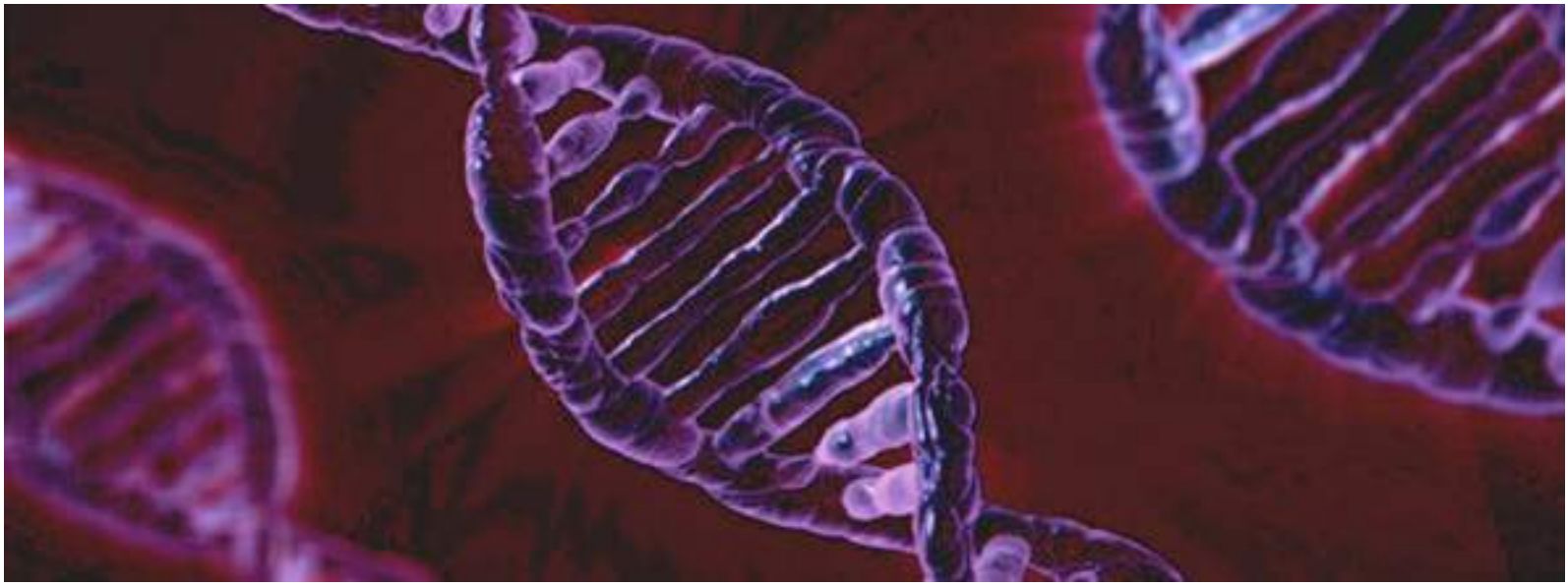
NephNews

Kidney Disease Research

SPRING 2016 VOLUME 1 ISSUE 2



Welcome to the Spring issue of NephNews: dedicated to keeping you informed about kidney disease.

In the Division of Nephrology at The Hospital for Sick Children, our mission is to educate, inform, and inspire our families about kidney-related topics and issues.

As always, delivering the highest quality care to patients is our number one priority. This edition of NephNews puts the spotlight on an important genetic disorder--tuberous sclerosis--and highlights the changes being made across disciplines at SickKids to better care for children with this rare disease.

Our hope is that by shedding light on some of the research we have been doing, and how our care and treatment of patients has been transformed as a result, we will motivate our patients and families to participate in our research projects. With more children involved, we can do better research, and ultimately help more children with kidney disease receive the best possible care.

We encourage you to share your thoughts and feedback about this newsletter with us, so that we can improve our content based on what is important to you.

Dr. Lisa Robinson, Division Chief, Nephrology

In this issue:

Spotlight on tuberous sclerosis complex (TSC)

Quick Facts:

- Tuberous sclerosis complex (TSC) is a rare, hereditary disease that affects multiple systems in the body. The kidneys, heart, eyes, lungs, brain, and skin can all be affected by TSC.
- TSC causes tumors to grow in these parts of the body. These tumors are not cancerous, which means they do not spread to neighbouring parts of the body.
- Children with TSC can experience seizures, developmental delays, behavioral problems, abnormalities of the skin, and kidney disease.
- 1 in 1-2 million people worldwide are affected by TSC.
- TSC affects 1 in 6,000 newborn babies in North America--but many cases go undiagnosed in infants. This is because symptoms are often mild at the outset of this disease.

