News from CureGN

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Sponsored by the National Institutes of Health (NIH)
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Thank you for your time and contributions to CureGN. As you may be already aware, the CureGN study is a huge effort aimed to further the understanding of rare forms of kidney diseases, including minimal change disease (MCD), focal segmental glomerulosclerosis (FSGS), membranous nephropathy (MN) and IgA Nephropathy (IgAN). Please find below some updates about this important, one-of-a-kind study.

The CureGN study is being performed jointly by 4 participating coordinating centers (PCC): The Midwest Pediatric Nephrology Consortium (MWPNC), Columbia University, University of North Carolina, and the University of Pennsylvania. Each center may have several enrolling sites. We would like to introduce you to each of our centers over the next several newsletters.

University of North Carolina Profile

The UNC PCC's efforts are centralized at the UNC Kidney Center. The UNC Kidney Center is known as a Center for Excellence for Glomerular Disease and Vasculitis, which means there is team of medical faculty dedicated to providing the highest quality of clinical services for these diseases.

The UNC PCC includes four academic medical centers and one community practice located in the southern United States and Canada. All of our sites enroll adult participants; the UNC Kidney Center enrolls both adult and pediatric participants. Our collaborating clinics are: University of Alabama at Birmingham, Vanderbilt University in Nashville, Tennessee, Virginia Commonwealth University in Richmond, Virginia, Columbia Nephrology Associates in Columbia, South Carolina, and Hôpital Maisonneuve-Rosemont/University of Montreal in Canada.

Enrollment

Clinical research studies like CureGN depend on you!

As of 11/28/2016:

Total Enrolled: 1633

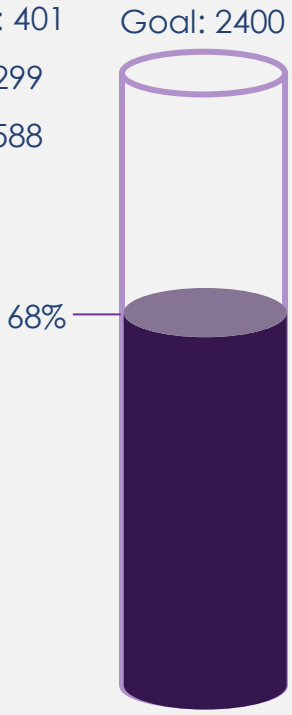
Totals by disease:

MCD: 345

FSGS: 401

MN: 299

IgA: 588



Additional content can be found on our website CureGN.org or at Nephcure.org.

The UNC PCC clinicians, researchers and study coordinators are dedicated to continued enrollment into the study and to working with the overall CureGN study to conduct high-impact research towards improving the lives of patients with these diseases.

Patient Profile - UNC

Karin, diagnosed with IgA nephropathy (IgAN) in November 2015, is glad to be a patient at the University of North Carolina Kidney and Hypertension Clinic. She recalls that after finding out about her diagnosis, she was afraid and had many questions and concerns about how IgAN would affect her and her family's lives. She spoke openly with her doctors at the clinic, and they were able to lessen her concerns and give her a solid understanding of IgAN.

Karin was pregnant with her first child when she started having symptoms of IgAN. At first, she thought the swelling in her hands and legs was simply due to being pregnant. She also had elevated blood pressure and migraine headaches. When the symptoms didn't improve as her pregnancy progressed, physicians decided to perform a C-section in the hopes that Karin's condition would improve. However, her symptoms persisted after the birth of her son and she was referred to the University of North Carolina.

Karin was hospitalized for a kidney biopsy and a series of various treatments. She admitted she was afraid at first and concerned how the disease would affect her in every aspect of life. This anxiety was magnified by the excitement and stress of becoming a new mother and concerns about how IgAN would affect this new role. Karin recalls being worried about her and her husband's careers; they were pursuing jobs that required moving internationally. The idea of finding nephrologists in all of those locations was scary and potentially an impossible task. Karin continued to come to all of her appointments, take her medications as prescribed and make lifestyle changes recommended by her physicians. Over time and after many office visits, Karin is much more comfortable with having IgAN and is an active participant in CureGN.

Today, Karin is doing very well and has adjusted to both living with IgAN and being a parent. When asked how her life has changed, Karin states that daily medication is one of the biggest adjustments. Regular doctor's visits have become a part of her life, but Karin is ok with that, especially because she is now doing well and the appointments have become less frequent. Becoming more aware of her health and making changes to maintain a healthy lifestyle have been positive outcomes. One of the biggest changes she has made is her diet. As a result, her family's diet has also changed as Karin has started to choose different healthy foods and prepare them for herself and her family. She is thankful for the support of her family; her diagnosis of IgAN has led her to bond with a family member who has a similar condition. At her last visit, Karin said that she and her family are relocating to the western part of the United States so her husband can pursue a job opportunity. Though it is not overseas, Karin is happy for the adventure that moving across the country will bring.

CureGN Disease Spotlight:
IgA Nephropathy (IgAN)

Immunoglobulin A nephropathy (IgAN) is the most common primary glomerulonephritis in the world. It is also known as Berger's disease, named for the French pathologist who first described the disease in 1968. Its prevalence varies across populations - highest among Asians, followed by Caucasians and very low among Africans. The diagnosis of IgAN requires a kidney biopsy. Special staining of the kidney tissue shows IgA stuck in the filtering structures of the kidney, the glomeruli. The tissue findings of a kidney biopsy are helpful for physicians to predict the long-term risk for more damage in the kidney that will lead to kidney failure requiring dialysis or transplantation.

Many patients with IgAN, especially those younger than 20 years, first become aware of a kidney problem when they see blood in the urine, usually during an infection of the nose, sinuses or throat (upper respiratory tract). In other patients, evidence of kidney disease includes protein in the urine (proteinuria) or blood in the urine that is only seen through a microscope. Rarely, in about 10% of cases, patients will have large amounts of protein in the urine, associated with swelling in the legs and a high cholesterol level. This is also known as having the nephrotic syndrome. In children and some adults, IgA collects in small blood vessels in the skin, intestines and joints, in addition to the kidneys. The resulting inflammation at these sites causes a red raised rash, joint pain, stomach pain, and the same type of IgAN kidney damage. The combination of these findings is called Henoch-Schoenlein purpura nephritis (HSPN), named for two German physicians who first described these physical findings in the 1800s. About 5% of patients with IgAN or HSPN have a relative with the same disease.

Patients with IgAN who have modest amounts of protein in the urine (less than 500 mg/day) will most likely have good kidney function throughout their life. Patients with greater amounts of protein in the urine may lose kidney function over time with about a third progressing to end-stage kidney failure within 20 years.

As for any other kidney disease, maintaining a healthy lifestyle is recommended: avoiding tobacco, exercising regularly, and maintaining control of weight and blood pressure. Physicians often prescribe medications called ACE inhibitors or Angiotensin Receptor Blockers (ARBs) to lower blood pressure and reduce the amount of protein in the urine. Keeping the protein level in the urine low will help slow the decline in kidney function. Occasionally, treatment that suppresses the immune system is needed to reduce the damage in the kidneys.

Research has helped physicians better understand why IgAN develops. IgA is made by our immune system, and we all have it circulating in our blood. We do not understand as yet why exactly it deposits in the kidney of some people to cause kidney disease. A lower content of a special sugar (galactose) on the IgA protein is recognized by the immune system as abnormal in those with IgA deposition in the kidney. Unfortunately, eating more galactose does not fix the problem. Ongoing research will improve the understanding of what causes IgAN, with the goal of developing future disease-specific treatments and hopefully preventing the disease altogether.