

## EXCITING NEWS

PKD FOUNDATION HONORS THREE NEPHROLOGISTS DEDICATED TO TREATING AND CURING CHILDREN WITH POLYCYSTIC KIDNEY DISEASE (PKD)

### • Award amount increases despite troubling economy

(Kansas City, Mo) Friday, March 6, 2009 – The PKD Foundation, the world's leader in catalyzing research and new therapies for those suffering from PKD, is once again teaming up with the International Society of Nephrology to honor three world renowned nephrologists with the 2009 Lillian Jean Kaplan International Prize for Advancement in the Understanding of Polycystic Kidney Disease.

Dr. Corinne Antignac (Necker Hospital, Paris, France)

Dr. Lisa Guay-Woodford (University of Alabama at Birmingham, USA)

Dr. Friedhelm Hildebrandt (University of Michigan, USA)

Each will receive the Kaplan International PKD Prize of \$50,000 cash – at the World Congress of Nephrology May 25, 2009 in Milan, Italy. The Kaplan PKD Prize is the largest for a medical sub-specialty.

For the first time –

All those selected for this prestigious prize are pediatric nephrologists specializing in finding a treatment and cure for polycystic kidney diseases (PKD) in children and young adults, and despite economic constraints affecting philanthropy worldwide, Thomas Kaplan (the benefactor behind this prize) increased his giving to a total of \$150,000 in order to honor three doctors instead of two as before.



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1-800-PKD-CURE  
[www.pkdcure.org](http://www.pkdcure.org)

Learn more about  
the PKD Foundation at  
[www.pkdcure.org](http://www.pkdcure.org)

## PKD Foundation

# ARPKD CHAPTER NEWSLETTER

SPRING 2009



## This is YOUR Chapter!

As we're getting ready for summer, we're happy to meet again in Chicago at the PKD Convention. The schedule is packed yet allows for a full travel day Friday if needed. Even lunch on Saturday is a session – nutrition for ARPKD attendees. We'll also be hosting the annual pizza party for families of ARPKD/ADPKD children, as well as speakers on Saturday. We hope to see you there.

Even with the economic challenges, the PKD Foundation is stable. In order to show their dedication to the research community the Foundation made sure to award grants during the January 2009 Board of Trustees meeting. In doing so, changes were made with the Foundation, which included canceling the 2nd Convention of the year as well as adjusting budgets from each department. It's comforting to know that the leaders of the PKD Foundation are as concerned about the economy as all of us are; and are making necessary adjustments in order to keep the research moving.

You will note that in this newsletter a new section is started to honor the memory of the families that have lost a baby to ARPKD. If you would like your baby's name included, please do let us know. This will be a regular feature and the names will be listed in each newsletter. The PKD Foundation typically forwards contacts to the two of us and we reach out to many other parents for support to newly diagnosed families. We thank you for your willingness to share your stories and offer support.

As always, if there is anything we can do for you, please let us know.

Julia Roberts (404-636-9382) & Michele Karl (914-522-6193)

Co-coordinators

[arpkdchapter@pkdcure.org](mailto:arpkdchapter@pkdcure.org)

Michele is still handling the newsletter, so if you have any inclusions or questions for Q&A, please let her know: [M3karl@optonline.net](mailto:M3karl@optonline.net).

## Links

**National organization committed to promoting reproductive wellness:**

[www.americanpregnancy.org](http://www.americanpregnancy.org) - increasing awareness of pregnancy needs through education, research, advocacy, and community awareness

**PKD online community:** [www.hopesquare.org](http://www.hopesquare.org) - The online PKD Community and social networking site that is up and running offering blogs, forums, live chat, video, personal profiles, connecting with friends, and more. There is an ARPKD/ADPKD in children group.

### CHAPTER INFORMATION

Chapter Coordinators:

Julia Roberts  
&  
Michele Karl

[arpkdchapter@pkdcure.org](mailto:arpkdchapter@pkdcure.org)

[www.pkdcure.org/arpkdchapter](http://www.pkdcure.org/arpkdchapter)

**Blog by ARPKDmom:** [www.caringbridge.org/visit/bennettmatthew](http://www.caringbridge.org/visit/bennettmatthew) telling the story of a boy who had one failed kidney transplant and is in desperate need for a second transplant.