While TSC is a rare disorder, it can strongly affect children's growth and development, and can lead to severe complications of the kidney. At SickKids, approximately 100 children are currently being followed for this disease, and 3 out of every 4 of these children have associated kidney problems.

The most common kidney problem encountered in these patients is ***renal angiomyolipomas***. These are non-cancerous, non-spreading tumours made up of abnormal blood vessels, muscle cells, and fat. Typically, the older these children are, the more tumours they have in their kidneys.

These tumours can cause pain and bleeding, and in severe cases they can lead to kidney failure. Because of this risk, patients with TSC who have kidney involvement are followed closely, to help prevent permanent kidney damage.

There is no cure for TSC. Treatment involves monitoring the child for developmental and kidney-related complications.

Assessing treatments for TSC kidney disease

A new research study has recently been started at SickKids, which aims to assess the burden of kidney disease in children currently followed for TSC, and how this burden changes when children are treated with a new medication.

Our bodies are made up of trillions of ***cells***, which are the building blocks for our different body systems. Inside each cell are thousands of ***genes***. These genes carry the information that determines each person's ***traits***: the features or characteristics about a person that are passed on from one's parents.

Each gene has a special job to do. Two genes, called TSC1 and TSC2, are important for turning off cell growth. When a child has TSC, there are problems in these genes (called ***mutations***). The genes no longer work properly and are unable to stop cells from growing. This means that cells enlarge abnormally, which causes tumours to develop.

A new class of medications called ***mTOR inhibitors*** work by helping to gain back control of cell growth. Researchers at SickKids are currently working to find out how well these drugs are working in children with kidney problems due to TSC. The hope is that by doing this research, treatment and management of children with TSC can be improved.

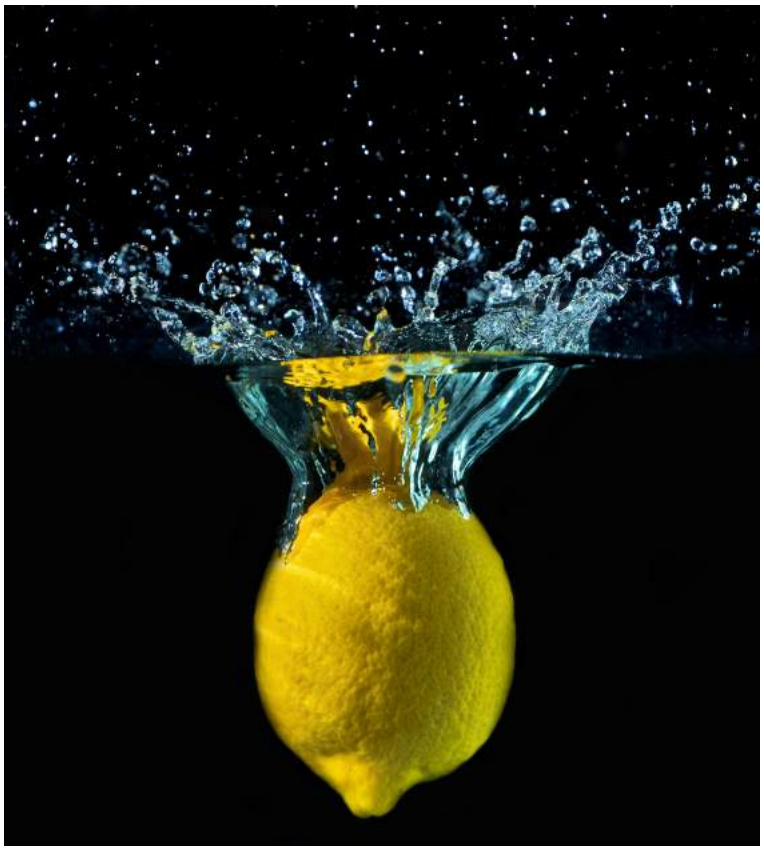
SickKids opens new comprehensive TSC clinic

TSC is a complex disorder that involves multiple organ systems. Because of this, children with a diagnosis of TSC are typically seen by specialists in many different areas of medicine. Brain lesions are treated by neurologists, skin lesions are treated by dermatologists, kidney problems are treated by nephrologists, etc. In the past, this often meant that children were required to come in to the hospital for many separate appointments with each of these specialties, in order to receive appropriate care for their disease.

