



WINGS

WAGR Information, News, Gorilla Stories

International WAGR Syndrome Association
P.O. Box 392 Allen Park, MI 48101

www.wagr.org
Spring/Summer 2009

From the President's pen

Dear Families & Friends,

I would like to begin by thanking all our donors. During these difficult and uncertain times your kindness and generosity is helping offset many organization expenses.

This includes the printing of our updated brochures, printing and mailing expense of our newsletters, gifts to hospitalized children, general operating expenses, rental space and meals for WAGR Weekend, and assistance for families to attend WAGR Weekend.

The IWSA purchased the QuickBooks online accounting software program to help Treasurer, Jeff Hefty manage our financial business. We also purchased Wrightslaw Webinar CD's. The CD's are available for our families to check out when needed.

Wrightslaw is the leading website about special education law and advocacy. Anyone interested in learning more or borrowing the CD's please contact Educational Consultant, Rhonda Sena at - Casasena5@yahoo.com

We continue to pursue grants as well as creative ways to raise funds for the organization. The IWSA "End of the Year Ask" letter was very successful and we plan to continue this every year.

Unfortunately the March of Dimes Michigan Chapter Community award was not renewed for 2009 due to cut backs. This \$1,000 grant was used in 2008 to offset some of the printing/ mailing expense of our newsletter.

Just a reminder that our online survey "WAGR Syndrome: The Kidney and Urinary Tract" is currently underway. For more information please visit our website at <http://www.wagr.org/survey2008.html>.

On behalf of the IWSA Board and Officers we would like to extend our deepest gratitude to Karen Rose and Rose Mallon, two of our founding members, for the time and energy they donated to make the IWSA what it is today.

The IWSA is a 501(c)(3) organization that accepts both financial and in-kind contributions. Some of the in-kind services we are in need of include: legal advisor, Certified Public Accountant, donated or discounted print services, office supplies, gift cards for sick children and supplies for our annual WAGR Weekend event. If you can help or know of somebody that can help with an in-kind contribution, please email CatherineLuis@msn.com

Calling all UK WAGR Families

The Aniridia Network UK will hold their annual gathering in May. Jenny Langley, ANUK Secretary and WAGR representative would like to know if there are any WAGR families interested in getting together?

Meeting space would need to be secured. If you are interested in a UK WAGR Gathering please contact Jenny at jl_27982@hotmail.com

Details of the Aniridia Network UK 2009 AGM:

Date: 16th May 2009

Time: 10:30am - 4:30pm and optional social event afterwards

Venue: The Resource Centre, Holloway Road, London

Cost: £10 for adults who are members, £15 for adults who are not members. Children go free.

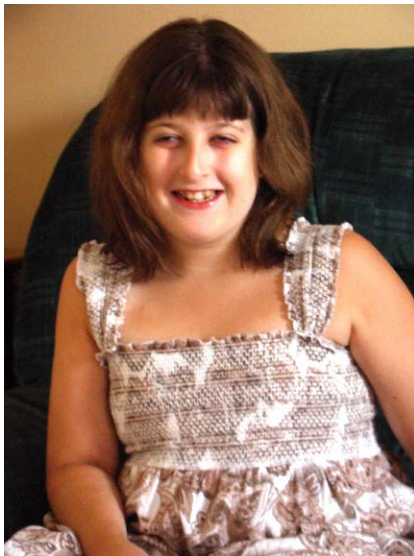
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Silver Linings

I'd like to share my daughter, Kaitlyn, with everyone. She is 12.5 years old and in 7th grade. Kaitlyn has 2 sisters, 1 brother, and a baby sibling on the way. She is the oldest. We don't know the gender of our baby, but Kaitlyn is hoping for a girl. She is getting excited for my induction date of March 19. Kaitlyn is a great sister, she loves to help out her siblings, but sometimes tends to be a little on the bossy side! She is the closest to her 8 year old sister, Lauren, and the two of them spend a lot of time doing things together. Joe and I are raising our family in Woodstock, IL. We are about 20 minutes south of the Wisconsin and Illinois border.

Kaitlyn is active in several activities. She has been in Girl Scouts since 1st grade; this year is her first year as a Cadette scout. She is actively working on earning her Silver Award, which is the highest award a Cadette can earn. Last year, she started taking swim lessons thru NISRA, which is a special education park district. She gets one-on-one assistance during her lessons. When she first started, she was afraid to get her face wet. In just one year's time, she has mastered going under, front float, and front glide. She's been working now on her back float and front crawl arm movements. This past fall, she got to



fulfill her dream of being a cheerleader. Our town had a new competitive, travel football league along with a new cheerleading program. It is a no-cut program, for kids thru age 13. She did really well learning her cheers and performing! Next fall, she will cheer again and is looking forward to it. Thru school, she has been in chorus since 5th grade. Kaitlyn loves to sing! Her other school activity is student council. Last year she was a representative and this year she is the fundraising co-chairperson. Next year she hopes to be the dance committee chairperson.

As I mentioned, Kaitlyn is in 7th grade. She enjoys middle school. Her school is from 6th-8th grade. It is hard to believe she's half way thru middle school. It seems like just yesterday I was so worried about her making the transition and if middle school would be too overwhelming for her. It has proven not to be! She does like school and her friends and activities there. Kaitlyn is mainstreamed, with the exception of reading and language arts. She is in a resource class for those two classes, and they are back to back taught by the same teacher.

