

Electrolyte Management After Kidney Transplant in Cystinosis: A Patient-Centered Approach to Adolescent Nutrition Care



Cystinosis Key Facts:

- ❑ Rare lysosomal storage disorder (~1–2 per million)
 - ❑ CTNS mutation → cystine accumulation and multi-organ damage
 - ❑ Early disease presents as Fanconi syndrome
 - ❑ Fanconi syndrome causes urinary losses of:
 - potassium
 - phosphate
 - bicarbonate
 - amino acids
 - glucose
- Short term:** dehydration, metabolic acidosis, poor growth
- Long-term:**
- ESRD
 - Kidney transplant
 - Transplant ≠ cure

Case Snapshot:

15-year-old female with nephropathic cystinosis
ESRD → living donor kidney transplant (March 2026)

This case highlights **how nutrition management in cystinosis evolves after transplant** and why **sustainable, patient-centered interventions** may be more effective than overly restrictive plans.

History of:

- Fanconi syndrome
- Electrolyte abnormalities
- Poor growth
- GI complications

Post-transplant improvements:

- Improved renal function
- Improved appetite
- Improved growth
- Improved laboratory values

Growth Improvement:

Weight 10th → 28th percentile
Height percentile: 10th → 24th percentile

Despite improved kidney function:

- cystinosis is a lifelong condition
- cysteamine continued
- immunosuppressive medications introduced new risks
- dietary priorities completely changed

Post-Transplant Medication–Nutrition Considerations:

- **Tacrolimus:** avoid grapefruit; food safety
- **Prednisone:** increased appetite, and bone health
- **Mycophenolate:** GI side effects
- **Cysteamine:** body odor
- **Phosphate binders:** liberalized diet

Nutritional flip post-transplant:

Kidney transplantation rapidly changed this patient’s nutrition priorities. Management shifted from replacing ongoing electrolyte losses and supporting growth to protecting the transplanted kidney, managing medication side effects, and creating a sustainable long-term diet.

*The transition from a liberalized, replacement-focused diet to one centered on **monitoring, restriction, and long-term adherence** created new behavioral and psychosocial challenges for the patient.*

Before Transplant	After Transplant
Electrolyte wasting	Electrolyte retention/monitoring
Replace potassium + phosphate	Monitor/manage phosphorus
Liberalized sodium intake	Sodium restriction
CKD fatigue + poor appetite	Prednisone appetite/weight gain risk
Growth failure	Catch-up growth possible
Dialysis/CKD management	Immunosuppression management
Fanconi syndrome symptoms	Medication side effects symptoms
Focus: survival + replacement	Focus: long-term graft protection
Nutrition support to maintain intake	Sustainable adherence + quality of life

Beyond Lab Values:

Challenges:

- new food rules
- sodium restriction
- fear of “doing it wrong”
- losing familiar foods and routines

Successful nutrition care requires balancing clinical goals with patient preferences, independence, and sustainable eating behaviors.

Team Goal: Create a sustainable plan for long-term adherence

Key Strategies

- Use phosphate binders instead of added dietary restriction
- Preserve diet normalcy as much as possible
- Encourage patient autonomy and shared decision-making

A manageable plan promotes long-term adherence.

Clinical Takeaways:

- Nutritional priorities shift rapidly post-transplant
- Adolescents need ownership for long-term adherence
- Sustainable plans may be more effective than highly restrictive ones

*As patients with cystinosis live longer after transplantation, nutrition care is increasingly focused not only on lab correction, but also on **sustainability, independence, and long-term quality of life.***

