Midwest ARPKD/CHF Family Social

Join us in Kenosha, Wisconsin on **Sunday, October 3, 2004** for the 1st Midwest family social for ARPKD/CHF families. Kenosha is located on the shores of beautiful Lake Michigan in southeast Wisconsin just 60 miles north of Chicago and 30 miles south of Milwaukee. This is a great opportunity for fellowship and fun with other ARPKD/CHF families. Forming a Midwest social group can also provide ongoing support for families when faced with the many concerns and challenges of this disease. Kenosha is home to great outlet malls, recreational, cultural and artistic attractions. Mark your calendar for a fun-filled fall family get-away! Here's a tentative schedule of the day's events:

1:00 pm:We'll meet at Apple Holler Orchard & Farm to get acquainted
2:00 pm—?: Group hay ride through the 50 acre apple orchard where we'll stop off for a family-style picnic in the orchard with hot dogs, burgers and all the fixins'. There'll be games and activities for the kids while parents talk and get to know one another.

Families are welcome to either come early or stay after the cook-out to enjoy the orchard. They have pony rides, a petting zoo, mini golf, train rides, a pumpkin farm and more.

For further information about the orchard visit: www..appleholler.com
For information on accommodations and other attractions visit: www.KenoshaArea.com

Advanced registration required, please respond by Sept. 17th
Please contact Linda Bevec
(765) 567-7485

Philadelphia Marathon Race

On December 3, 2003, Ryan Hutchison ran in the Philadelphia Marathon. Ryan and his wife Wendy are parents to two year old Abby with ARPKD/CHF. Although this race was not planned as a fundraising event, Ryan and Wendy sent out letters asking family and friends to sponsor Ryan's race. This letter generated $3,600! A huge thank you to the Hutchinson's for taking the opportunity to support the ARPKD/CHF Alliance's mission and purpose. The ARPKD/CHF Alliance is deeply grateful for their support.

"Walk/Run for Your Cause" and "ARPKD/CHF Social"

It was with great regret the "Walk/Run for Your Cause" and the "ARPKD/CHF Social", both scheduled for September 20, 2003 at Spring Gulch Camp Resort in New Holland, PA was canceled. Although impending Hurricane Isabel dominated the news, another significant weather event occurred days prior. A severe storm produced torrential rain, significantly damaging the "Walk/Run" course and areas set aside for the "ARPKD/CHF Social". Repairs were delayed to prepare for Hurricane Isabel's arrival, which caused further damage. The ARPKD/CHF Alliance would like to thank our sponsors: Spring Gulch, Quarryville Family Dentistry, New Holland Auto Group, Shady Maple Smorgasbord, and Susan Lawrence, NY, NY. Sponsors and signed up participates were notified of cancellation and although the event was a washout, our sponsors were not.

Pennsylvania ARPKD/CHF Social

WHEN: August 14, 2004 beginning at 10 am

WHAT: A fun day filled with children and adult activities in a relaxing atmosphere for ARPKD/CHF family, friends & professionals. A $15.00 donation is suggested per family to attend, for food and activities.

WHERE: Mill Bridge Village & Camp Resort (www.millbridge.com), is the oldest continuously operated historic village in Pennsylvania Dutch Country. Strategically located within minutes of all major Lancaster County shopping and attractions, including a children's theme park, in beautiful Lancaster County (www.padutchcountry.com).
Mark your calendars and make a weekend of it!

RSVP to: 717-687-8181 or 1-800-645-2744 **by July 31st**
QUESTIONS: 717-529-5555 or info@arpkd.org

Hidden Grief when Parenting a Chronically Ill Child

The grief of parents whose child has a serious or chronic illness is often hidden. In the busyness of caring for all the medical and emotional needs of a sick child, parents can often put their feelings on hold. Because few people, including family, friends, and professionals, take the time to inquire about those feelings, parents can begin to believe that their emotional responses are abnormal and that no one feels as they do.

However, it is very common for intense feelings of grief to be triggered for parents, not only at the time of diagnosis, but also over and over during the illness. This can happen at medical crises, at times of missed opportunities for their child, and even at simple occurrences such as seeing another child who is healthy.

Parental grief is not simply feelings of sadness and pain. It includes times of shock, confusion, numbness, memory loss, anger, fear, anxiety, and guilt. These and many other feelings are normal responses to dealing with an abnormal situation that has so many unknown factors. Because all parents seem to have a basic belief that a "good parent" should protect a child from illness and suffering, many parents blame themselves when their child gets sick. It is hard work to unlearn and ignore this kind of myth.

Family members also express their grief in many different ways. Often this can be a source of friction, especially if, for example, one parent needs to talk, while another needs to withdraw. It helps if each parent can recognize his or her style of dealing with grief, and can give one another the freedom to be different. Having other people to share with often helps parents to do this more easily.

Other methods for parents to deal with grief include: becoming as informed as possible about the illness and ways to live with it, being a part of the treatment team, joining a support group of other parents of chronically ill children, and learning to reach out for help to others in the community.

