Professor Emeritus Visith Sitprija, MD, Ph.D. Endowed Lectureship: Kidney Diseases Across a Lifespan

Honorable Lecturer: Prapaipim Thirakhupt, MD, RTA Major General (Retired), Clinical Professor

Pediatric Nephrology Division, Phramongkutklao Hospital and College of Medicine

Short Biography

Professor, Major General, Dr. Prapaipim Thirakhupt is a Senior Advisor in the Pediatric Nephrology Division at Phramongkutklao Hospital and Phramongkutklao College of Medicine. She earned her medical degree from Mahidol University, completed her pediatric residency training at Phramongkutklao Hospital, and pursued pediatric nephrology training at Children's Hospital, Free University of Berlin, and the Pediatric Nephrology Department, Heidelberg University Hospital, Germany. She has over 30 years of experience in caring for children with kidney disease. She previously served as the Director of Academic Affairs at Phramongkutklao College of Medicine, Royal Thai Army; President of the Pediatric Nephrology Association of Thailand; and Council member of the Asian Pediatric Nephrology Association. Currently, she serves as a Council member of the Nephrology Society of Thailand, a Council member of the Royal College of Pediatricians of Thailand, and an Assessor for medical schools and pediatric nephrology training accreditation. Her professional interests include both pediatric nephrology and medical education.

Keywords: CKD; chronic kidney disease; renal failure; congenital diseases; kidney; AKI; pediatrics; genetics

Corresponding author: *Prapaipim Thirakhupt*
Email: *prapaipimt@yahoo.com*

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ปาฐกถาเกียรติยศ ศาสตราจารย์เกียรติคุณนายแพทย์ วิศิษฏ์ สิตปรีชา: โรคไตในแต่ละช่วงวัยของชีวิต

องค์ปาฐก: พลตรีหญิง ศาสตราจารย์คลินิก แพทย์หญิง ประไพพิมพ์ ธีรคุปต์

กองกุมารเวชกรรม โรงพยาบาลพระมงกุฎเกล้า วิทยาลัยแพทยศาสตร์พระมงกุฎเกล้า

ประวัติโดยย่อ

พลตรีหญิง ศาสตราจารย์คลินิก พญ. ประไพพิมพ์ ธีรคุปต์ เป็นกุมารแพทย์โรคไตที่ปรึกษาของหน่วยไต กองกุมารเวชกรรม โรงพยาบาลพระมงกุฎเกล้าและวิทยาลัยแพทยศาสตร์พระมงกุฎเกล้า พล.ต.หญิง ประไพพิมพ์ จบการศึกษาแพทยศาสตร์บัณฑิตจากคณะแพทยศาสตร์ศิริราชพยาบาล มหาวิทยาลัยมหิดล จากนั้นได้ฝึกศึกษาเป็นแพทย์ประจำบ้านสาขากุมารเวชศาสตร์ที่โรงพยาบาลพระมงกุฎเกล้า และได้ไปศึกษาต่อด้านกุมารเวชศาสตร์โรคไตที่โรงพยาบาลเด็ก มหาวิทยาลัยฟราย และ โรงพยาบาลเด็ก มหาวิทยาลัยไฮเดลเบิร์ก ประเทศสหพันธ์สาธารณรัฐเยอรมนี พล.ต.หญิง ประไพพิมพ์ มีประสบการณ์การดูแลรักษาผู้ป่วยเด็กโรคไตมากว่า 30 ปี เคยได้รับตำแหน่งผู้อำนวยการกองการศึกษา วิทยาลัยแพทยศาสตร์พระมงกุฎเกล้า ประธานชมรมโรคไตเด็กแห่งประเทศไทย กรรมการบริหารสมาคมโรคไตเด็กเอเชีย ปัจจุบันพล.ต.หญิง ประไพพิมพ์ ดำรงตำแหน่งกรรมการบริหารสมาคมโรคไตแห่งประเทศไทย กรรมการราชวิทยาลัยกุมารแพทย์แห่งประเทศไทย ผู้ตรวจประเมินหลักสูตรแพทยศาสตร์บัณฑิต และหลักสูตรกุมารเวชศาสตร์โรคไต พล.ต.หญิง ประไพพิมพ์ มีความสนใจทั้งทางด้านกุมารเวชศาสตร์โรคไต และแพทยศาสตร์ศึกษา

คำสำคัญ: โรคไตเรื้อรัง; กุมาร; เด็ก; โรคไตเฉียบพลัน; กรรมพันธุ์; พันธุกรรม

Kidney diseases can affect people across their lifespan, from congenital disorders present at birth to conditions that are more common in old age **(Table 1)**. Some conditions are acute and reversible, while others are chronic and progressive, which may lead to end-stage renal disease (ESRD). This article provides an overview of kidney diseases in different age groups, with a focus on two common conditions: congenital anomalies of the kidney and urinary tract (CAKUT) and nephrotic syndrome. The transfer of children with chronic kidney disease from pediatric to adult nephrology care, a crucial process, will also be discussed.

