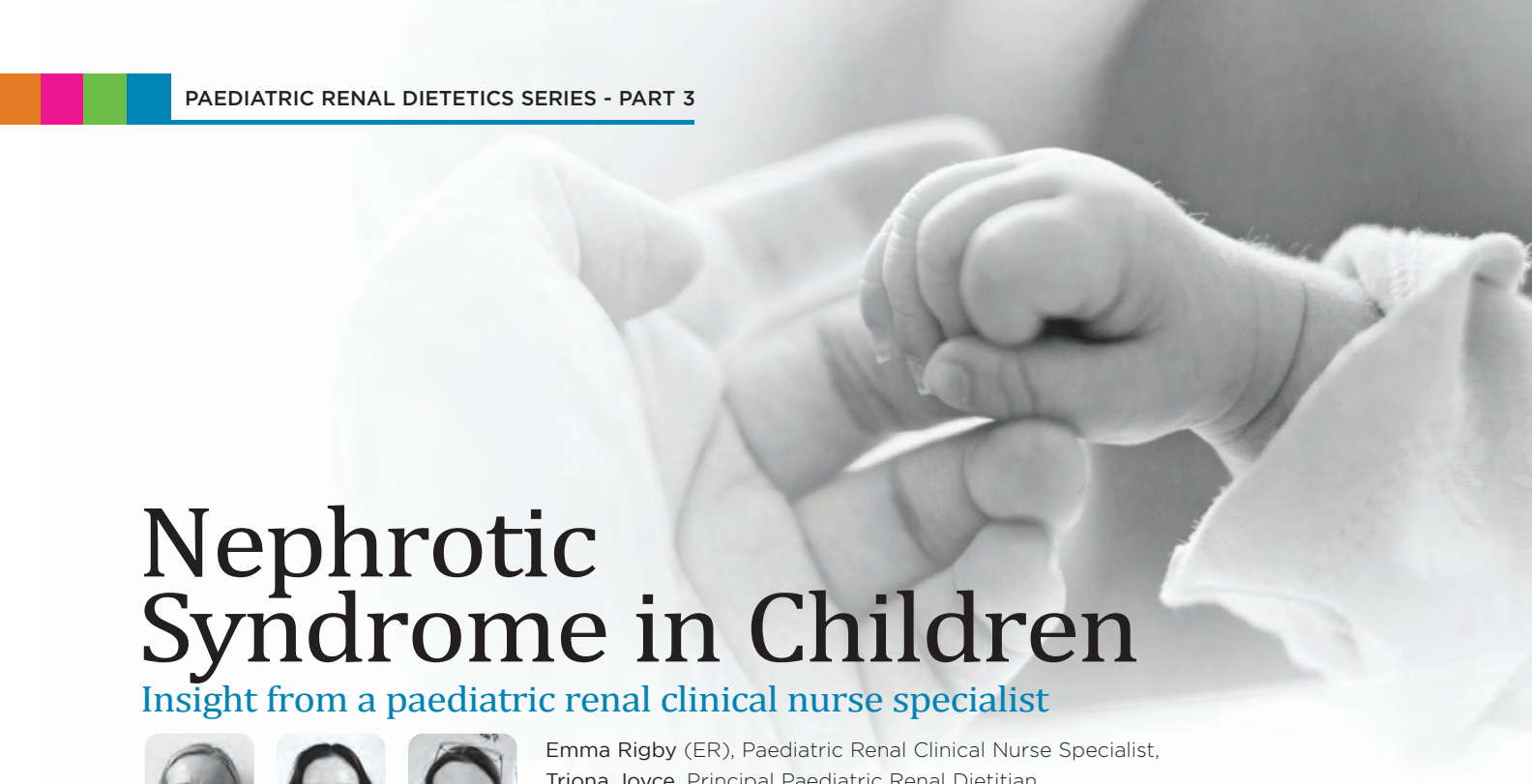

# Nephrotic Syndrome in Children

## Insight from a paediatric renal clinical nurse specialist



**Emma Rigby** (ER), Paediatric Renal Clinical Nurse Specialist,
**Triona Joyce,** Principal Paediatric Renal Dietitian
& **Tara Hall,** Senior Paediatric Renal Dietitian, Evelina London Children's Hospital, UK