~ Reprinted from the HOME CARE FAMILY NEWSLETTER, Children's Hospital, Philadelphia, PA

Editor's Note: This article first appeared in the ARPKD/CHF Newsletter in 1996 and is now located on the ARPKD/CHF Alliance's website. It generated much discussion at the time.

ARPKD/CHF Alliance T-Shirts are available for $15.00. To view, see them at www.arpkd.org. Get them while they last!

Afghan Raffle

A new and beautiful 7' X 7 1/2 ' afghan, handmade and donated by Susan Walushen, aunt to an ARPKD child, will be raffled off. Tickets are $5.00 each and a maximum of 500 tickets will be sold. Proceeds will benefit the ARPKD/CHF Alliance. Please complete the form and mail it to: *Beth Hall, 290 Girard Ave., East Aurora, NY 14052.* Your name will be entered according to the number of tickets bought. Winner will be announced. *(Please print clearly.)*

Name: _____

Address _____

Phone: _____

Number of tickets entered: _____

The ARPKD/CHF Alliance is grateful to it's supporters for their generosity. We extend our deepest thanks to our 2003 contributors

Domenick & Mary Ellen Abbate
Gary & Dawn Adelman
AM Silver Linings Inc.
Beth Bailey
Robert & Patricia Bailey
Jon & Beverley Benda
Frank & Linda Bevec & family
James H. Borg
Phillip and Lisa Brudos
Eugene & Lillian Bryson
Laurence & Janet Carroll
John & Kristen Carter
Modestino, Aka, Donna Carullo
Lawrence M. Cohen
Joe Costanza
Helga Dammann
S.M. Delanoy
Alfred & Annamaria Dilapi
Charles Dolle
Robin Helfritch & David Euler
Debi Miller
Mary L. DeLanoy
Gloria Dengler
James & Noreen Dengler
Loretta Dengler
Alfred & Annamaria Dilapi
Donna Greenly
James & Carol Dvorchak
Julie Brandt & Eric Coger
William & Hazel Ernestine
Raymond & Peggy Ann Ernst
Janet Taubin
Christine Filippini
Michael F. Gaughan
Howard & Julia Glasser Parker
James & Julie Gombas
Antone R. Granada
Zachary & Judy Lee Greenhill
Robert Grosshandler
Scott & Melissa Hames
David & Ruth Harrison
Herbert & Diane Heffner
Dean & Kristin Huibregtse
Dale & Dorothy Ishol
Gila Ivey & Christopher Selicious
John & Ada Kennedy
Wayne & Roberta Kladerman
Stefan Kopka & Barb Wetula
Glenn Kressner & Marcia Pepper
Anil & Tracey Knight Narang
Glen & Marcia Kressner
Jonathan & Nicole Kuhn
Theodore Kuliczkowski
Rick Lengold
Jeffrey H. Lipton & Randi S. Rosner
Warren & Darlene Lea
Stephen & Dipika Mascarenhas
McDonald Investments Foundation
Keith S. Marks & Kristen E. Neary
Sarah Martemucci
Stephen Mascarenhas
James McKillop
Christopher & Cheryl McGrath
Stan Kastrup
Melissa Keiser
Larry & Debra Menaker
Bob & Marilyn Miles
Harold & Linda Moehringer
Frank & Roe Mondaruli
Michelina Mondaruli
Patrick & Sharron Mulvihill
Stephen Nagal & Heather Hoffpauir-Nagel
Anil & Tracey Narang
Vanessa Nastri
Michael & Rosemarie Navatta
Kristen & Keith Neary
New Holland Ford, Lincoln, Mercury
Nicole Williams
William & Lori O'Neill
Mark A. & Diane M. Nelson
Donna Ochenkowski
Todd & Beth Parmington Hall
Mike & Lorrie Patton
William & Ernestine Polland
Joseph Principato & Nancy Dengler
Quarryville Family Dentistry
Christopher & Julie Reising
R. Renzetti
Julie Reising
Debbie Relis
Cynthia P. Saxman
Frank & Carol Seidl
Frank & Patricia Seidl
Shady Maple Smorgasbord, Inc.
Lillian Smith
Mark & Lori Spier
Spring Gulch Inc.
State Bank of Long Island
Russell & Daryl Stern
Stoltzfus Feed & Supply
Janet Eskolsky Taubin
Barry & Debbie Telis
Heather Trussell
George & Karen Tzanetopoulos
David & Loretta VanHoose
Volunteer Center Of Sonoma County
Stephanie Wagner
Lucille & Robert Walker
Barbara K. Wetula
David & Cindy Wittels
Sue Yearick
Chris & Michele Young
Gerald Zak

In honor of Maria Bernardo's 70th birthday:
Marge Charlton
Warren & Darlene Lea

In honor of Mr & Mrs. James S. Brown:
Weldon & Allyson Showalter

In loving memory of Hannah Deming:
Janice Croisetiere
Gerard & Elaine Daigle
David & Stephanie Deming

