PAEDIATRICS

A NATIONAL MODEL OF CARE FOR PAEDIATRIC HEALTHCARE SERVICES IN IRELAND

CHAPTER 32: PAEDIATRIC NEPHROLOGY

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ROYAL
COLLEGE OF
PHYSICIANS
OF IRELAND

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32.0 INTRODUCTION

The national paediatric nephrology programme has evolved over the past 30 years, and now provides high-quality tertiary paediatric nephrology care to children with kidney problems in Ireland. The service has met the challenges of an increasing birth rate, increased survival of children with complex problems, as well as the demands of technological and surgical advances.

Paediatric nephrology is a key paediatric tertiary specialty providing expert care for children with diseases of the kidney and urinary tract. Paediatric nephrology integrates directly with paediatric urology – a tertiary specialty providing surgical intervention for the correction of abnormalities of the kidney and urinary tract – and with the kidney transplant team who provide the surgical expertise to support a kidney transplant programme directed by paediatric nephrologists.

Disease Incidence and Prevalence

Renal diseases are rare in children, with incidence and prevalence varying depending on the condition. Examples of diseases the service deals with include the following:

- 100 children in Ireland have received a kidney transplant in childhood in the past 10 years.
- 60 children with a kidney transplant are under active follow-up in the clinic.
- Each year, eight children a year develop end-stage renal failure requiring dialysis/transplantation.
- 22 children are currently waiting for a kidney transplant.
- Each year, 25 children develop haemolytic uraemic syndrome.
- 200 children in Ireland have nephrotic syndrome.
- Each year, 1,000 children are born with a congenital abnormality of the kidney or urinary tract.

32.1 CURRENT SERVICE PROVISION

The paediatric nephrology service to children in Ireland includes:

- acute assessment
- management of chronic disease
- dialysis
- and, ultimately, renal transplantation for children who require it

32.1.1 Acute Care

Paediatric nephrology is highly specialised, and involves an acute model of service delivery. Conditions encountered by the service often involve acute and critical illness in children. The expertise required to deal with kidney conditions suffered by children is not available in the adult nephrology services, and is not a part of routine paediatric training.

The national paediatric nephrology service cares for approximately 750 new outpatients per year. In addition, 2,500 review patients are seen in clinics annually. Clinic waiting times vary according to the basis of the clinical indication. For example, children with acute illness requiring urgent review are seen almost immediately; children requiring routine review may have to wait up to one year. Approximately 30% of outpatient referrals are from

Dublin-based general practitioners (GPs), and work is currently taking place with colleagues in general paediatrics and general practice to try and better define referral pathways, and also to provide guidelines as to how some of the more common paediatric nephrology conditions might be managed in the community.

Conditions which require direct immediate access to specialist paediatric nephrology care include:

- Haemolytic uraemic syndrome (HUS) – a complication of infection with E. coli 0157, a common bacteria encountered mainly through contact with cattle or via food/drink contamination. Ireland has a high incidence of HUS, which is associated with the dairy industry. (Germany has 60 cases reported nationally each year; the comparable figure for Ireland is 20 cases). HUS occurs predominantly in children, and presents with acute renal failure and multi-organ dysfunction. Treatment is supportive, and involves dialysis in approximately 50% of cases. Outcomes in Ireland are excellent, compared with international reports: indeed, our death rate is particularly low in this regard.
- Acute kidney injury (AKI) – caused by a variety of illnesses and often occurring as part of multi-system disease. AKI is an immediately life-threatening loss of renal function that requires expert assessment as to the underlying cause. Assessment includes specialist imaging and investigation, which may include a kidney biopsy. Children with AKI may require dialysis.
- Nephrotic syndrome – a kidney condition associated with loss of protein in the urine. Nephrotic syndrome was universally fatal prior to the development of treatment with steroids. Although treatable, this condition can be very dangerous, especially around the time of initial presentation. Approximately 20% of children respond well to initial treatment and can be managed in the community; the remainder can be difficult to treat, and require expert input. Shared care links have been developed with many of the paediatric units in Ireland, in order to manage children with more complex types of illness.

32.1.2 Long-term Care

In addition to providing acute care, paediatric nephrology provides care for children with a wide range of long-term illnesses that affect the kidney. Congenital abnormalities of the kidney and urinary tract are the commonest form of congenital abnormality seen in children internationally – occurring with greater frequency than abnormalities such as developmental dysplasia of the hips or congenital heart malformations. Ireland has a high incidence of children born with congenital abnormalities, and the service has significant expertise in the management of babies born with such conditions. Potential interventions include operative intervention, radiological intervention, and dialysis.

Other conditions that require access to specialist paediatric nephrology care include:

- chronic/complex glomerular disorders
- vasculitis
- tubulointerstitial disorders, including renal tubular transport disorders that are primary or secondary to acquired or metabolic disease
- complex/severe hypertension
- nephrolithiasis
- complex neuropathic bladder, particularly in children requiring other specialised services
- extremely rare diseases such as cystinosis or atypical HUS

32.1.3 Dialysis

The national paediatric nephrology service provides a dialysis programme for children throughout Ireland. Where possible, dialysis is provided in the community using overnight continuous cycling peritoneal dialysis, which can be performed in the child's home. Where home-based dialysis is not possible, a dedicated hospital-based paediatric haemodialysis service is provided.

