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Family Reflections Series

Family Reflections: Living with, but not defined by: Posterior urethral valves, chronic kidney disease and kidney transplant

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My son is a caring, energetic and clever 3.5 year old. He has learned to ride a bike, has done his first ballet show, now attends pre-school and we've just submitted applications for primary school. They are milestones which to us demonstrate that regardless of his health needs he will be who he wants to be. He will not be defined by his health.

BACKGROUND

My son was born with posterior urethral valves (PUV), a blockage in his urethra that prevented him from urinating. This blockage meant during pregnancy urine built up in his bladder. The volume was so great that pressure built inside his bladder forcing the urine back into the kidneys, distending the bladder, both ureters and both kidneys. At birth, he required resuscitation and special care. At 2 days of age, a blood test detected an abnormal creatinine and an ultrasound confirmed the presence of valves and kidney damage.

He was transferred to a specialized Neonatal Intensive Care Unit for treatment and then to a Specialist Paediatric ward. He was catheterised and had a double nephrostomy inserted to drain the bladder and kidneys. He was attached to multiple machines and his blood work indicated that although the pressure in his system had been relieved the damage caused resulted in chronic kidney disease (CKD). The extent of the damage meant that alongside medication he required tailored milk low in potassium and he had to drink a very specific amount each day; the milk was made up of 2 powders, water and either breast milk or formula milk. After an initial 6 weeks, my son was discharged in a heat wave and returned to hospital for a further 2 weeks as he was dehydrated. My son was diagnosed with Stage 5 CKD and at this point we knew he would need a kidney transplant in childhood.

At 6 months, my son was started on Oxyboutin to relax his bladder; however, it relaxed it too much. It prevented him from voiding properly and the reflux into the kidneys began again. He had surgery to create a ureterostomy (stoma created from a ureter), which drains his left kidney directly by-passing the bladder. For him, this hugely helped and kept him stable for a while. At this point, we psychologically began to prepare that he would need a transplant in the next few years.

The core parts of my son's care following his initial hospital stay were directed towards treating and maintaining kidney function. Treatment involved a large fluid target, specific nutritional input, medication and regular monitoring. Pre-transplant, he had a very specific diet, breastfeeding was possible; however, because of his level of kidney damage and the high potassium content of breast milk, the breast milk needed to be diluted with different powders and water. I was encouraged to breastfeed on demand, but the

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knowledge of the impact of potassium on my son made me feel I was poisoning my child. To add context, it was also an incredibly physically and psychologically gruelling 3 months of every 2–4 h bottle feeding (taking an hour), expressing (taking an hour) and starting over again.

I suspect that even at a young age my son felt the pressure of drinking enough milk. He would vomit frequently and at 11 months flatly refused milk so we started weaning early. To wean a child with CKD is difficult. Many foods were excluded from his diet or had to be cooked in a specific way to remove the potassium. CKD and the medication to manage it makes children thirsty, to a point, but also has a side effect of reduced appetite. To be eligible for transplant, children need to be above specific weight and size, and to get to this point is difficult when a child's nutritional input is so specific. The kidneys are responsible for growth and so when they are damaged, they need lots of care to grow. In some cases, children need dialysis and what I've heard from others, this can be a very gruelling process for patients and families. To put the impact of working kidneys into context, pre-transplant my son needed monthly erythropoietin injections and at 2.5 years old and just over 10 kg. Post-transplant, he is now 3.5 years old and weighs 18 kg.

In 2019, we started the build up to transplant. My husband was assessed so that he could be a living donor to our son. At the start of 2020, we thought the transplant would be in June 2020, but then coronavirus disease (Covid) hit and the donor programme was shut down. While the transplant programme was paused, my son's condition worsened. Discussions were had about a deceased donor. The clinical team did everything to get the transplant back up and running and in September 2020 my son received my husband's kidney. The experience of paediatric kidney transplant is hard and within the context of Covid it is incredibly isolating. My son and husband were in separate hospitals and because of Covid restrictions couldn't see each other after surgery and recovered separately. Every emergency admission following transplant has been 1 parent only. We live with an emergency bag in the car, knowing that any change could mean a dash to hospital. Multiple lengthy emergency admissions are hard on a patient and families, in particular when you are unable to get any support beyond the hospital. For the families who have experienced this situation, I am sure they will be impacted. Research into how best to support this group and inform them of appropriate precautions/provisions in future would be essential. Post-transplant, an assumption is that my son's health is 'fixed'. Please be aware a kidney transplant isn't a solution, it is a treatment option, my son needs to manage his health every day and it is likely my son will need another transplant in the future. My son and we as a family live with the uncertainty of his health every day.

TREATMENT AND IMPACT

Both before and after transplant, my son's everyday treatment plan involves fluid, good diet, medication, regular monitoring and now catheterisation. As before, treatment involves the management of kidneys and bladder. The impact of this is that all aspects of daily life, any activity such as meeting up with friends or attending nursery/ pre-school/school, comes with a risk assessment and a caveat to cancel. We do not tell my son what he's doing until the day we do it; the reason is to prevent him from being disappointed and feeling different. He also has many routine clinical visits and scans, alongside any emergency appointments, all of which can change the trajectory of a day.