Table 1 Kidney diseases across a life span

Life stage	Common kidney conditions
Neonate	CAKUT, ARPKD, AKI
Childhood	Nephrotic syndrome, HUS, VUR, inherited diseases
Adolescence	Glomerulonephritis, systemic diseases, hereditary nephropathy, IgA nephropathy
Adulthood	CKD, ADPKD, acquired glomerular disease, AKI

ผู้ประพันธ์บรรณกิจ: *ประไพพิมพ์ ธีรคุปต์*
อีเมล: *prapaipimt@yahoo.com*

รับบทความ: 7 ตุลาคม 2568; รับตีพิมพ์: 24 ตุลาคม 2568

1. **Kidney Diseases Present at Birth (Congenital or Neonatal)**

• **Congenital anomalies of the kidney and urinary tract (CAKUT):** The most common cause of pediatric kidney disease in young children, encompassing a range of structural malformations. About half of chronic kidney disease (CKD) patients below 18 years of age are thought to result from CAKUT.[1]

• **Polycystic kidney diseases:** A genetic disorder that can cause cysts to form in the kidneys and liver. Autosomal recessive polycystic kidney disease (ARPKD) often appears in early childhood, leading to severe kidney problems early in life.

• **Acute kidney injury (AKI):** AKI is an important contributing factor to the morbidity and mortality of critically ill neonates. Causes of neonatal AKI are categorized as functional AKI, intrinsic AKI, and postrenal AKI.

2. **Kidney Diseases in Childhood**

• **Nephrotic syndrome:** Characterized by heavy proteinuria, edema, and hypoalbuminemia. Minimal change disease is the most common cause in children, which often responds to corticosteroids.

• **Hemolytic Uremic Syndrome (HUS):** Often caused by *E. coli infection (especially O157:H7)*. Patients usually have hemolytic anemia, thrombocytopenia, and acute kidney injury (AKI).

• **Vesicoureteral Reflux (VUR):** Urine flows backward from the bladder into the ureter/kidneys. This may lead to recurrent urinary tract infections and renal scarring.

• **Inherited diseases:** Certain genetic conditions, such as cystinosis and other inherited metabolic disorders, can manifest in childhood and damage the kidneys over time.

3. **Kidney Diseases in Adolescence**

• **Glomerulonephritis:** This group of diseases continues to be a significant cause of chronic kidney diseases (CKD) in young people. Some types, such as C3 glomerulopathy (C3G), can have different outcomes depending on whether the disease develops in childhood or adulthood.

• **Systemic diseases:** Certain systemic conditions that affect the entire body can also damage the kidneys over the years.

- **Systemic lupus erythematosus (SLE)** is an autoimmune disease that frequently affects young people and can cause severe inflammation of the kidneys, a condition called lupus nephritis.

• **Hereditary Nephropathies**

- Alport syndrome is a genetic disease that causes progressive kidney disease, hearing loss, and eye abnormalities. X-linked inheritance is common.

• **IgA nephropathy:** This disease, which involves the deposition of an antibody (immunoglobulin A) in the glomeruli, is one of the most common causes of glomerulonephritis and can be diagnosed in young adults.

4. **Kidney Diseases in Adulthood**

• **Chronic kidney disease (CKD):** As people age, their risk of developing CKD increases significantly, often driven by common medical conditions.

- Diabetic kidney disease (DKD): The most common cause of CKD in Thailand is diabetes. High blood sugar levels damage the kidneys leading to impaired function.

- Hypertensive nephrosclerosis: Uncontrolled high blood pressure can damage the small blood vessels in the kidneys, leading to impaired function.

• **Polycystic Kidney Disease (ADPKD):** Presents in late adolescence or adulthood. Symptoms include hypertension, flank pain, hematuria, and kidney stones.

• **Acquired Glomerular Diseases:** Membranous nephropathy, Focal segmental glomerulosclerosis can occur later in life.

• **Acute Kidney Injury:** Sudden loss of kidney function may be caused by sepsis, dehydration, nephrotoxic drugs, or obstruction.

Congenital anomalies of the kidney and urinary tract (CAKUT)

Congenital anomalies of the kidney and urinary tract (CAKUT) encompass a broad range of malformations that result from abnormal embryonic kidney and urinary

tract development, including kidney parenchymal malformations, abnormalities in kidney migration, or abnormalities in the developing collecting system. The clinical spectrum ranges from severe malformations, such as kidney agenesis, to potentially milder manifestations, such as vesicoureteral reflux. Almost 50% of cases of chronic kidney disease that manifest within the first three decades of life are caused by CAKUT. A study from 8 university hospitals in Thailand (107 cases) revealed that congenital KUB anomalies (obstructive uropathy and hypo/dysplasia) were the main causes of chronic kidney failure in children (49%) [2]. Evidence suggests that many CAKUT are genetic in origin. To date, mutations in 54 genes have been identified as monogenic causes of CAKUT. CAKUT may also occur in conjunction with other organ defects. Environmental and epigenetic factors can also increase the risk of CAKUT. The discovery of novel CAKUT-causing genes is challenging owing to variable expressivity, incomplete penetrance, and variable genotype-phenotype correlation. This leads to improvements in accurate molecular genetic diagnosis, prognosis assessment, and patient management with CAKUT. Children with CAKUT require individualized, long-term follow-up to monitor renal function, growth, blood pressure, and urinary health. Early identification and management are critical to preventing progression to CKD and optimizing long-term outcomes.