Because of the demand for extensive care from multiple disciplines, the Hospital for Sick Children has now instituted a new, designated Comprehensive TSC Clinic for patients with this complex diagnosis. This clinic represents a coordinated effort from many fields, including: nephrology, cardiology, oncology, urology, dermatology, genetics, neurosurgery, gastroenterology, psychiatry, social work, and psychology. As of January 2016, physicians from each of these specialities are working together to provide more comprehensive care for children living with TSC.

The TSC Clinic at SickKids represents the third paediatric clinic of its kind in Canada, and is an important step forward in the care and management of children with TSC.

What can you do for your kidneys?



There are several easy ways to reduce your child's risk of developing kidney disease, and to promote overall kidney health:

1. Maintain an active lifestyle
   Keeping fit reduces your blood pressure and risk for kidney disease.

2. Reduce sugar and salt intake
   Reducing your risk of high blood pressure can help your kidneys.

3. Monitor blood pressure and keep weight in check
   High blood pressure is the leading cause of kidney damage. Keep your heart and kidneys healthy.

4. Stay hydrated
   Drink plenty of water per day (add lemon) to help the kidneys clear toxins from the body.

5. Do not smoke
   Smoking slows the flow of blood to the kidneys, impairing their ability to work properly.

Kidney research at SickKids

In the Division of Nephrology, we are committed to giving our patients the highest quality care. This means that we need to continue to expand our understanding of kidney diseases, their causes and treatments, and their implications on overall health and well-being. To do this, we conduct research studies.

Our division has several ongoing studies:

Patient Registries (TRACKD study)
Our registries are some of our biggest resources when doing research. They help us identify which children can help us to answer our research questions in the best way possible. Our goal is to enrol every child into a comprehensive registry called TRACKD, to keep track of each one of our patients and know which children will be eligible for future research studies.

Genetic Studies
Several of our research studies aim to explore the genetic causes of kidney diseases affecting children. By understanding the mechanisms by which these diseases cause kidney damage, we will be better able to target our treatment and management. Currently, several projects are underway to understand the genetics of rare diseases, including atypical hemolytic-uremic syndrome, undiagnosed congenital tubulopathies, and more.

Dialysis Outcomes
Infants with kidney failure who are less than 2 years of age are generally too small to receive adult donor kidneys, and thus may require dialysis for a period of time before being transplanted. We are currently undertaking a study to understand the outcomes of chronic dialysis in these infants.

Transplant Outcomes (PROTECT study)
Several of our patients with kidney failure ultimately go on to receive a kidney transplant. The goals of one of our projects are to study the risk of cardiovascular and kidney conditions in children who have received a transplant, in order to determine treatment and lifestyle modifications that can improve their overall outcomes.

Get involved:
participate in our research studies

If you have been diagnosed with:
Tuberous Sclerosis Complex,
Atypical Hemolytic Uremic Syndrome,
IgA Nephropathy,
Glomerulonephropathy,
Rare Kidney Stone Disease,
Nephrotic Syndrome,
or **Renal Dysplasia**

or if you are currently undergoing **dialysis**

you may be eligible for one of our research studies.

Please contact *nephrology.research@sickkids.ca*
if you are interested in learning more about participating in research.

Upcoming learning opportunities

INSIGHT Family Appreciation Night

An evening to celebrate the patients and families participating in the INSIGHT project.

Date and time to be determined.

Please contact: insight.study@sickkids.ca for more information.

Peter Gilgan Centre for Research & Learning
686 Bay Street, Toronto

Chronic Kidney Disease Family Education Event

An afternoon information session to inform patients and families about chronic kidney disease (CKD) in children.