In loving memory of Alexis Hall
Darlene Frost
Jon & Shannon Hall
Merle Frost

In honor of Hannah Hall:
Merle Frost

In loving memory of Matthew Huibregtse:
John & Karen Carroll
Dean & Kristen Huibregtse
Warren & Darlene Lea

In honor of Abigail Hutchinson's 2nd birthday:
Ryan & Wendy Hutchison
David Euler & Robin Helfritch
Denise Fujii

In loving memory of Chloe Fujii:
Laura Alvarez
Karen Brandt
Eric Coger & Julie Brandt
Helga Dammann
Steve & Mary Fujii
Brian & Kelly Fujii
Gail R. Gunter
Tiffany Halterman
Anita Helfritch
Dennis & Iris Helfritch
Garret & Jen Helfritch
Aloha Jackson
Kevin Missak
Maureen Motzny
Marissa Pflueger
John Prestia
Carol Robertson
Mike & Natalie Sanaghan
Eva Sanchez
Mike & Kate Skibbie
Carole Skibbie
Ron & Sue Skibbie
Dale Solyom
John Spiteri
Jennifer Struif
Laurie Wit
Carleen Witmer

In honor of Lucy Gombas:
Silver Linings

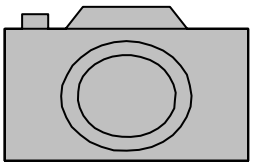
Ryan's run for ARPKD, to honor daughter Abigail:
Chuck & Leanne Augustine
William & Nancy Barba
Bryan & Heather Benson
Jason & Rebecca Bernstein
Gordon & Gail Bilsland
Rich & Jennifer Boyd
Harry & Laurie Brown
Richard & Lynne Brown
Shawn & Melissa Burger
Pamela Burke
Jared & Jared Butler
Jennifer Clark
Dennis & Diane Coston
Stephen & Donna Cupitt
Kenneth & Sally D'Amico
Jeff & Debra Dearie
Kathy Diodati
Rhodell Domilici
Harford & Bonnie Drexler
Brent & Jenny Drollette
Pete & Jenny Eibeck
Raymond & Peggy Ann Ernst
Clare Fannon
James & Linda Frank
Bruce & Deborah Finn
Glenn & Jodi Frazier
Helen & Paul Gillespie
Donna Greenly
Christina Greenwood
James & Kathryn Howard
Chung & Susie Huang
Ryan & Wendy Hutchison
Steve & Michelle Hutchison
William & Hazel Hutchison
Dennis & Sheila Hutchison
Marc & Beverly Kauffman
Brian & Jennifer Keehbler
Melissa Keiser
Alan & Henrietta Kiel
Jennifer Kiel
Kathleen Kiel
Jennifer Kiel
Brad & Barb Kiel
Diane & Richard Krause
Lynn Kress
Robert & Darlene Osmun
Jennifer Outland
Elizabeth Langan
Lori Lennon
Deborah Lepone
Nordis Lindbloom
Todd & Beth Longreen
Harry & Marie MacLacklin
Debi Miller
Jarret & Lisa Miller
Steve & Kathy Moelter
Tom & Christine Newman
Marjorie Perry
Stephen & Rosanne Rabbitt
Bob & Lisa Reiser
Bruce & Sandy Roberts
Bryan & Lisa Rosica
Tom & Jennie Roule
Judith Russell
Paula Scott
Scott & Linda Sepsy
George & Dody Shaw
Thomas & Linda Terry
Walt & Judy Samsoe
Victor & Frances Sassano
Mark & Blanche Speiser
Lee & Jane Schondelmeyer
Carrie Schreck
Steve & Mary Spoerle
Jim & Anne Shute
Ken & Paulette Staub
William & Laura Viel
Robert & Lucille Walker
Robert & Karen Wallace
Phil & Joanne Waldron
Nicole Williams
Dennis & Dolores Wismer
Denise & Gerry Wurst
Sue Yearick
David & Melissa Yates
Chris & Michelle Young
Paul & Michelle Zotter

In loving memory of Wm. George N. Sanders:
Mr. & Mrs. T.E. Bulley
Mr. & Mrs. B. Sanders

In honor of Rita Seidl:
Mary Beth Selfert

In loving memory of Larry Spier:
Fred E. Ahlert, Jr
Daniel & Kara Alpert & family
Carmine & Robin Esposito
Sharon D. Feldman
Natalie Gordon
Fred Ahlert Music Corp.
Harry Fox Agency
Holee Music Co.
Glenn Kressner & Marcia Pepper
Robert Margolies & Carolyn Schatz
Mordechai & Miriam Mayerovitz
Robert Moon
Lila Nathan
Prager & Fenton
Gia PrimaEdward
June Robinson
Dale Solyom
Barry & Barbara Synder
David Sugita
Melodie Der Welt

In loving memory of Jason Thomas:
Robert & Linda Thomas



PHOTOS NEEDED

The ARPKD/CHF Alliance is working on website & educational material and we would like to include your ARPKD/CHF child's picture. If you have a clear picture you would like us to include—please email it to: info@arpkd.org . Include a brief statement giving us permission to use the photo(s).