32.1.4 Kidney Transplantation

Over the past 10 years, the national paediatric nephrology programme has taken over the provision of renal transplantation for children in Ireland. The results of the paediatric kidney transplantation programme are exemplary, and compare well with the best international standards. Over the past five years, as the demand for paediatric transplantation has increased, the National Paediatric Renal Transplant Programme has worked closely with Beaumont Hospital to develop a live related renal transplant programme to enable the parents and close relatives of children with end-stage renal failure to receive a pre-emptive renal transplant. Where children are already on dialysis, work is taking place to facilitate live related donation, in order to minimise the time a child spends on dialysis.

32.1.5 Transition Programme

Structured transition from the care of paediatric services to adult services is vital for children with chronic illness. A paediatric transition programme has been developed over the past five years, and a process is underway to develop partnerships with adult nephrology teams in Cork and Beaumont to formalise these arrangements. Outcomes for young adults with chronic illness are significantly improved with the provision of robust transition programmes.

32.2 PROPOSED MODEL OF CARE

The following are the key features of this proposed model of care for paediatric nephrology:

- National tertiary service
  - access through secondary care
  - consultation at tertiary level

- Single-site focus with strong regional links
  - a central hub with outpatient and inpatient care delivered on a single site
  - integrated access to paediatric urology, interventional radiology and transplant surgery
  - development of regional clinics and link paediatricians, in order to support secondary care
  - clearly established home care packages for chronic and end-stage renal failure
  - establishment of a national home haemodialysis programme for children

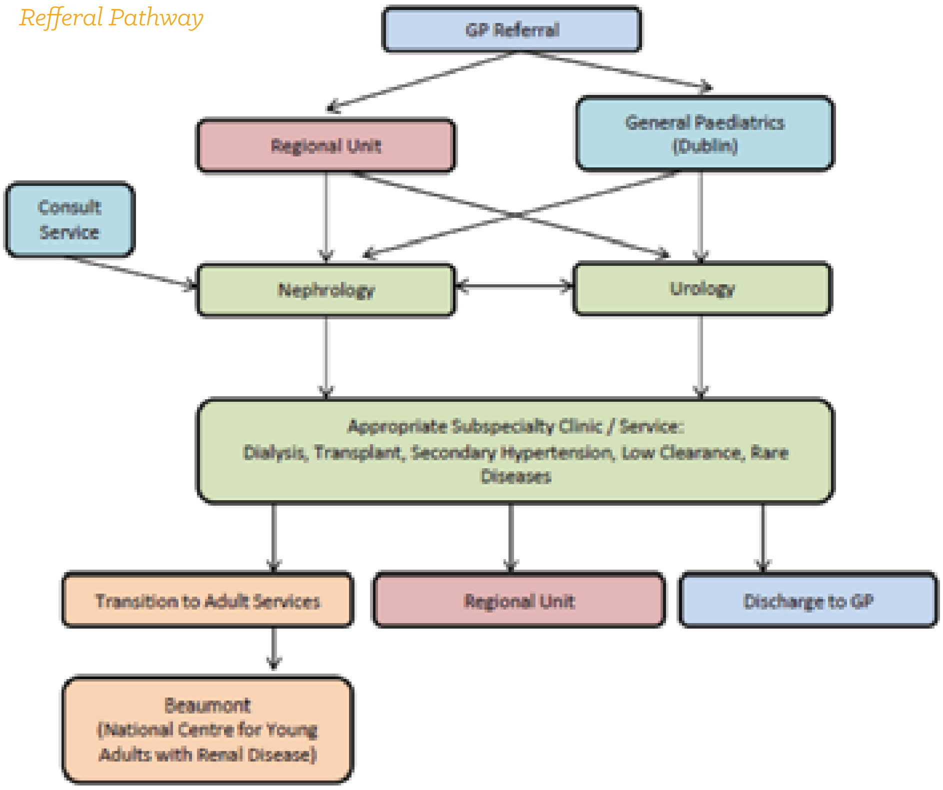
- Standardised shared care pathways
  - support for community-based primary care through the development of a nephrology network, education and guidelines
  - implementation and use of agreed referral pathways with secondary care

- Structure to support dependencies
  - access to support services to include specialist paediatric radiology, specialist paediatric laboratory, renal histopathology, vascular surgery, transplant immunology
  - co-location with services requiring specialist input from paediatric nephrology, such as paediatric intensive care, paediatric cardiology, paediatric oncology, paediatric haematology
  - multidisciplinary care of the young person transitioning to adult nephrology services