Idiopathic nephrotic syndrome, which is one of the most common renal conditions in children, is known to newly affect 1 to 3 per 100,000 children per year.[1] Although it is a common syndrome seen by paediatric nephrology teams, there is a low incidence amongst the general paediatric population.[2] The overall incidence of idiopathic nephrotic syndrome, which varies by region and ethnicity, has been reported as ranging from 1.15 to 16.9 per 100,000 children.[3] The Evelina London Children's Hospital has one of the largest dedicated paediatric nephrotic syndrome outpatient clinics in the UK, with over 260 children with complex nephrotic syndrome managed by this service. However, as most children with idiopathic nephrotic syndrome will receive input and management from general paediatricians, paediatric dietitians in primary and secondary care settings may receive referrals for dietetic input for this patient group. Paediatric Renal Clinical Nurse Specialist, Emma Rigby, who manages the dedicated paediatric nephrotic syndrome outpatient clinic at the Evelina London Children's Hospital, provides an insight into this syndrome. This article focuses on idiopathic nephrotic syndrome (no known cause) and not nephrotic syndrome linked with other conditions (e.g. lupus, glomerulonephritis).

### How do children usually present with idiopathic syndrome?

ER: Children tend to present a couple of weeks after a viral illness, with parents/carers often initially noticing paleness, lethargy and puffy eyes. Most commonly parents/carers will take their child to the GP and quite often the initial impression will be an allergy and an antihistamine will often be recommended. As the nephrotic syndrome progresses, parents/carers will notice increasing oedema, in which case they will either bring their child back to the GP or their child will be referred to their local hospital for further investigations. The key characteristics of idiopathic nephrotic syndrome are the presence of protein in the urine (proteinuria), low blood albumin levels (hypoalbuminaemia) and/or the presence of oedema.[1] Idiopathic nephrotic syndrome often starts between the ages of 1 and 4 years and is more common in boys than in girls.[4]

### What is the first line treatment for idiopathic nephrotic syndrome?

ER: The first line medical treatment used in the management of idiopathic nephrotic syndrome is with the immunosuppressant prednisolone (steroids). The International Pediatric Nephrology Association recently published clinical practice recommendations for the management of children with idiopathic nephrotic syndrome,[1, 3] including a standard initial starting dose for prednisolone. The minimum prednisolone duration is 8 weeks and dosage/regimens are individualised thereafter depending on the patient and hospital protocols.

## Are all children with idiopathic nephrotic syndrome managed by specialist paediatric renal centres?

ER: Most children with idiopathic nephrotic syndrome will receive medical input and management from general paediatricians. However, those that are more complicated to manage are referred to specialist paediatric renal centres. Children tend to be referred to a paediatric nephrologist if there are additional issues other than urinary protein losses. For example, if there is the presence of both blood and protein in the urine, if the child has hypertension, or if the child is slightly older. For the latter, children above the age of ten should be referred to our nephrotic syndrome clinic because it is unusual to present at that age. Most children with nephrotic syndrome (approximately 85%) will experience complete remission to prednisolone treatment.[1]

## What are the main medical concerns with idiopathic nephrotic syndrome?

ER: Once diagnosed the main concerns relate to controlling the protein loss and treating the side effects of the leaking of protein. These side effects include those associated with oedema and the protein loss, such as increased risk of infection, increased risk of blood clots[1] and the risk of hypovolemia if not managed and treated. In addition, when trying to control the protein loss, children receive immunosuppressants which further increases their risk of infections. Some children may need to be admitted to a tertiary hospital with specialist paediatric renal services for intravenous albumin. In most cases fluid restrictions are not required, and it is known that inappropriate fluid restrictions could lead to an acute kidney injury, hypovolemic shock and thrombosis (blood clots).[3]

## What do you find are the main challenges for parents/carers?

ER: One of the most difficult aspects of idiopathic nephrotic syndrome for parents/carers is the unpredictability of this syndrome. We are unable to predict what might trigger relapses, how their child is going to respond to the medications used, and how many relapses their child is going to experience throughout their childhood or into adulthood.

Steroids are known to have an effect on increasing appetite and can cause behavioural changes.[2] From my experience, families often find this increase in appetite alongside the challenging behaviour due to the steroid medication difficult to manage. Often if their child is demanding a particular snack – whether this is biscuits, sweets, chocolate – if giving this helps manage the behaviour, appetite and also calms the situation, then that's quite often what families will do. I often talk about the importance of distractions, for example, going for a walk, playing a game to help with the behaviour and having fruit and vegetables easily available as a snack, to help encourage this from the start.

When their child eventually achieves remission and the steroids are weaned/stopped, their child's behaviour and appetite returns to normal and understandably parents/carers often worry about restarting this medication again during additional relapses. Another side effect of the steroids that families are concerned about is the effect on their child's weight. The steroids make you store fat differently; therefore, it needs to be explained to families prior to starting that children can get a round face but that this is normal. If we don't see that happening, then this is a concern and we would worry about compliance with taking the steroid medication.