The first successful transplant occurred in 1954! It has been 50 years since the first successful kidney transplant!

LISTSERVS:

Here is how they work... register for free, then submit messages or participate silently (just "listen" in). Submitted messages are distributed to all list members for reading and potential reply, with the exception of **"ARPKD/CHF Reporting".**

ARPKDLOSS ONELIST- for parents who have lost an infant, register at: http://groups.yahoo.com/group

ARPKD ONELIST—for parents and affected individuals, register at: http://groups.yahoo.com/group. Subscribers must be approved.

ARPKDadults-for adults living with ARPKD and CHF, register at: http://groups.yahoo.com/group/ARPKDadults. Subscribers must be approved.

ADpkdchildren-for families with infants and children affected by ADpkd, register at: http://groups.yahoo.com/group/ADPKDchildren

NEW: ARPKD/CHF Alliance Reporting". This listserv will provide electronic newsletters and information. It is not an interactive listserv, where members can post messages, only the ARPKD/CHF Alliance will post information. See page 2 for more details.

You can help spread the word about life-saving organ and tissue donation. April is National Donate Life Month.

Tell your family about your decision to donate!

Tidbits of information on Congenital Hepatic Fibrosis

Liver biopsy and endoscope are no longer considered routine procedures to diagnosis CHF. Both procedures involve some risk and don't effect or change course of treatment.

Varices can be visualized thru an endoscope. Some GI specialists suggest banding or using scleratherapy every six months, but if pressure is removed, it may go elsewhere and create varices. Do you become proactive, by scoping and treating varices year after year, or leave them alone? Some varices exist for many years without rupturing.

Inderol (propananil) can help ease portal hypertension, however, in order to be effective, a sufficient dose is needed to drop the heart rate by at least 25%. This produces a significant decrease in heart rate, which can cause light headedness, and may not allow the heart rate to speed up and compensate during a varice bleed, vital in shock.

There are no treatments or therapies to help ease portal hypertension, a complication of congenital hepatic fibrosis, with the exception of liver shunting/transplantation. For more information, visit our website: www.arpkd.org.

This newsletter is sponsored entirely by donations. The ARPKD/CHF Alliance does not charge for membership and all services/programs are free . Contributions allow us to continue our important mission–to educate, advocate, support and advance research specific to ARPKD/CHF, our purpose is to improve the lives of those affected. Here are some ways you can help:

- Make a tax deductible contribution to the ARPKD/CHF Alliance. Make checks payable to: ARPKD/CHF Alliance. Mailing address: P.O. Box 70, John Drive, Kirkwood, PA, 17536
- Designate the ARPKD/CHF Alliance on your pledge form to the United Way. (United Way administration take out fee is approximately 10%, depending on your location.)
- Double your donation, arrange for a matching fund donation through your employer.
- Place collection cans in stores (with store owner's permission) marked: ARPKD/CHF Alliance. Follow through and regularly empty cans.
- Sponsor/organize a Fund Raiser: a Benefit Yard Sale, Book, Craft or Bake Sale, a Car Wash, Bike-A-Thon, Raffle, Auction, Nickel/Dime Drive, or Black Tie event.
- Plan a Fund Raiser in conjunction with another organization: Lion's Club, Kiwanis, 4-H, Elks, Rotary, American Legion, Moose Lodge, VFW, Chamber of Commerce, Nights of Columbus, Boy/Girl's Club, and Youth & Women Groups.
- Help fund the "Professional Educational Project" - Getting information to as many professional communities as possible. See www.arpkd.org for more information.
- Talk to family, friends, co-workers and neighbors about ARPKD and CHF! Tell them about the ARPKD/ CHF Alliance, our purpose and mission – someone may be looking for an organization to help!

Honoring or Remembering Someone in a Special Way

Do you know someone you would like to honor or remember in a special way?
A donation made *in memory of* or *in honor* of a special someone or occasion is a wonderful way to give a gift and support the ARPKD/CHF Alliance at the same time. We will acknowledge your kind donation with a thank you letter and receipt. Please complete the form below to make a tax-deductible gift. Thank you for your support!

A GIFT MADE...

In memory of _____

In honor of_____

In celebration of _____

(Birthday, Graduation/Congratulations, Anniversary, Retirement, Job Well Done, Other)

Please send an acknowledgement to:

Name_____Address_____

City_____State_____Zip_____

From _____

May we acknowledge your gift in our newsletter or on our website? Yes No

Would you like to be on the 'ARPKD/CHF Alliance Reporting' listserv? (See page 2 for details)

Yes, my email address is: _____

Clip or copy and send to ARPKD/CHF Alliance, P.O. Box 70, Kirkwood, PA 17536

In Loving Memory of Calla Coetzee 1999/12/21 to 2003/07/11

I was married at the age of 26 knowing that I will never have my own kids – my female insides were "all wrong". Yet, my beautiful healthy baby boy "Calla" was born 34 weeks premature due to absence of amniotic fluid - on 21st, December 1999. During my prenancy, none of the ultrasounds indicated large echogenic kidneys.