- Consultant-delivered
  - Inpatient and outpatient care needs to be delivered directly by consultants.
  - appropriate number of consultants to provide a full on-call rota for paediatric nephrology, paediatric urology, and a full paediatric renal transplant programme
  - service planning to recognise the shortage of specialist staff at a national and international level
  - development of a fellowship programme to support training

- Supported by specialist staff
  - support and develop the roles of senior specialist nurses to foster leadership, development and communication across the service
  - dedicated ward and outpatient staff with highly specialised skill mix
  - specialist renal nursing training for all nursing staff
  - development of specialist clinical nurse specialist (CNS) and advanced nurse practitioner (ANP) roles to support provision of specific haemodialysis, peritoneal dialysis, hypertension, specialist voiding disorder and transplant clinics
  - focused areas of care for health and social care professionals: renal dietitians, psychological support for chronic illness/body image issues/organ transplantation
  - Coordinate the establishment of a national plasma exchange service which provides treatment for children and young people supporting all specialities

- Quality driven/evidence based/research oriented
  - The national paediatric nephrology programme continues to innovate and to adapt in a complex and evolving medical environment. Recent developments include the use of monoclonal antibody-based therapies to treat atypical haemolytic uraemia syndrome and children with complex forms of nephrotic syndrome.



Refferal Pathway

Multidisciplinary Support

A well-developed, appropriately staffed and highly trained multidisciplinary team (MDT) is essential for the delivery of high-quality care to children with kidney disease. The following table highlights the involvement of key members of the MDT in the outpatient setting:

For every 100 children seen by a consultant nephrologist in clinic:

- 66 will also see a non-consultant hospital doctor.
- Two will also see another consultant.
- 39 will also see a clinical nurse specialist.
- Four will also see a clinical psychologist.
- 18 will also see a renal dietitian.
- Eight will also see a social worker.

For every 10 consultant appointments, there will be 14 additional visits with members of the MDT on the day of the clinic

Key members of the nephrology MDT include:

Child Psychologist

The diagnosis of a chronic renal illness, particularly one that is going to lead to the need for renal replacement therapy and transplantation, impacts on every aspect of a child or young person's life. The impact is felt by parents, carers, siblings and the extended family. The effects also extend to the child's social and educational life. The demands of the treatment regimens are arduous, and the burden of care is high.

The number of patients with chronic and end-stage renal failure continues to increase worldwide. Notably, the complexity of these patients has also increased significantly. At present, a number of patients have had previous kidney transplants previously, with consequent complications. Many have other medical or developmental disabilities. The majority of these children have had disease from early childhood, which is a likely indicator for a higher incidence of psychological adjustment problems. As many of these children are tube fed, oral aversion and feeding difficulties are common. Important areas for therapeutic psychological input also include adherence to necessary treatment issues, and support for the child's parents and siblings.

Many psychological, coping, stress and allied problems accompany chronic kidney disease in children. Disruption to school and education, chronic illness, poor growth, altered body image, renal replacement interventions and invasive medical procedures all contribute to considerable psychological challenges for children and families burdened by chronic renal failure.

Key issues/areas	Clinical psychology interventions
Adherence and concordance issues	The majority of children with end-stage kidney disease have had disease from early childhood, which is a likely indicator for a higher incidence of psychological adjustment problems. Psychological support will include adherence/concordance issues, anxiety, as well as support for parents and siblings. Clinical psychology has an important role in the prevention of treatment adherence failure and in the identification of children at risk, as well as the preparation and management of children and families.

Transplantation programme	A key area for psychological support is around psychological screening and transplant preparation. This is often a time of high anxiety for children and parents, and disruption of mood and behavioural and adaptation difficulties can occur. Informed, developmentally appropriate understanding and expectations of the transplant process is crucial.
Transition to adult services	Research has shown that up to one-third of transplanted kidneys may be lost, due to non-adherence within the first few years following the transition from paediatric to adult nephrology services. Many surveys have highlighted a substantial decrease in the quality of healthcare following patients' transfer to adult services. Therefore, it is crucial that a process of preparation and a high level of support are considered key in preserving graft function as well as good psychological functioning and well-being for young people.
Urogenital abnormalities	Psychological support for children born with complex urogenital abnormalities is a crucial area for development, as there is a high incidence of psychological adjustment difficulties and mood disorders in this group, particularly with the onset of puberty and throughout adolescence and young adulthood.
Feeding problems	Many children with kidney disease have disrupted feeding. Some are tube fed prior to achieving an acceptable weight for transplantation. Children with long-term feeding difficulties are among the most difficult to treat, both from a medical and psychological perspective. As a result, this can be an area of immense stress for families. The role of clinical psychology is multi-faceted, with an emphasis on both prevention and treatment.
Teamwork	The psychologist contributes to discussion and consultation on both routine and complex cases. There is a role for skill sharing and upskilling with other team members, as well as providing a psychological perspective on cases.
Research	Psychology has a significant role to play in future research, particularly with regard to psychological and emotional adjustment, coping and resilience factors in children and young people with acute kidney disease, chronic renal conditions, as well as in children requiring dialysis and, ultimately, renal transplantation. Targeted research on these topics could lead to significant savings in health costs for this cohort by identifying high-risk groups, and reducing the impact and costs associated with poor treatment adherence, as well as reducing the risk of failed transplants.