## 2009 National Convention

This year's National Convention on PKD will take place in Chicago the weekend of June 26-28. The conference will take place at the DoubleTree Hotel Chicago – Oak Brook •1909 Spring Road • Oak Brook, IL 60523;• Phone: 630-472-6000

Here is the schedule for the ARPKD tracks:

- **ARPKD Parent to Parent Roundtable Discussion / Introductions;**  
Speakers: Julia Roberts & Michele Karl, ARPKD Chapter Co-Coordinators
- **The Science of ARPKD;**  
Speaker: Lisa M. Guay-Woodford, MD, University of Alabama at Birmingham
- **Top 10 Questions to Ask Your Child's Doctor;**  
Speaker: Lisa M. Guay-Woodford, MD, University of Alabama at Birmingham
- **Nutrition for ARPKD Children;**  
Speaker: Nancy Spinozzi, RD, LDN, Children's Hospital Boston, Clinical Nutrition Services, Division of Nephrology
- **Dialysis & Transplantation for Children;**  
Speaker: Scott Williams, MD, University of Texas Southwestern Medical Center
- **Congenital Hepatic Fibrosis;**  
Speaker: Theo T. Heller, MD, University of Maryland Medical School/National Institute of Diabetes and Digestive and Kidney Diseases (NIDDK), Liver Diseases Branch
- **Emotional Impact Chronic Illness Has on Children;**  
Speaker: Teri Sullivan Lutz  
She will be sharing information based on the book, *Trauma Through a Child's Eyes* by Peter A. Levine and Maggie Kline  
"A truly remarkable book that captures the essence of what it is to be a traumatized child, while simultaneously helping us understand, appreciate, and facilitate their natural capacity to heal. Written with a deep sense of compassion and wisdom, this book offers clear insight to those who care for and about children."  
—John Stewart, PhD
- **Questions & Answers on ARPKD;**  
Speaker: Panel of Experts

*NOTE: There will also be a hospitality suite for ARPKD families on Friday from 4:00pm – 7:00pm and an ARPKD Family Party on Saturday evening from 5:00pm – 6:30pm. Families of ADPKD children are also welcome at any and all ARPKD listed sessions/events.*

### Is attending the PKD Convention a financial hardship?

Scholarships for a caregiver in an ARPKD family are being offered for families that find it difficult to attend the PKD Convention because of the financial hardships that can and often do hit with a diagnosis of ARPKD and the on-going treatment of a chronic disease.

A special anonymous donation, designated for caregiver scholarships for National Convention attendance has been established. Depending on need, the National Convention registration, travel and hotel may be covered.

Please contact [taraf@pkdfoundation.org](mailto:taraf@pkdfoundation.org) or 800-PKD-CURE for a simple application!  
We hope to see you there.

## FEATURE ARPKD FAMILY: THE NEARY FAMILY



Helen's Story:

My husband and I were so excited to find out we were expecting our first child. Until the 7th month, the pregnancy was very normal. At 28 weeks, we went to have a Level 2 ultrasound and found out the devastating news. Our first child, a girl, most likely had ARPKD. The doctors grimly told us that our baby's kidneys were very large and looked "bright" which meant a high possibility that she had ARPKD and it looked severe. They gave us a poor prognosis and sent us home with very little information or resources. Along with our family, we began to research and found other families who had children who were doing well with ARPKD. Life wasn't easy but they had beat the odds and we found hope.

We found doctors who were willing to fight with us. They were very realistic about our daughter's specific case but said they were there to support us and our daughter. We went for weekly appointments and had ultrasounds and non stress tests done to monitor our baby's growth especially watching the size of her kidneys. I had little amniotic fluid and they also were watching her lung development.

At 38 weeks, I went into labor naturally and delivered our daughter, Helen Grace. She was 6 lbs and 11 ounces and gave out a loud scream when she was born. We were so relieved to hear this sound because we weren't sure how developed her lungs were. Quickly, Helen began showing signs of distress and was taken to the NICU and put on a ventilator. Helen's lungs were severely underdeveloped and she needed the ventilator to live. We went to the NICU and spent as much time with her as we could. The doctors were very honest with us about Helen's underdeveloped lungs and the severity of her kidney disease but we still had hope. Many friends and family were able to come to the hospital and meet Helen.

On the second night, Helen developed a pneumothorax and it was very clear that she would not be able to recover. The doctors took her off the ventilator and we held her for the first time and rocked her as she passed away. Helen's short life was full of love and she taught us all so many lessons in that time. It was devastating to lose Helen but we know that we are better for having her in our life even for such a short time. The process of grief was difficult but we knew that to honor Helen and celebrate her life we needed to live our life to the fullest.

In remembrance of Helen, we have an annual picnic at our house to raise money for the PKD Foundation's Walk for PKD. Over 200 people come to our house to celebrate and donate to this special cause. In seven years, we have raised close to \$80,000 in Helen's memory. We are hopeful that someday a cure will be found. We were also blessed to have two daughters who are healthy. Josie is five and Tess will be two. They are blessings in our life and we feel so fortunate to have them.

-Written by Kristen Neary ([marksneary@comcast.net](mailto:marksneary@comcast.net))



Josie and Tess Neary



### Walk for PKD

Even though the economy had started to turn, the 2008 Walk for PKD was still a success raising \$2.9 million. For the 2008 Walk, the PKD Foundation set up a new feature during registration to have your team listed as a "Walking for ARPKD" team. This was very exciting because anyone who looked at the site could see which teams were ARPKD teams.

Sixty-six teams registered as "Walking for ARPKD" and raised more than \$202,866.