After just 12 days he started crying – according to the Doctor nothing was wrong. On Sunday the 9nth of January 2000 he was operated on for a hernia inguinal unilateral and the pediatrician discovered that there was something terribly wrong with his kidneys the were grossly enlarged and still getting bigger! Fortunately, for us she studied under a brilliant Professor Peter Thomson for a few months and she told us that the possibility of Calla being ARPKD positive is 50 %. Little did we know what that mean and what an effect this will have on our lives. We visited the Professor for the first time on the 2nd, February 2000, and it was confirmed that Calla was ARPKD positive and had a blood pressure of 160/140. ,He was immediately put on Lopressor, Minipress, Norvasc, Renitec, Propranolol, and Triprim and everything came in pills and had to be given twice daily. So we went home with this small little baby and I started giving him this handful of crushed pills and he started getting better. The next blow came when we were told that he also suffered from Congenital Hepatic Fibrosis (enlarged liver) and splenomegaly (enlarged spleen due to increase pressure in the liver) and esophageal varices having the potential to bleed suddenly and unexpectedly into the esophagus. When Calla was eight months old, my husband was seperated and I finally filed for divorce (big mistake). I asked my mother if Calla and I could live with her.

What nobody prepared us for was the four weekly visits to the specialists in Johannesburg. They drew blood every time and as he grew older he started crying every time we walked into the specialists rooms because by then he knew what was going to happen. If they were not satisfied with the results, we went home, came back after two weeks and had to send blood samples every week. Urine samples was taken every second day. Finally, my mom gave me a blood pressure monitor and I didn't have to go to the local hospital for blood pressure monitoring. If he ate something funny (he hardly eats at all), he would start vomiting. He weighed a mere 8kg on his 1st birthday. He never crawled but moved around on his but and finally started walking at 19 months.

Calla was in and out of Hospital with peritonitis, rotavirus and lung infections. In March of 2002, he was again in hospital for the rotavirus and when he vomited, blood came with it. According to the doctor, the amount was not sufficient enough for the varices that might be bleeding but he was operated on for "sclerotherapy" a week later. He hardly gained any weight as for his features he's got the typical small jawbone and lower set ears. Until the 30th of April 2002 nothing really astronomical happened but on the night of 30th of April 2002 our real nightmare started. Calla started vomiting blood. We went to our local hospital where they stabilized him and we took him to Pretoria to another specialist the following day – luckily for us we found another because our dear Professor Tompson already left South Africa in February so we were between doctors. In Pretoria on that dreadful Wednesday we met a Dockter Ida van Biljon and she booked us into a wonderful hospital "Medforum" in Pretoria. Calla was in the Neunatal Pediatric Intensive Care Unit from the Wednesday 1st of May until the 5th of May 2003. A Pediatric Surgeon by the name of Maartin van Niekerk gave him extensive endoscopic sclerotherapy and according to me saved his precious little life. However, Calla also received four units of blood – he was still bleeding from his varices when they operated on him. Dark tarry like blood stool was still to be found in his diapers for about two weeks after he received sclerotherapy. Exactly two weeks later, we had to go back for more sclerotherapy. From that time on, we had to go back every four weeks for the endoscopic sclerotherapy. We now also visited he's knew pediatric nephrologist doctor Lionel Mattheyse every 4 weeks. Therefore, every four weeks we made an appointment for the Monday for the schlerotherapy and on the Tuesday, we would visit the nephrologist. So with blood and urine samples and a crying little two and a half year old we sat of to Pretoria every four weeks.

When we came home after that ordeal in May my son just never really recovered. His list of medicines looked like this: Propranolol 10mg 3x per day (portal hypertension), Hyperphen 10 mg 2x day, Lopressor 50 mg at night, Minipress 1 mg 2x per day, One alpha 1 mgc. in the morning, Titralac 3x per day, Norvasc 5 mg 2x per day, Renitec2,5 mg at night, Losec mups 5mg at night, Folic acid (pure vitamins) and he had be injected with Eprex 2x a week. All except for the Titralac and One alpha was for he blood pressure and the Losec mups (antacids) so that his stomach acids will stop eating away at his insides.

Life started being a real hellhole. Between him crying every time I had to give him his medicines and being injected on either his

(Continued on page 18)

Life's Adventures With ARPKD/CHF – From a Fifty-Year-Old Woman . . .