The National Service Specifications for Renal Services which were adopted in the UK (2013) describe the core standards that need to be in place for any provider in order to ensure safe and effective care and excellence in the field. In relation to renal dialysis (haemodialysis), it is recommended that psychology services should be available to all children, as required. The British Psychological Society bases its workforce recommendations on both population size and population needs. Acute and chronic renal failure in children is characterised by 'low-density but high-intensity work', i.e. the number of children affected is small, but they have complex needs. The caseload and complexity characteristics in renal healthcare are similar to other chronic health conditions, such as cystic fibrosis. Here, the recommendation is 0.4 WTE consultant clinical psychologists per 50 patients. It is important to bear in mind that in the UK regional services are far more developed and better resourced in terms of shared care, whereas in Ireland regional services for children and young people with chronic kidney disease are almost non-existent outside of Our Lady's Children's Hospital, Crumlin (Crumlin) and the Children's University Hospital, Temple Street (Temple Street). The National Renal Strategy Review commissioned by the Department of Health and Children (2006) recommended two WTE senior paediatric psychologists, in order to adequately support the national paediatric nephrology programme and the kidney transplantation service.

Renal Dietitian

The renal dietitian plays an essential role in the treatment of children with kidney disease – promoting optimal growth by ensuring adequate calories and micronutrients; controlling symptoms such as uraemia by moderating protein intake; and ensuring safety by helping to control potentially life-threatening variations in electrolyte and fluid intake. There is good evidence to show that good dietary control in children with chronic kidney disease can slow progression of the disease. Children with chronic kidney disease can pose a significant dietetic challenge – the aim being to maximise quality of life in the face of nutritional management, which often needs to be aggressive in order to achieve optimal nutritional status and acceptable biochemistry. The challenge of providing practical and age-appropriate advice means that a significant input of time is required for each child seen, and active follow-up is essential in order to keep up with developing milestones in eating behaviour.

The dietetic care delivered to children with chronic kidney disease is highly specialised and includes:

- maintaining growth in chronic kidney disease
- potassium restriction
- detailed protein/phosphate balance
- fluid restriction
- management of polyuria
- management of nephrocalcinosis
- chronic kidney disease – bone mineral disorder
- salt restriction in relation to hypertension
- management of renal tubulopathies
- long-term feeding disorders including oral aversion
  – in collaboration with the speech and language therapist
- nutrition in the context of dialysis/end-stage renal failure
- vitamin D metabolism in chronic kidney disease
- metabolic acidosis
- weight management in post-transplant patients

Competing aspects of care can pose the biggest challenge, e.g. poor growth in the face of strict fluid restriction makes delivering calories to a child who cannot eat solids very difficult. Paediatric nephrology relies heavily on expert dietetic input to provide safe and effective care for all children with chronic and end-stage renal failure. Renal dietetics is highly specialised, deals with critical safety issues, and needs to be primarily delivered by senior dietitians.

Play Services

The renal unit play service is a vital component of the care offered to children with kidney disease who are cared for by the renal service in both inpatient and outpatient settings.

The play service aims to:

- engage with children to provide support, encouragement and reassurance
- provide age-appropriate play, specific to a child's development level and medical condition
- assist with procedures, by providing play and distraction, both during and after such procedures
- identify fears and help children master negative feelings by developing appropriate coping strategies
- ensure that children understand their illness and treatment
- support families
- participate in the assessment of the holistic development needs of the child

The service plays an important role in providing consistent support to children and their families, building up relationships over time, and making children with chronic life- threatening illness feel special.

The provision of play programmes for children on haemodialysis is paramount. Haemodialysis sessions occur on three or four days a week for each child. These children have to travel to hospital, often over considerable distances, and then sit still for between three and four hours for the duration of their haemodialysis session. As such, haemodialysis is extremely tedious, even for older children, and it is very important that children have something nice and worthwhile to look forward to after their dialysis session. The play service develops programmes of activity that are individually tailored to children on haemodialysis. One of the roles very much appreciated by children attending the unit is the recognition of milestones such as birthdays and achievements, and the celebration of events such as holidays with activities and decorations.

The renal unit play service specialises in helping children to cope with painful procedures such as phlebotomy, which are unavoidable in the care of children with chronic kidney disease:

- Preparation empowers the child to gain some control over a potentially frightening procedure.
- Distraction therapy focuses on modifying the brain's perception of pain and enabling children to manage their anxieties.
- Post-procedural play helps to draw a line under a procedure and helps children to relax.

Relationships built up over time can be important in allowing communication with young adults who have moved beyond the realm of 'play' in isolation. The renal unit play service continues to be involved in the support of many of young adult patients who have issues such as medication compliance, body image and sensible lifestyle choices in the context of renal disease.