## What do you find are the main challenges from a clinical nurse perspective?

ER: From a clinical nurse perspective, the main challenge is managing parental expectations. It is important to prepare and equip patients/carers with the coping strategies to help them with managing the unpredictability of idiopathic nephrotic syndrome and ensuring that they have a good understanding of this syndrome. I find that if parents have a good understanding of idiopathic nephrotic syndrome they are better able to be more proactive and reduce problems before they progress further. Ensuring that parents are monitoring their child's health regularly and know who they should contact if any concerns is important, so that any potential issues can be identified early. Some useful information resources that I direct our parents/carers to include the online resources available on both the infoKID and the Nephrotic Syndrome Trust (NeST) websites (www.infokid.org.uk & https://nstrust.co.uk).

## What changes would you like to implement into paediatric nephrotic clinic services?

ER: The ideal dedicated paediatric nephrotic syndrome clinic service, for me, would involve an initial multidisciplinary team (MDT) appointment at referral for every child with a new diagnosis of idiopathic nephrotic syndrome. This would include a paediatric dietitian, a paediatric renal clinical nurse specialist, a paediatric renal consultant and a psychologist. I believe that if you start off with good habits initially, and help prepare parents/carers to manage potential behaviours on steroid medications, it could help make the management of nephrotic syndrome relapses easier.

## What dietary advice is important for children with idiopathic nephrotic syndrome to receive?

TJ & TH: In their recent clinical practice recommendations, the International Pediatric Nephrology Association recommended that a dietitian should provide advice to patients and families on dietary salt intake.[1, 3] To help in the management of oedema, it is recommended that excessive salt intake should be avoided,[1] with normal salt intake, in line with dietary salt intake recommendations for the general paediatric population, advised during remission.[3] There are excellent resources available to help support with this education including video and written resources on the Action on Salt website (www.actiononsalt.org.uk) and the paediatric-specific 'All about salt' video on the My Renal Nutrition website, produced by Vitaflo International Ltd (www.myrenalnutrition.com).

A common dietetic query is whether children with nephrotic syndrome require additional dietary protein intake above the recommended nutrient intake levels. As highlighted by the International Pediatric Nephrology Association, there is insufficient evidence to recommend an increase in dietary protein intake in children with nephrotic syndrome.[2] The current recommendation for children with nephrotic syndrome is that dietary protein intake should be the same as for the general paediatric population.[3]

It is important for the MDT to discuss the importance of regular physical activity alongside encouraging a healthy balanced diet. The International Pediatric Nephrology Association has made a strong recommendation for supporting regular physical activity to help prevent thromboembolic events during relapses, prevent potential weight gain whilst receiving prednisolone treatment, and to help prevent the loss of both muscle and bone mass.[1] In addition, the International Pediatric Nephrology Association have recommended ensuring adequate dietary calcium intake in children with steroid sensitive nephrotic syndrome[1] as part of their advice addressing preservation of bone health.

It is known that dyslipidaemia occurs during the initial presentation of idiopathic nephrotic syndrome and during relapses.[3] The lipid abnormalities in idiopathic nephrotic syndrome are thought to be mostly due to impaired clearance of lipids.[5] As dyslipidaemia usually resolves during remission, routine lipid-lowering agents (e.g. statins) are not recommended during relapses.[1] In children with both steroid resistant nephrotic syndrome and persistent raised blood low-density lipoprotein cholesterol levels, lifestyle changes including dietary modifications, enhanced physical activity and weight control in those that are overweight, has been suggested.[1]

Interestingly, whilst a small proportion of children with nephrotic syndrome may require multiple immunosuppressants (e.g. prednisolone, tacrolimus, mycophenolate mofetil and rituximab) and as a result could be more immunosuppressed than children post renal transplant, there is a lack of evidence and guidance on providing food safety advice to these children.

## Summary

In summary, idiopathic nephrotic syndrome is one of the most common renal conditions in children. As the majority of children with idiopathic nephrotic syndrome will be looked after by general paediatricians, paediatric dietitians within primary and secondary care settings have an important role in providing dietetic input and support for these children and their parents/carers. Dietetic advice should be individualised to each child and should focus on following a healthy balanced diet. Dietary advice to support with the avoidance of excessive salt intake at initial presentation is important with normal salt intake, in line with dietary salt intake recommendations for the general paediatric population advised during remission. With regards to protein intake, the current recommendation for children with idiopathic nephrotic syndrome is that dietary protein intake should be the same as for the general paediatric population.

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