April 19th, 1953. Exactly three months after Lucy gave birth to "Little Ricky" on T.V. and Desi Jr. in real life, I made my own appearance in Chicago at 6 lb. 3oz. back in the day when there were no ultrasounds and my parents were blissfully unaware that anything was amiss. Their firstborn, I seemed perfect to them. As a youngster there were only a few things that I can ascribe to the ARPKD/CHF in hindsight. Colic as an infant. Always thirsty (still am). I also seemed to get sick more often than my sisters and couldn't keep down anything too acidic. I mix juices with half water. That was about it until I had a blood test at the age of twelve (almost thirteen) and it was discovered that I had an alarmingly low white count, or leucopenia. This led to hospitalization for blood tests and eventually a bone marrow aspiration that revealed no cause. It was discovered that my spleen was sticking out from under my rib cage. At the time it was decided that "it was just natural for me." This was back in 1966. I've read since then that this condition was not given a name until 1961, and so the lack of knowledge at that time is understandable.

The next issue to surface was at fifteen when I tried to join G.A.A. or the Girl's Athletic Association in high-school. There was a cursory physical and the school nurse was concerned about my blood pressure saying it was way too high for someone my age. She wanted me barred from participating. I was so aggravated, and got a note from my family physician saying it was O.K. for me to join. Now looking back I realize that this nurse was the only one on the ball and a very good nurse. No one paid any attention to her. Finally at seventeen I was put on medication for high BP. This was continually increased without achieving control so that by the time I was mid-twenties my then doctor said it would be morally irresponsible to increase it further without more investigation into the cause considering my youth. Again a lot of blood tests including one where a catheter was threaded up from my leg to shoot dye right into the kidneys looking for a blockage in the veins or arteries. I was told nothing of that sort was seen and so that was that. My BP medications were continually raised for a number of years and helped lower it but never really got it under the kind of numbers they wanted. By that time I smoked too (still do, I know..... I don't eat fruit or vegetables either.....)

During these years I started getting kidney stones, the first one when I was 18. A nursing student I worked with told me a lot more than any doctor ever had, saying to drink lots of water, never put off going to the bathroom, and avoid all colas. My kidneys felt better than they ever had, they often caused me pain and discomfort that I didn't realize wasn't normal. I continued to get the stones infrequently at first, and then eventually at least once a year in each kidney or twice a year. I almost lost my job over the time missed. I would have to go to the hospital for the pain shots and always passed the stone on my own the third day. It was during the kidney x-rays with dye one of these times that I first learned my kidneys were oversized. An x-ray tech asked me how tall I was and I told her not that tall, 5'5&1/2". She said it was unusual that my kidneys were big enough she couldn't fit them on one x-ray plate and if I had more in the future to tell them to use two plates sideways. I forgot to mention, at the age of 22 I had been told that I had “sponge kidneys" and that it was nothing to worry about.

Finally the stones became such a problem I was sent to the Stone Clinic at the University of Chicago Hospitals. This was very detailed. They asked if I had collected any stones and I said yes I had quite a little stash of them which were sent them off to be analyzed. There were two days of very exacting urine collections and blood tests. Meantime a CT scan of the kidneys was ordered. It was during this CT scan that I first learned the seriousness of the problem with my kidneys. The tech came over and started asking a lot of questions, like had anyone ever said anything to me about cysts in my kidneys? I said no and he said hold on, he was going to call my doctor. Then he came back and told me they were going to do the whole thing again after I drank some contrast material. You have to remember that during these years imaging techniques were getting better all the time. Eventually the doctor told me that the PH of my urine was slightly acidic, and the composition of my stones was not calcium as it usually is. I was put on a prescription of a systemic alkalizer and have not had another stone since. He also told me the CT scan showed that I have Polycystic Kidneys. Whoa, I had heard of this before when I looked up "sponge kidneys" and remembered thinking "I'm glad I don't have that." But then I realized that nothing had really changed, my kidneys were the same as they had always been. The only thing different was the fact that I was a lot more informed about them now. And with that I went back to not thinking about them unless a problem came up.

Moving up to 1993, I was supposed to remind my Doctor that I was to have a follow-up scan of my kidneys, six months after the last one. So I did remind her, and she said well she didn't know if it really mattered if it were six months or a year. I just shrugged my shoulders and said O.K. But then on the way home I was thinking about it, what had the original shown that prompted the suggestion of a repeat anyway? I called the office when I got home and requested a copy of the original report. [This was in the advent of HMO's]. I quickly learned the value of asking to see a report for myself. Now I was to see the doctor to get the report and meantime make an appointment for another scan. Turns out when the original was done whoever read it said something on there did not look right about my liver. The words were big and medical and I didn't know quite what to make of it. But after the scan I was scheduled to see a gastroenterologist, who ordered a detailed MRI of the liver. This was followed by the news that a biopsy of my liver was next, only the architecture of my liver was so screwed up it could not be done in the usual way by a needle through the side, but by abdominal laporoscopy, otherwise they wouldn't be sure if they hit the liver or not.

(Continued on page 19)

*(Continued, "*In Loving Memory of Calla Coetzee", *from page 16)*

legs or very thin little arms and me still being in a full time 8-17:00 day job he was slowly but surely slipping away from us. He was totally anemic. He had terrible nightmares, hardly ever played any games and didn't want anything to do with other people except myself, my mother and at this time he and he's daddy's relationship was starting to pick up beautifully.