Play is central to the healthy growth and development of the child, and is recognised as a basic human right of all children (UNCRC, 1996, Article 3) and the value of play for children in hospital is an intrinsic part of their care and treatment. Despite the obvious benefits, the provision of play for children in Irish hospitals is largely volunteer-led and the number of dedicated hospital play specialists currently employed falls short of the recommended level. The proposed model of care aims to support the renal unit play service to expand and improve the level of care offered to children with kidney disease.

Renal Social Worker

Renal social workers offer services for children and their families who attend the renal unit on both an outpatient and inpatient basis. The social work service recognises the fact that a child's renal illness and subsequent attendance at a hospital can be a stressful time for families, and offers both practical and emotional support with the aim of minimising these difficulties. The renal social worker's primary focus is the emotional and social impact of illness and hospitalisation on the lives of patients and their families. The aim is to provide a holistic service to patients and their families.

Key issues/areas	Social work interventions
Childhood illness	• It is recognised that a diagnosis of a long-term renal condition or illness impacts on the whole family system. Support and counselling is offered to children, parents and families who have received a new renal diagnosis. Assistance is provided to families to make contact with local services which may offer support following the child's discharge, e.g. local HSE services or specialist voluntary services. • Children living with a long-term renal illness face their own difficulties and challenges. Specific attention is given to children who are on haemodialysis or peritoneal dialysis. These children and their families need regular long-term support in order to deal with the impact that this treatment regimen has on the child and their family. • A serious diagnosis may have an impact on the patient's siblings. Children are likely to have a number of concerns relating to their brother or sister's attendance at the hospital, the patients' long-term prognosis, and how the illness/condition will affect family life in general. The siblings of children who attend the hospital on an ongoing basis may feel alienated from the hospital. In order to address this, we have established a special sibling camp has been established to be held on an annual basis. The aim of this project is to provide an opportunity for siblings to spend some time in the hospital, to learn more about their sibling's condition and to receive therapeutic support.
Bereavement	• In the event of the death of a child, a support and counselling service is offered to families, both at the time of the bereavement and on a follow-up basis. Parents are offered one-to-one support and assistance on both an emotional and a practical level. Parents are also invited to attend parent support groups organised by the department. These therapeutic groups provide an opportunity for bereaved parents to meet and offer support to each other. Bereavement groups for siblings are also run; these groups facilitate an expression of feelings, and help children to normalise their loss experience.
Child welfare and protection	• From time to time, families may have difficulty caring for their children, or difficulty in meeting all their needs. Counselling and support is offered to families who have problems relating to family separation, homelessness and relationship difficulties. We accept that the child's welfare is paramount and we work closely with parents, other colleagues and statutory authorities where these issues arise. Information on social welfare entitlements is also made available to parents.
Pre-transplant preparation	• The renal team aim, whenever possible, to ensure that a child is transplanted pre-emptively. The team recognise that a structured programme needs to be in place in order to ensure that parents and children are given all necessary information and support before the child is called for the transplant. It is the role of the renal social worker, as part of the wider renal MDT, to be part of this structured pre-transplantation preparation programme.

As the renal unit plans to expand its services, a new senior social worker position will be required in the department. Two social workers will be needed in order to maintain the existing service available in the department, as well as develop new services such as the pre-transplant preparation programme. It is also hoped that some children may be able to start on home haemodialysis. Delivering support to these families will be extremely important to the long-term success of this treatment, and home visits may be needed.

Education

Chronic renal failure impacts significantly on children's ability to learn both physically and socially.

- Physical impacts on children's education include:
    - low energy related to anaemia
    - low attention span and nausea related to the build-up of toxins in the blood, which are normally cleared by the kidneys
    - short stature
    - physical restrictions related to lines/transplant
    - poor exercise tolerance

- Social impacts on children's learning include:
    - the effect on school attendance due to regular hospital visits
    - the need for medications to be taken at school
    - physical differences from other children in terms of lines or scars
    - restrictions on engaging in sports activities
    - continence problems

Children with end-stage renal failure face the additional burden of fitting dialysis into their lives.

The majority of children with chronic renal failure access education services through the community. The hospital-based education service provides support for children with chronic renal failure by:

- ensuring effective communication between stakeholders, including the renal team, parents, social workers, psychology, local schools, home schools and the Department of Education and Skills
- engaging in multidisciplinary collaboration with professionals to ensure that the development of the whole child is considered during a hospital stay
- continuing education and minimising the disruption caused by repeated or long-term admissions
- establishing links with both the hospital community and the wider community
- aiding transition to adult services

Effective hospital-based schooling is essential for children on haemodialysis who will spend three days a week in hospital. For these children, a hospital-based education service:

- allows primary pupils on haemodialysis to access their right to education by providing a full curriculum of carefully graded educational activities in line with the 1999 primary school curriculum
- enables post-primary pupils on haemodialysis to access their education and continue their Junior and Leaving Certificate programmes
- ensures a vital sense of normality for children and young adults with medical needs
- provides an appropriate learning environment
- facilitates post-primary pupils to sit State exams by providing an exam centre under the auspices of the
- State exam centre.