In the first year of life, the impact of his diagnosis, various hospitalisations and care plan meant that the amount of time he spent with other children and in groups was limited. The uncertainty of his condition made it difficult to explain to friends and family and that too became isolating. We attended baby signing classes to try and support communication and attended what we could. However, some days it was overwhelming and we couldn't strike a balance. He started nursery at 10 months but this was limited and multiple infections and hospital admissions meant that the option of nursery was limited as well as expensive. We have made a conscious effort to support him to get the most from life and make friends and attend groups, but everything has come with caution and the potential for it to change on the day.

When Covid hit, we shielded as a family until 3 months post-transplant. During the year following transplant, he had urosepsis twice, his transplanted ureter calcified and he had many more admissions. Approximately 1 year post transplant, things started to settle properly and, after a false start with childcare he was able to start socialising and attend pre-school. He loves being around other children, and he loves learning. As he gets older, he is more aware of his differences and the physical scars of what he has been through, at times this affects him more than others. Sometimes catheters are difficult as he is frustrated that he needs them and others don't, he asked to make a traffic light for the bathroom door to prevent others coming in whilst he was having an intermittent catheter; at 3.5 this is a heart breaking insight into how he feels. As a child he is learning and growing, it would be great if there was research and support to help support children with all of these big emotions, those stemming from infancy as well as from the health needs of an enduring health condition. He will also need more involvement from medical teams throughout his life. He will need appropriate provisions in school and he will always need to make a risk assessment and take appropriate precautions.

WHAT SHOULD BE RESEARCHED?

There is some incredibly exciting research being done in PUV and recently the first International Posterior Urethral Valve conference was held by Evalina Children's Hospital London, with the aim of supporting clinicians internationally to give equal care to all children with PUV. The British Association of Paediatric Nephrologists Clinical Study Group too are committed to quality research for children with renal problems, including linking those with urological and renal comorbidities. I mention these two groups, because in addition to being committed to research they are committed to including both the patient and caregiver in the research experience. I've been fortunate to be able to give input to them. However, more needs to be done.

PUV are found in 1 in 8000 boys but often clinicians in local hospitals do not know about it nor have the knowledge to give the right information, diagnosis or treatment. This means that throughout the world there are children and families who have this diagnosis and feel totally isolated, overwhelmed and scared by what is to come and local clinicians who do not understand it. I've been told the valves are there antenatally but not always detected. I wonder if this is the case for other conditions, and so surely there is justification generally for more frequent antenatal screening?

There are many practical everyday aspects of my son's care that I feel are not addressed or understood. As an example, at one point my son had 9 daily timepoints to take medicines and this was alongside a fluid target of 2 l and catheterizations every 2.5 h. In between all of these activities was making meals and doing daily activities. This hectic schedule reduced his chances to be a fun-loving child. My plea for researchers is to consider the everyday experience of children with medical complexities and look into sustainable, appropriate and manageable approaches to care. As an Occupational Therapist myself, I would certainly make the case for Occupational Therapists to be involved with negotiation of complex health management and the everyday. Considering things in this way would support a child going to school and taking ownership over their health. Clearer and unified (international) advice needs to be given about potty training; incontinence and potty training are used in conjunction and these terms seem completely opposite.

As you read this, you may think I'm discussing the urological side of things, and the reason for this because the kidneys and bladder are connected organs, what happens to one can affect the other. Bladder management is crucial to kidney management; however, the specialists are separate and the organs are treated separately. Therefore, when issues with either the bladder or the kidneys are explained to patients, their care is explained separately. In my opinion, this approach does not work and is confusing. In recent months, the link between renal and urology appears to be improving but I think research needs to be done in understanding how best to explain the complexities of bladder and renal conditions to patients and caregivers. Otherwise, the learning curve is steep, confusing and further isolating.

Please change the narrative around PUV and CKD. It is often so fatalistic which massively impacts a child's and caregiver's mental health. Please do research into what is known about PUV and CKD in order to learn what is known and what needs to be relayed. Share it with all hospitals, sonographers and the general public so that risk factors are known and that treatment can be considered earlier. Please learn and share knowledge about this condition, connect families, understand their everyday activities and share rough treatment plans so patients and families can prepare and fill the child's childhood with positivity. PUV and CKD can be comorbid with other conditions, things such as blood pressure and cardiac function is regularly monitored. Post-transplant my son developed an anaphylactic reaction to bananas; pre-transplant he was not allowed bananas. The relationship between allergies and immunosuppression doesn't seem to be known. Research in this area would be very helpful.

So, to finish, the practical, realistic everyday management of the care of babies with PUV varies between patients. Having support will make diagnosis and the possibilities for the child so much better. Please do more research into the everyday of a child with PUV and generate frameworks that make a treatment plan manageable and understandable; if non-compliance comes to mind ask 'Why? Is it because the treatment plan isn't realistic?' then ask 'How can we make it more appropriate for this child?'. Co-produced research would be absolutely wonderful, in particular research involving play so that every child has a voice. The narrative around PUV and CKD is sometimes fatalistic and not all of the information is given at once. Please be open and collaborative with patients and families as the more people share their understanding, the more understanding there is.

COMPETING INTERESTS

I have started to write a blog called The Hidden Everyday (www.thehiddeneveryday.com). There wasn't anything to help my son manage his ureterostomy, so I started making belts to support this and was encouraged to sell them. I've started to write the blog to share knowledge and support others as a parent. I started volunteering for Kidney Research UK as a parent research representative and help out where I can to make sure the patient/carer voice is represented in research.

ADDITIONAL INFORMATION

Correspondence and requests for materials should be addressed to Lucy Wray.