For me a lifesaver had always been the fact that he was still on formula milk so at least I knew that he got in some food even if it was just in drinking form. Next thing the doctor asked me to either take away his formula milk or see to it that he only takes in 400ml per day. He wanted this little guy to gain weight because know we were starting to move in to the kidney transplant direction and they want him to weigh at least 20kg. My heart stopped but I went home and we tried it. By that time, he weighed 13kg and I started taking away the milk. Within 4 weeks time he lost 2kg. 21st December he turned 3 years. January 2003 we went back again to the specialists and he decided that Calla now should be injected with Eprex (in order to get some kind of better red blood count) 3 times per weeks. By then he had 23 % kidney function left. January and February 2003 passed us with no better results and February 2003 we decided that test should be done so that he can go on dialysis.

On Monday the 3rd of March, 2003 Calla went into renal failure. Tuesday 4th of March 2003 we left for Pretoria, was admitted to the Zuid-Afrikaanse Hospital pediatric ward and the real hell started. At that stage Calla didn't walk by himself anymore, wasn't eating at all and just taking in fluids. On the Thursday they drew 14 small biles of blood and put him on a catheter in order to see if he was still passing "good" urine. For 48 hours my son did not move, talk or ate anything, he was just too ill. After 48 hours, they took the catheter out. The results came back and they confirmed that he was in renal failure!

On Friday 7th of March, they operated for dialysis. Calla did not take the operation very good and they had a few hick-ups during the procedure. It took quite a while to wake up and then the professor who operated on him, Professor Karriseitte, came to tell me the last of the "tale". He inserted the dialysis pipe but while operating he discovered that Calla's "membrane" was overgrown with tissue. The tissue scarring is so bad that it is covering his stomach (explaining why he is always complaining of tummy ace and does not want to eat); he is liver, both kidneys and spleen. The tissue might cause the dialysis to fail and surely, the tissue will start over-growing the pipes. However, the Professor explained that he inserted the pipe and that we will know for sure within the next 48 hours if the dialysis will work or not. The opportunist in me said but if that does not work then we will try blood dialysis. But the Professor explained that he saw Calla's poor hardened spleen and that blood dialysis would not work because of the fact that he's not producing enough white and red blood cells and that we can inject him with, as much Eprex that we want to but it won't help.

During the next 48 hours, we pumped 2 liters of fluid into Calla and only 25ml came out. At that time, his belly outline was 65cm and he was not able to sit or stand. He just lied there. On Sunday morning 9th March, the pediatric nephrologist said that we should once more try to pump 100ml of fluid in. That is when I said no! No more. No more crying no more nothing. This is where it stops! I informed my mother, and the doctor's that I want to see all the pipes been taken out. I then again spoke to more specialists. The truth was on the map. Medically speaking there was nothing more they could do for my son. Not even a transplant would do – he would not survive the operation anyway!

Tuesday 11th March 2003 another surgeon removed everything and told me exactly what Professor Karriseite told me. Nothing could be done; it will better for me take Calla home. He might have 6 weeks to live but that is it. The medical staff advised me to try and give Calla he's medicines as long as possible and to keep morphine in the house for the end stage… all was said and done. On Wednesday 12th of March, 2003 we left the hospitals and the doctors behind and came back home (Potchefstroom). That day I also promised my 3 year, 3 month old son that I will never take him back and never will let anybody harm him again. I will never hold him for somebody to take blood or anything else. When we got home, we held a party on the next Sunday for the family. I started cutting down on all his medicines – he was vomiting every time I gave him the medicines. Since we left the hospital he was injected only once with Eprex and then I stopped that also. When we left the hospital, he weighed a mere 10kg. He had 3 % kidney function left and only passed 1 % of waste through his urine. All his tests were positive of total renal failure.

I am not telling you about the medical bills – in South Africa there is no such thing as free medical assistance. Fortunately, for me I have a very good medical aid but 70 % of my salary every month went for the medical aid. What the medical aid didn't pay the fund I started in January paid for. I'm not telling you about every time I cried for my son for being so frail and helpless and about every time my heart stopped when he vomited or every time we both cried because of the medicines.

Calla died on Friday the 11th of July 2003. When he died his heart was 11cm big and his lungs flooded. We took him to the hospital at 7 o'clock in the morning of Friday the 11th of July, 2003 and he died at 17 minutes to 12. We gave him his first shot of morphine at 9. He died in my arms.

When he died he was off all medication for 3 months, in the first two months he gained 2kg. *(Continued on page 19)*

(Continued, "In Loving Memory of Calla Coetzee", from page 18)

He still was not eating but he drank as much formula milk that he wished. He didn't get nightmares anymore, he laughed and when he felt up to it he played with his toys. The last three months of his life he was happy, we we're happy. A total stranger gave him a Labrador puppy – Calla named her Lilo. So when I started work again in June, she kept him company along with the wonderful black women named Anna, his nanny since he was 3 months old.