Service levels for the hospital-based education service are determined by the Department of Education and Skills. There is currently one primary teacher; one post-primary teacher covering Mathematics, English, History, CSPE, and Music; and one shared post-primary teacher covering Irish and French. The education service also facilitates mathematics assistants from Trinity College Dublin, and supports outreach programmes in music, art and science. Currently, the principle challenge to providing education to children with kidney disease is the lack of dedicated facilities. It is planned to develop a satellite renal classroom near the nephrology ward, which will:

- allow education to be tailored to the needs of children who are immuno-suppressed
- facilitate on-site monitoring of blood pressure/fluid restrictions
- avoid acute mobility problems that limit educational opportunities, i.e. following transplant
- allow flexibility around the child's medical condition
- bring stability/normality into children's lives and reduce isolation
- facilitate the delivery of high-quality education
- provide an education service that is adapted to the pupil's individual needs and abilities
- accommodate pupils using wheelchairs, drip stands and other medical apparatus
- provide a space for group learning, shared reading and cooperative teaching socialisation is a high priority and teachers make every effort to normalise the learning situation

In addition, we are currently working to ensure that pupils have access to the Schools Broadband Service provided by the National Centre for Technology. This service is filtered specifically for the educational needs of pupils.

32.3 REQUIREMENTS FOR SUCCESSFUL IMPLEMENTATION OF MODEL OF CARE

32.3.1 Infrastructure

Immediate

- To actively develop the renal wards in Crumlin and Temple Street, including the provision of:
  - wireless access for patients
  - entertainment systems
  - murals and child-friendly decor
  - clinical and office space for staff
- In order to secure adequate systems to monitor renal patients and ensure safe and effective communication, there is a plan to develop IT-driven systems. We are working to develop the paediatric component of the National Renal Database which it is anticipated to be in operation in Temple Street in 2016 and in Crumlin in 2017. Improved IT resources are essential, in order to enable the monitoring of morbidity and outcomes in this complex patient group.

Medium term

- To build a new outpatient facility in Temple Street
- To redevelop the haemodialysis unit, in order to facilitate infection control requirements
- To redevelop the nephrology ward in Temple Street, in order to provide en-suite cubicles appropriate for the care of children with kidney disease in a comfortable and safe environment

Long term

- To move to a single integrated base within the main site of the new children's hospital
- Facilities should encompass:
    - 16 inpatient beds
    - an adequately sized haemodialysis unit comprising a four-station bay and two isolation bays with flexibility to increase isolation capacity, as needed, in the future
    - a specialised nephro-urology, transplant and dialysis area compromising eight individual consultation rooms, a conference room and a multidisciplinary base for a minimum of 10 staff
    - a four-bed day ward – this facility will facilitate assessment and treatment of children with renal disease under the direct supervision of the team of nephrology and urology clinical nurse specialists. The day ward will reduce the need for inpatient admission and streamline the delivery of complex multidisciplinary review/therapies.
    - adequate access to theatre facilities to support renal biopsies; dialysis access procedures; catheterisation of children for urodynamic assessment; and renal transplantation.
- Paediatric urology services need to be incorporated into the plans for the new children's hospital. Departmental space should include office space for the multidisciplinary team; an appropriate outpatient space and four dedicated urodynamic assessment suites, each equipped with toilet facilities which can be accessed by a child/young adult in a wheelchair. The urodynamic assessment suites should be designed with privacy for children and young adults as a primary focus.

32.3.2 Staffing

Immediate

- Adequate consultant numbers to ensure continuous and safe cover for acute paediatric nephrology at a national level
    - A consultant-delivered service is currently provided across a split site. There is no dedicated middle grade cover and consultants are frequently called throughout the night. Consultants attend the hospital during the night on a regular basis to assess critically ill children and to support the busy transplant programme. With increasing pressure on the renal service and inadequate resources there is concern that levels of clinical risk will be significantly elevated in the context of a reduced daytime consultant presence. To maintain the current level of service and on-call activity in a safe and structured manner requires an urgent review of our rota arrangements and the appointment of two new consultant nephrologists.

- To ensure a full complement of specialist nursing staff
    - Funded posts are required to be filled and full maternity cover to be provided in all key areas.

- To continue to develop our multidisciplinary team in accordance with best international practice
    - A full-time psychologist is required to provide support to children and families in dealing with chronic life-threatening illness, which has a huge impact on daily living. In addition, psychological support is vital for children who have received a transplant – particularly in relation to medication compliance.
    - A data manager is required to facilitate data capture and enable the unit to compare outcome results with international registries.

- Support and develop paediatric urology services
    - Paediatric urology is a highly specialised surgical field closely allied with paediatric nephrology. At present there are 1.2 WTE paediatric urologists working in Ireland; this number is extremely inadequate. Waiting lists for urology are unacceptably long and the primary focus is on dealing with emergencies.

