

Innovations in Nephrology

Rady Children's

Hospital
San Diego

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focused solely on children.*



PEOPLE

Dr. Nourbakhsh: Easing the transition to adult care



Dean Nourbakhsh, D.O., a member of the Division of Nephrology at Rady Children's Hospital-San Diego and an assistant clinical professor at UC San Diego School of Medicine, has a special interest in treating pediatric patients with a range of kidney conditions who are transitioning to an adult nephrology clinic. He cares for patients in the Transition Clinic at UC San Diego Health. The clinic coordinates care for patients in their postteen years who are making the move to UC San Diego Health's adult nephrology clinic.

The transition of pediatric patients to adult nephrology care can be challenging, especially for those with special needs and chronic illness. Patients and their families face uncertainty and apprehension about the loss of a longtime provider, the absence of familiar faces, a new healthcare system to navigate, a new location for care, access to medications and insurance coverage. Some may also have negative perceptions of adult healthcare.

The Transition Clinic's goal is to ease anxiety and address concerns – for both patients and providers. At the patient's initial visit, Dr. Nourbakhsh begins the planning process for the transition. As part of this process, pediatric clinic staff communicates with the adult nephrology clinic staff on a weekly basis to evaluate and overcome the insurance barriers for transition. At subsequent clinic visits, other obstacles are addressed and the framework for the transition is established.

Along with his clinical work, Dr. Nourbakhsh teaches residents at Rady Children's. Among his numerous honors and awards over the years has been the Whitehill Prize Award for Excellence in Teaching.

His research focuses on sepsis-induced acute kidney injury (AKI), one of the most common causes of AKI. Other research interests include metabolic alkalosis, protein-energy wasting in children with chronic kidney disease and the role of renal oxygenation and mitochondrial function in the pathophysiology of AKI. Dr. Nourbakhsh's work has been published in various peer-reviewed medical journals, including *Nephron Clinical Practice*, *Pediatric Nephrology* and *The American Journal of Medicine*.



PROGRAMS

TSC Clinic: Expertise in therapy for kidney-associated issues

The [Tuberous Sclerosis Complex \(TSC\)](#) Clinic at Rady Children's, a designated TSC Center by the Tuberous Sclerosis Alliance, cares for children through adulthood. The multidisciplinary team includes pediatric nephrologist [Peter Yorgin, M.D.](#), a pediatric neurologist, a pediatric pulmonologist, a pediatric cardiologist, a pediatric neuro-oncologist and a nurse manager. Specialists in pediatric ophthalmology, psychology, dermatology and neurosurgery are also involved.

Dr. Yorgin treats and monitors patients who have renal cystic disease or angiomyolipomas, which can become large enough to cause severe and life-threatening bleeding. To treat these tumors, he has been using two drugs primarily used in kidney transplant patients: the mTOR inhibitors everolimus and sirolimus. Both have been effective in reducing tumor size, and there is early evidence that they also shrink kidney cysts. Additionally, the medications have been successful in treating SEGA (subependymal giant cell astrocytoma), a noncancerous brain tumor associated with TSC. The team members are experts in using these therapies, Dr. Yorgin says, and patients as young as 5 have received the drugs.



The clinic is also involved in research on everolimus. It has been one of the sites for a multicenter everolimus treatment trial of intractable epilepsy in TSC and continues to participate in the long-term follow-up phase of the study. The clinic is also exploring basic science opportunities related to the m-TOR pathway and collaborates with other TSC centers around the country.



RESEARCH

Associations between weight loss, kidney function decline and risk of end-stage renal disease in children

[Robert Mak, M.D., Ph.D.](#), chief of Rady Children's Division of Nephrology and a professor of pediatrics at UC San Diego School of Medicine, has found associations among weight loss, kidney function decline and risk of end-stage kidney disease in children. The findings are part of the Chronic Kidney Disease in Children (CKiD) study, a National Institutes of Health-funded prospective cohort study of children with CKD and adolescents ages 1 to 16 with estimated glomerular filtration rates (eGFRs) of 30 to 90 mL/min/1.73 m² that began in 2005 and is ongoing.

Anorexia and malnutrition are associated with poor outcomes in



innovation
belongs in every moment



children with chronic kidney disease (CKD). Whether weight slowly declines in a linear fashion with advancing CKD or remains stable until the late stages of CKD is unclear. In the study, during a mean longitudinal follow-up of 3.4 years, BMI z scores remained stable until eGFR decreased to <35 mL/min/1.73 m². When eGFR decreased to <35 mL/min/1.73 m², a mean decline in BMI z score of 0.13 (95% CI, 0.09-0.17) was noted with each 10mL/min/1.73 m² further decline in eGFR. This was statistically significantly different from the weight trajectory when eGFR was ≥35 mL/min/1.73 m² (P < 0.001).

Among children and adolescents with significant weight loss (defined as decline in BMI z score > 0.2 per year) after eGFR decreased to <35 mL/min/1.73 m², the odds of end-stage renal disease (ESRD) was 3.28 (95% CI, 1.53-7.05) times greater compared with participants with stable BMI z scores (BMI z score change per year of 0-0.1). Dr. Mak's data suggests that more frequent monitoring of protein-energy wasting may be especially important among children and adolescents with glomerular causes of CKD, in whom weight loss was observed to be more profound compared with those with nonglomerular causes of CKD. He and his colleagues speculate that greater attention to nutrition (by [naso]gastric feeding or caloric supplementation) due to the chronicity of kidney disease and the younger age of onset of CKD may occur in this subset of patients.

In contrast, due to the older age of most children and adolescents who acquire CKD due to glomerulonephritis, it is possible that less attention is paid to nutrition among this subgroup, leading to more profound weight loss with advancing CKD. It is also possible that children and adolescents with glomerulonephritis have more generalized inflammation and therefore increased protein-energy wasting and weight loss. Further research is needed to determine reasons behind the association between weight loss and risk for ESRD in children and adolescents. [View the abstract in the American Journal of Kidney Diseases.](#)

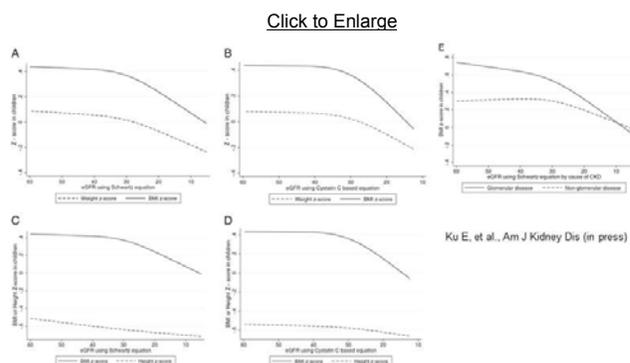
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Association between longitudinal repeated measurements of body mass index (BMI), weight, and height z scores with repeated measurements of kidney function over time in the Chronic Kidney Disease (CKD) in Children Study in the overall cohort and by cause of CKD (glomerular vs nonglomerular disease).

Repeated measures of

(A) BMI and weight z scores with repeated measures of estimated glomerular filtration rate (eGFR) by serum creatinine;

(B) BMI and weight z scores with repeated measures of eGFR by cystatin C;

(C) BMI and height z scores with repeated measures of eGFR by serum creatinine;

(D) BMI and height z scores with repeated measures of eGFR by cystatin C; and

(E) BMI with repeated measures of eGFR by serum creatinine by glomerular versus nonglomerular causes of CK.

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