Yes, it is true that he suffered from a runny bloody nose from the high toxic wastes in his little body. His urine had a very foul smell, as did his breath. He was a kind of yellow/brown color, and when he died, he weighed only 9 kg. But - he was alive and content. He never cried anymore – yes I do think he suffered, and yes I had to inject him with morphine in June because he got heart cramps from all the excess fluids, BUT this is the part I want to bring over to you – never in this 3 ½ years since his birth had we been this happy. None of the doctors believed what was going on. The signs of death only started the Thursday night before his death. But, after we came back from Pretoria I stopped worrying and wondering when he will die and now more than ever I am glad for each day that I had with him. KNOW THIS IF YOU BELIEVE IN THE FATHER, THE SON AND THE HOLY SPIRIT always remember, no matter the storm, when you're with God, there's always a rainbow waiting.

With all my love to my son – without him, I would never have been this happy, and if I had to choose, I would take him again with every bit of hardship that we went through. To my son- I love you with all my heart!

Annamart Coetzee
(Calla's mom)

(Continued, "Life's Adventures With ARPKD/CHF – From a Fifty-Year-Old", from page 17)

When I woke up there was ALL of my family looking at me seriously. I knew this wasn't a good sign. So I looked at the sister I knew would tell me the truth if she knew that that's what I wanted. "Well?" "Are you sure you want to know, it's not good." "Of course." "Well he said it's bad, really bad. I asked him what about a transplant and he said well that takes time, as if you didn't have enough time. So I asked him how much time are we talking? And he said as if he wasn't sure, two years?" Needless to say, this was sobering news. I was 40 years old. It took me about three days to get over the shock and then you get caught up in the everyday life you were in before you knew this and just accept it, what else can you do? So I made my funeral arrangements complete with a headstone, I designed which is already in place. A cousin who puts artificial flowers on all the family graves each year always puts some on mine and we had the biggest laugh about it the first time I saw it. I demanded nicer flowers for the following year. How often does THAT happen. I can joke about it here because that was all 10 years ago, and I don't seem to be any worse off now than I was then. That doctor told me that my liver biopsy slides had been sent out for another opinion, and came back confirming the initial diagnosis, Congenital Hepatic Fibrosis. He said it was very unusual and my slides were being passed around the city hospitals. My kidneys are the same, 50% compromised, I always say that since one is a spare anyway that puts me at 100% ! I have had the experience of talking to a few parents of young children with this through the years, and no one is happier than them each time I get another decade older.

I asked a Hematologist a few years back how come I can only find info on kids with this disease, does that mean that everyone else is dead by the time they are my age? He just looked at me, he didn't know. And most specialists I have seen are used to seeing patients who are in desperate need of a transplant for other reasons and just blow me off. That is why it is so hard to find information. I think now that there are probably a lot more adults out there who are like me, only they don't know it, just as I wouldn't if I hadn't had all those kidney stones and then someone noticed something about my liver on a scan of my kidneys. I told Colleen I want all the parents out there to hear my story, and see how long I have lived and am still here, and realize that because I was born back when a lot of the technology that informs them today was not available neither I nor my parents went through the anguish I keep reading about. I DO understand that a lot of people with this are a lot sicker than I, but just the same I want you to take heart in my true story.

On December 10th 1999 I was held up at gunpoint by two guys in ski-masks in my own garage when we still lived in the city. I came a lot closer to dying that night than from anything else in my life. They didn't want money or my car, they wanted me to start my car and then ordered me to "get in the trunk." I was very calm and polite and said "no." After they insisted in a menacing way, I calmly told them, "you will have to shoot me then because I am not getting in that trunk." Mentally I reminded myself of what to expect from what I had heard, a trip down a long dark tunnel toward a bright and warm and loving light. I even laughed inside at a joke, thinking about the saying "don't count your chickens before they're hatched..." and how I had been congratulating myself about making it to the year two-thousand. How could I be so calm? I have been prepared to die for a long time now thanks to my ARPKD/CHF. So in a way if I wanted I could say that this disease saved my life by keeping me calm in the face of death. It depends on how you look at things. None of us knows how long any of us has. So please don't act around your kids as if their life is a tragedy. Enjoy the moment, that's all there really ever is. The past is a memory and the future a dream. The only thing real is right now. And right now I'm feeling pretty good! Any questions, write me in the columns at the site! Take Real Good Care. ~ K.P.S.

K.P.S. participated in the NIH study: "Clinical Investigations into Autosomal Recessive Polycystic Kidney Disease and Congenital Hepatic Fibrosis". To hear more from her, join the new listserv: 'ARPKD/CHF Alliance Reporting' - see page 2 for details.

ARPKD/CHF Alliance
PO Box 70
Kirkwood, PA 17536
717-529-5555
www.arpkd.org

For Summer 2004 National Kidney Foundation Transplant Games visit www.NKF.org

For Camp Sunshine: www.campsunshine.org
(Supports children and families with diseases.)