- As an acute surgical specialty, the current number of paediatric urologists in Ireland is unsustainable and unsafe. Acute urological emergencies in childhood include dealing with obstructed and infected urinary systems and complex congenital malformations where drainage options are limited.
- Ireland has one of the highest incidences of live births with spina bifida in Europe. Paediatric urology is a key component of the care of children with neuropathic bladders. Access to the appropriate level of specialist urology review for children with spina bifida is currently unacceptable.
- Paediatric urology services need to be developed. A minimum number of four consultant paediatric urologists are required, along with support staff, to include six urology clinical nurse specialists, a psychologist, a social worker and four secretaries.

Long term

- In the longer term it is anticipated that full staffing for the new renal unit to comprise: consist of:

MDT member	**WTE**	**MDT member**	**WTE**
Consultant nephrologists	8	**Renal nurses:**	
Paediatric transplant surgeons	3	CNM 3	1
Live related transplant co-ordinator	1	ANP	2
Dietitian	3	CNM 2	2
Clinical technologist	1	CNM 1	4
Social worker	2	Staff nurse	26
Pharmacist	2	Clinical nurse specialists	6
Psychologist	2	Clinical education facilitator	2
Play therapist	2	Haemodialysis CNM 2	1
School teacher	1	Haemodialysis CNM 1	2
Secretarial/Administration	6	Haemodialysis staff nurse	3
Data manager	2		
Paediatric urology / Neuropathic bladder:			
Consultant paediatric urologist	4		
Urology clinical nurse specialist	6		
Urology psychologist	1		
Urology social worker	1		
Secretarial/Administration	4		

32.3.3 Strategic Requirements

- Nurse education programme with backfill of posts, in order to allow specialist training in paediatric nephrology/urology
- Dublin City University advanced renal nursing course
- Support the development of CNSs and ANPs in areas to include nephrotic syndrome; hypertension; haemodialysis; and specialist disorders of bladder function
- Support the ongoing growth and development of the live related and pre-emptive renal transplant programme
- Continue to support the appointment of a new consultant urologist and urology nurse specialist

- Secure HSE funding for novel and expensive treatments for atypical HUS (e.g. Eculizumab)
- Further improve the transition programme for adolescents
- Encourage active participation in research/data collection/trials, and continue active audit and research
- Develop clinics in Cork, Galway, Letterkenny, Limerick and Waterford (regional adult dialysis centres)
- Reduce outpatient waiting times to 90 days for new patients
- Further develop transition services for young adults and review the care pathways pertaining to their progress into adult services, including weight management and healthy lifestyle implantation post-transplant, in order to optimise graft outcomes; and to develop a national transition programme for young adults with chronic kidney disease
- Develop a national paediatric home haemodialysis service for children who are likely to face a considerable wait for renal transplant due to unfavourable HLA typing or high panel reactive antibody titres
- Expand involvement in translational research – particularly in the areas of post-transplant lymphoproliferative disease, the early detection of acute renal insufficiency, and in drug trials.
- Work to improve renal outcomes for children with spina bifida in Ireland
- Work to have paediatric nephrology recognised as a subspecialty by the Irish Committee on Higher Medical Training (ICHMT) and the Medical Council
- Incorporate our service into the European Network for Rare Diseases – allowing us to build on our current network of expert support, and share experience and expertise with other European centres
- Services for children with enuresis in Ireland are inadequate, with an over-reliance on tertiary specialist referral for problems that could be dealt with at a community level. Problems with enuresis are distressing to children and their families – if not dealt with in a timely and appropriate manner. Complications of abnormal bladder habit can result in infections and more serious continence issues in later life, particularly in adult women after childbirth. At present, nephrology/urology services are not staffed or equipped to meet the demands on services to assess and treat children with enuresis, and referrals for treatment are being refused. If a community-based solution to the requirement for paediatric enuresis services is not forthcoming, then it is proposed the development of a National Childhood Continence Service is run by our combined services. Staffing would require the appointment of six consultant paediatricians with a special interest in paediatric continence and a team of 30 CNSs to run a nurse-led assessment service for childhood continence problems.

32.4 GOVERNANCE

Clinical governance arrangements in the department are as follows:

Education and Training

All of the consultants are registered with the Royal College of Physicians of Ireland (RCPI) for CME and have up-to-date certificates of compliance. Both sites are actively involved in local and national education programmes, and are recognised sites for specialist registrar training.

Clinical Audit

The department is compliant with the Irish Medical Council's requirements for ongoing audit. There are rolling audit projects covering areas such as renal biopsy yields and complications, line infection and peritonitis rates, and vitamin D supplementation in haemodialysis. An audit of our outpatient letters is being developed in an effort to optimise use of limited secretarial resources. In addition, rates of weight and height gain post-transplant in relation to dietetic interventions are being examined.

Clinical Effectiveness

Data is currently being collected on line infection and peritonitis rates. Performance is monitored against internationally agreed standards (KDOQI). We have recently reviewed our peritonitis protocol in terms of efficacy and safety, and changes were made subsequently which it is hoped will optimise diagnosis and treatment. The renal dietitians have developed specific paediatric renal dietetic competencies for their profession with their colleagues in the UK, and are in the process of developing clinical outcome measures.