# REMEMBRANCE

*We decided to add a section to the newsletter to remember those babies who have lost their lives to ARPKD. If you would like your child's name mentioned here in future newsletters please email us to let us know.*

## Remembrance

Remembrance is a golden chain

Death tries to break,

but all in vain.

To have, to love, and then to part

Is the greatest sorrow of one's heart.

The years may wipe out many things

But some they wipe out never.

Like memories of those happy times

When we were all together.

~ Author Unknown

- Helen Grace Marks May 24-May 26, 2002
- Noah Samuel DeRycke September 16, 2003
- Juniper Isabelle Wakely December 10, 2008
- Bowen Lee Urban March 26-March 29, 2009
- Jody Michael Terian April 10th 2009
- Van Fredrick Weisshaar May 7- May 8, 2009
- Noah Raynham Lyons December 29, 2004
- Carley Devin Grove October 22-October 23, 2002

## Q&A with DR. ASKENAZI & DR. HELLER

*The answers provided below are not necessarily the views of the PKD Foundation, it's staff or constituency. Always consult a physician who you trust and is familiar with these issues. If you have a question you would like answered here please email it to Michele at m3karl@optonline.net.*

### I. How can you preemptively plan for a transplant and avoid dialysis?

Planning is key... Most institutions start evaluation process when the GFR is about 20 ml/kg/1.73 m<sup>2</sup> which is determined by serum creatinine and patient size. Depending on the situation and institution, kidneys may need to be removed prior to transplantation which would commit the patient to dialysis therapy for a short time.

### II. What are some important questions parents should ask at their first appointment with a pediatric nephrologist after the initial diagnosis of ARPKD?

What is the current renal function? How do we decrease the progression of CKD? Are there consequences (anemia, renal osteodystrophy, electrolyte abnormalities, hypertension, acidosis, growth failure) at this time and if so, what do we do to control these? How is the liver doing? When should we follow up with you (the nephrologist)? How often should we monitor the blood pressure? Are there any restrictions at this point? What are things we should look out for and/or be concerned enough about to call the pediatrician?

### III. What is your opinion on doing a liver biopsy in a child with ARPKD/CHF?

Liver biopsy in CHF is controversial; in general we do not recommend a liver biopsy to make the diagnosis of CHF. If it is completely unclear what disease a person has then it would make sense to do a liver biopsy. We would also do a liver biopsy if there was a mass present to diagnose the mass. We do not think a liver biopsy is necessary to stage or follow CHF. There is no validated system to do stage CHF by biopsy and we can assess severity using noninvasive means.

# RESEARCH GRANTS

The following 2008 ARPKD:

- Yale University School of Medicine: pathogenesis of congenital hepatic fibrosis and biliary cyst formation in ARPKD, \$75,000
- Brigham and Women's Hospital: Molecular Mechanisms of the Autosomal Recessive Polycystic Kidney Disease; \$65,000
- Aachen University of Technology: Characterization of the PKHD1 gene and genotype-phenotype studies in autosomal recessive polycystic kidney disease; \$75,000
- Midical College of Wisconsin: The role of 20-HETE in polycystic kidney disease ; \$75,000
- Identification of PKD markers: Transcriptome-profiling based approach; \$75,000
- Yale University School of Medicine: Mechanism of polycystic liver disease in Hhex mutant mice; \$75,000
- Swiss Federal Institute of Technology: The role of BICC in PKD and cilia signaling; \$75,000
- Hokkaido University School of Medicine: Molecular genetic basis of kidney and liver cyst formation in ADPLD; \$50,000

## KIDNEY WISE NUTRITION

**Brilliant Eats** by Kelly L Welsh, R.D., C.D. is an easy to follow cookbook that was written by a renal dietitian, who is also a kidney disease patient. There are nutritious and simple meals that the entire family can enjoy. To purchase Brilliant Eats you can go to [www.pkdcure.org](http://www.pkdcure.org) or to [www.kidneywise.org](http://www.kidneywise.org).

Here is a sample recipe from Brilliant Eats: Spicy Chicken

Prep time: 1 hour

Servings: 4

Ingredients:

2 tablespoons balsamic vinegar

1 tablespoon olive oil

½ teaspoon dried rosemary

1/4 teaspoon ground cumin

1/8 teaspoon ground coriander

1/8 teaspoon black pepper

dash of ground red pepper

8 small green onions, cut into 2 inch pieces

2 cups hot cooked rice

2 tablespoons peach or apricot preserves, melted (optional)

Preparation:

In a small bowl, combine vinegar, olive oil, rosemary, cumin, coriander, black pepper and red pepper. Thread onions on eight 6 inch skewers. Set aside. Grill the chicken on the rack of an uncovered grill directly over medium heat for 12 to 15 minutes or until chicken is tender and no longer pink, turning and brushing once with the vinegar mixture. Place kabobs on grill rack next to chicken the last 5 minutes of grilling, turning and brushing once with vinegar mixture. Serve chicken and kabobs over rice. If desired, drizzle with preserves (Michele cooked this recipe by marinating the chicken in the vinegar mixture and then broiling it and it was delicious).