Nephrotic Syndrome

Nephrotic syndrome (NS) is characterized by nephrotic range proteinuria, hypoalbuminemia, and edema. It is caused by increased permeability across the glomerular filtration barrier. In children, minimal change disease (MCD) is the most common histopathology found in primary NS, which often responds well to corticosteroids. However, some children may have frequent relapses, be corticosteroid-dependent, or may not respond to corticosteroids, especially children with other histopathologies such as focal segmental glomerulosclerosis (FSGS). Evaluation of children with primary NS should focus on clinical presentation, corticosteroid responsiveness, and in some cases, kidney histopathology. Genetic testing is recommended in early onset, familial, extrarenal features, or steroid-resistant cases to refine diagnosis and treatment planning [3]. Laboratory panels that test mutations in genes linked to NS or kidney disease are increasingly available. The identification of the most frequent variants (ie, *NPHS2, WT1, NPHS1*) has important clinical implications. The key monogenic causes of pediatric nephrotic syndrome are shown in **Table 2**.

Management of children with NS (new cases) required corticosteroids for a minimum of eight weeks (4 weeks of daily glucocorticoids followed by 4 weeks of alternate-day glucocorticoids) or 12 weeks (6 weeks of daily glucocorticoids followed by 6 weeks of alternate-day glucocorticoids)[4]. Steroid-sparing agents such as mycophenolate mofetil, cyclophosphamide, calcineurin inhibitors, and rituximab should be considered in nephrotic children with frequent relapses, steroid-dependent, or steroid-resistant. In children with hereditary nephrotic syndrome, corticosteroid therapy is generally ineffective; however, disease recurrence is not observed following kidney transplantation.

Transition from pediatric to adult kidney care

As some kidney diseases progress across the life span, many children with CKD now survive into adulthood. Adolescents and young adults are at a higher risk of poor outcomes after transfer. Data from Phramongkutklao Hospital of 35 kidney transplant patients who transferred from pediatric to adult nephrology care revealed that rejection and calcineurin inhibitor intoxication are common despite the same immunosuppressive regimen. The differences between pediatric and adult kidney care are notable, with family-centered care in pediatric care, more intensive support, and a holistic approach. In contrast, adult care is patient-centered, more independent, and has a faster pace, with less psychosocial support. The goals of transition are to ensure continuity of care without gaps, promote self-management skills, address education and vocational planning, and prepare families to shift responsibility from parent to patient. The transition process should be done step by step.

1. Start early (age 12-14): Introduce the concept of transition, assess the patient's knowledge
2. Structured education and skill building (age 14-17): Teach self-management (e.g., medication, appointments)
3. Creating a transition plan (age 17-18): Develop a written medical summary and readiness assessment
4. Transfer to adult nephrology services (age 18-21)
5. Schedule a joint meeting with the adult provider, and arrange overlap of care

Table 2 Key monogenic causes of pediatric nephrotic syndrome

Gene	Protein	Function	Inheritance	Associated syndrome
NPHS1	Nephrin	Podocyte slit diaphragm	AR	Congenital NS (Finnish type)
NPHS2	Podocin	Podocyte integrity	AR	Steroid-resistant NS
WT1	Wilms tumor 1	Podocyte & gonadal development	AD	Denys-Drash, Frasier syndrome
LAMB2	Laminin β2	GBM structure	AR	Pierson syndrome
COQ2, COQ6	Coenzyme Q10 pathway	Mitochondrial energy metabolism	AR	NS plus neuro or muscular features
PLCE1	Phospholipase Cε1	Podocyte signaling	AR	Early SRNS, occasionally responsive to steroids
INF2	Inverted formin 2	Cytoskeleton regulation	AD	FSGS, also with Charcot-Marie-Tooth disease
TRPC6	Cation channel	Calcium signaling in podocytes	AD	Late-onset FSGS
SMARCAL1	DNA helicase	DNA repair	AR	Schimke immuno-osseous dysplasia
CD2AP	CD2-associated protein	Podocyte slit diaphragm	AR/AD	FSGS, variable expression
ACTN4	α actinin 4	Active binding protein	AD	Adult-onset FSGS

Summary

Kidney diseases can occur from birth to adulthood across the lifespan. Some diseases are acute and reversible, while others are chronic and aggressive, which lead to ESRD. Many diseases in children are inherited and require genetic testing. When children with CKD get older, a carefully planned transition process to adult kidney care should be done to achieve the proper outcome for the patients.

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