Openness

Nephrology services are actively involved in hospital risk reporting in both Crumlin Temple Street. There is an excellent record in terms of patient care, and we strive always to reduce risk. In addition, there is active involvement in quality improvement programmes.

Risk Management

Monthly cross-city meetings are held, which have a teaching theme but also provide a forum for discussion on developing cross-city guidelines, reviewing protocols, and discussing ethical issues or adverse events.

Information Management

Flow charts of specialist nephrology patient results are kept in Crumlin and Temple Street. Weekly meetings are held in Crumlin and Temple Street to review critical results. We are engaged with the National Renal Office on the development of a national renal database, which we hope will be introduced in 2016.

32.5 RESEARCH

Current research projects include:

1. Prevention of post-transplant lymphoproliferative disorder (PTLD) by development of EBV-specific cytotoxic T cell assays and analysis of lytic versus latent gene expression of EBV in post-renal transplant patients. The research team comprises Atif Awan, Michael Riordan (Children's University Hospital Temple Street, Dublin), Julie Moran (UCD), Jeff Connell (UCD), Jaythoon Hassaan (Conway Institute, UCD).
2. Urinary biomarkers of acute kidney injury in paediatric oncology patients. The research team comprises Michael Riordan, Michael Capra, Atif Awan, Niamh Dolan and Mary Waldron, Crumlin.
3. Increased PD-1 expression and monofunctional EBV-specific CD8+ T cells in paediatric renal transplant patients with chronic high Epstein-Barr viral loads. The research team comprises Atif Awan and Michael Riordan, Temple Street, and Julie Moran, Jeff Connell and Jaythoon Hassaan (UCD).
4. Function of exosomes secreted by EBV-infected cells and their potential role as a biomarker in transplant patients. The research team comprises Atif Awan, Michael Riordan, Niamh Dolan and Mary Waldron, Temple Street, and Jeff Connell and Jaythoon Hassaan (UCD).
5. Clinico-pathological features of C3 Glomerulopathy and associated acquired and genetic complement abnormalities in the Irish population. The research team comprises Atif Awan, Michael Riordan, Temple Street, in collaboration with Professor Peter Conlon, Beaumont Hospital and Royal College of Surgeons in Ireland.
6. Review of growth in infants with end-stage chronic renal failure. The research team comprises Niamh Dolan and Mary Waldron, Crumlin.

32.6 PROGRAMME METRICS AND EVALUATION

32.6.1 Key Service Outcomes

To ensure that:

- a sufficient skilled and competent multidisciplinary workforce to manage children with renal disease
- children are treated as close to home as possible
- to ensure that children have their healthcare and any social care plans coordinated
- to ensure children with renal disease are treated in line with national guidelines and agreed local pathways
- to ensure equity of access to care for all aspects of renal disease

32.6.2 Key Performance Indicators

- transplant survival data
- growth adequacy
- effective treatment of anaemia in chronic renal insufficiency
- renal biopsy audit
- central line infection
- peritonitis audit
- vitamin D audit
- iron in haemodialysis unit audit
- patient/family experience of the service

The aim of the service is to improve both life expectancy and quality of life for children with renal disease by providing optimal specialised care, and specifically to:

- identify children at risk of renal disease
- prevent progression of disease where possible
- provide appropriate care to those with established renal disease, including support from full multidisciplinary teams, so as to ensure support for the physical, psychosocial and emotional needs of the patient and their family
- prevent complications of renal disease, in order to provide equity of access to care
- provide care as close to home as possible
- ensure smooth and managed transition to adult services
- support parents and families of children with renal disease as well as the affected child
- ensure effective communication between patients, families and service providers

32.7 KEY RECOMMENDATIONS

- Support the provision of adequate staffing across all grades and disciplines within the service.
- Develop service infrastructure to support children and families in our care; and optimise the use of the available resources.
- Secure adequate systems to monitor patients and ensure safe and effective communication.
- Support and develop paediatric urology services.
- Build and open a new children's hospital as quickly as possible.
- Focus on quality by ensuring an evidence-based/research-oriented approach to the care of children with kidney disease.

32.8 ABBREVIATIONS AND ACRONYMS

AKI	acute kidney injury
ANP	advanced nurse practitioner
CME	continuing medical education
CNM	clinical nurse manager
CNS	clinical nurse specialist
HSE	Health Service Executive
HUS	haemolytic uraemic syndrome
ICHMT	Irish Committee on Higher Medical Training
KDOQI	Kidney Disease Outcomes Quality Initiative
MDT	multidisciplinary team
NHS	National Health Service
NICE	National Institute for Health and Clinical Excellence
OLCHC	Our Lady's Children's Hospital Crumlin
PTLD	post-transplant lymphoproliferative disorder
RCPI	Royal College of Physicians of Ireland
UCD	University College Dublin
WTE	whole-time equivalent

32.9 REFERENCES

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