Saturday, June 4, 2016
9am - 2pm

8th Floor, Burton Wing
The Hospital for Sick Children
555 University Avenue, Toronto

World Kidney Day

World Kidney Day (WKD) took place on March 10, 2016. The Hospital for Sick Children participated in this global awareness event by hosting the first ever SickKids World Kidney Day symposium: a day of educational and informative kidney-related games, displays, and interactive demonstrations.

Patients, families, and staff alike were able to join in the day's celebrations by learning about kidney function with seive models of filtration and a fun-facts jeopardy game. Participants were also able to learn about what happens when kidney's are not functioning at their best, with interactive displays on kidney dysfunction, kidney stone removal devices, dialysis machines, and more. Research into improving the care of children with kidney disease was also highlighted.

We would like to thank everyone who stopped by to participate in this inaugural event.

Dr. Lisa Robinson signs the WKD kidney-shaped canvas.

Karlota Borges and Tonny Banh, Research Assistants, help with the educational displays and demonstrations.

Left to right: Drs. Valerie Langlois, Lisa Robinson, and Mina Matsuda-Abedini show their support for WKD.

Organ Donation Awareness Day

On April 21, 2016, the Transplant & Regenerative Medicine Centre will be hosting an organ and tissue donation awareness event.

Join us on the ramp outside the cafeteria on the main floor from 10am-3pm, and WEAR GREEN to show support!

Kidney-friendly recipes

French Toast
(low phosphate, low potassium)

Recipe from Andrea Aquilina, Dietician, The Hospital for Sick Children

Ingredients:
4 large eggs
1/2 cup rice milk (not enriched; e.g., Rice Dream Classic)
4 tsp sugar
2 tsp vanilla extract
2 tsp cinnamon
8 slices white bread (slightly stale is best)
Butter or margarine

Directions:
Beat eggs, rice milk, sugar, cinnamon, and vanilla together with a fork. Melt butter or margarine in a frying pan and heat to medium. Dip bread in egg mixture and flip to coat both sides. Fry bread, flipping once, until golden on both sides. Serve with butter and syrup.

Chicken Tikka
(low sodium, low potassium, low phosphate)

Recipe from Andrea Aquilina, Dietician, The Hospital for Sick Children

Ingredients:
600 g (4 breasts) boneless chicken breasts
1 tsp ground ginger
1 clove garlic, minced
1 tsp ground cumin
1/4 tsp chili powder
1/4 cup plain yogurt
2 tsp canola oil
Skewers

Directions:
Cut chicken into 2 cm (3/4 inch) cubes. Stir together remaining ingredients to make a marinade. Stir in chicken to coat and marinate for at least 2 hours in the fridge. Preheat oven to 400F or start barbecue. Thread chicken onto skewers. Bake for 6 minutes or barbecue on both sides until firm to touch and no sign of pink.

Rice Pudding
(low sodium, low potassium, low phosphate)

Recipe from Andrea Aquilina, Dietician, The Hospital for Sick Children

Ingredients:
2 1/2 cups vanilla almond milk (not enriched, e.g., Silk True Almond Vanilla)
1/3 cup short grain white rice or Arborio rice
Pinch of salt
1 egg
1 tsp vanilla
1/2 tsp cinnamon
2-3 tbsp sugar or brown sugar

Directions:
In a medium saucepan, bring almond milk, rice, and salt to boil over medium-high heat. Reduce heat to low and simmer, uncovered until rice is cooked (25-30 min). In a small bowl, beat egg. Whisk a ladle of hot rice mixture into beaten egg. Add egg mixture back to saucepan and stir over low heat until thickened (10-12 min). Remove from heat and stir in vanilla, cinnamon, and sugar. Serve warm or cold.

We want your drawings!

Bring your artwork in at your next visit, and give it to one of the staff in clinic.
We will showcase your art in the next issue of our newsletter.
We can't wait to see your masterpieces!

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CHRONIC KIDNEY DISEASE

SickKids®

Saturday, June 4, 2016

9am - 2pm

8th Floor, Burton Wing
The Hospital for Sick Children
555 University Ave.
Toronto

Chronic Kidney Disease (CKD) occurs when the kidneys are not working properly.

Agenda:

- Overview of CKD
- Diet
- Medications
- Dialysis
- Transplant
- Coping with CKD

Family Education Event

Food, child care, and parking will be provided.
Space is limited. Please RSVP by May 15th:
nephrology.events@sickkids.ca or 416-813-8326