Kaitlyn reads about 2 grade levels behind, and she has a hard time comprehending what she's read. Some of her IEP requirements are large print textbooks, with an extra set at home so that she doesn't have to bring them back and forth to school, a CCTV, another device which has a camera she points at things (like the board) and then it transmits the image to a flat screen/panel TV for her to see at her desk, enlarged assignments on large size paper, extra time for tests, reduced assignments (but most of the time she ends up doing the entire assignment), and two desk spaces so that she can sit at one and her textbook and materials at the other. She is assigned a one-on-one aide who is with her most of the day. That aide was assigned to her at the beginning of 6th grade and made a dramatic difference with how well she transitioned. Kaitlyn also

receives services from her vision itinerant, who is also her mobility teacher. She has an adaptive PE teacher who helps to modify things for her in PE so that Kaitlyn can safely participate.

Kaitlyn works hard for her grades, sometimes she does struggle. I'd like to brag on her behalf and let you know that last year she made high honor roll for 1st quarter and then was on honor roll for the rest of the year! This year, she is following the same pattern. It is wonderful to see her hard work paying off and knowing that she really can do well in school!

Lastly, I'd like to talk a little bit about her health. Her vision is 20/400 and cannot be corrected with glasses. She does have nystagmus, aniridia, bi-lateral cataracts present since birth, and she developed glaucoma about four years ago. She did have Wilm's tumor, which was discovered on March 17, 1999. We are rapidly approaching the 10 year anniversary of the rollercoaster that goes along with a Wilms diagnosis. This year when we celebrate her becoming a teenager, we will also be celebrating ten years of her being cancer free! She did end up having a complete left nephrectomy, followed by chemo of vincristine and dactinomycin. We still follow up with ultrasounds every six months. She also gets ultrasounds of her ovaries, and there is no sign of them being abnormal. Kaitlyn has hallux valgus on both feet, which makes it hard to find her shoes that fit. So far, she doesn't complain of any pain. This spring, we have to take her to a pediatric rheumatologist to get a diagnosis on her hands. She may have rheumatoid arthritis. Both ring fingers are showing characteristics of it and she can't straighten them. Another journey she is about to embark on is braces. She goes to the orthodontist at the beginning of March to get them put on. Right now she is looking forward to them, saying how cool they are. We'll see how long that attitude lasts once the pain sets in.

Last summer, Kaitlyn was sponsored by our city's Lion's Club to attend Camp Lions, a camp for visually and hearing impaired children. It was an overnight camp; she was there from a Sunday thru Saturday. I was so worried she'd be homesick being away that long, but I was wrong. She loved it! The campers got to do crafts, swim, have campfires, horseback ride, etc. It was such a nice feeling to send her off to a camp that was specifically designed for children with needs like hers. I just got a phone call that they want to sponsor her again this summer!

Thank you for taking the time to read about Kaitlyn. I've enjoyed sharing her with you!

Karen Wietermann

A Perfect World

Nine and one half years ago I had a fantastic pregnancy, my second. Two weeks over due and along came my beautiful daughter, Grace. Uneventful labor and delivery healthy baby girl. She wouldn't open her eyes. My midwife encouraged me to see someone and so we did. In fact we saw two doctors within the first few days of her life and all was "normal". Grace's eyes were swollen shut at birth so we didn't think anything of the fact that she did not open her eyes and when she finally did three days later, she wouldn't keep them open for very long. I attributed it to the fact that she was head down and flexed for so long that her frontal bones were swollen. Me being a chiropractor, I was checking and adjusting my child and doing cranial work on her. She was ok. That's what I kept telling myself, when in all reality my instincts were telling me different. I cried for 7 weeks everyday in the shower so no one could hear me begging God that there was nothing wrong with my perfect baby.

The day came when my perfect world came to a crashing halt. I was at my chiropractor's office and we were working on Grace when

he said "Jen she is not looking at me, you need to take her in to see someone". Those words hurt but also made me quit denying. I knew, oh God how I knew. In his office I pulled out the phone book and made an appointment with a pediatric ophthalmologist. No referral no searching no nothing. Just a cold phone call and by the grace of God I had picked the most amazing well respected doctor in southeast Michigan, Dr. Baker. My appointment with Dr. Baker was in 2 weeks. We started preparing ourselves for a blind child. I was ok with that as long as she was healthy in every other way. We even joked about having a "real" dog finally.

My once again perfect world came tumbling down the day we met Dr. Baker. He said she has aniridia. My medical background clued me into the fact she was born without irises. She has cataracts, and nystagmus. Is there an end to what he was saying? I mean how much can one absorb as the tears started flowing freely. Ok get a grip she has some eye problems I mean what the heck I was already prepared for a blind child. Now the real kicker came when he started mentioning all the other complications such as Wilms tumor, genital urinary problem, and mental retardation. Did I hear him correctly? My daughter could have cancer and be mentally impaired? Walls were closing in. WAGR syndrome. Never heard of it in all my studies, and certainly had never been exposed to someone with it. I left there feeling worse than ever, and was told we needed a full body CT scan. Once at home with the power of the internet, we researched WAGR and everything about all the issues surrounding it. Our wonderful Dr. Baker had the kindness in his heart to call us that night at home to see how we were doing and if we had any questions. Holy @#\$% we had questions.

A CT scan later which was another experience for the inexperienced, and getting involved with the VI support in our area and we were back to our perfect life. Routine ultrasounds, eye patches, home visits for VI support, eye exams

and we had this beautiful blossoming daughter. Ahh, life was great again! We had gotten on the mailing for the WAGR group when we initially were told about the possibilities. However, at 5 months of age we saw the geneticist and were told that the likelihood that Grace would develop Wilms tumor was less than 1%. Grace was not like those kids I read stories about in the newsletters. Those stories made me sob everytime I read one. Grace had aniridia with some complications. I was living in my perfect world or denial. We got involved with an aniridia group and went to a conference in Louisville, Kentucky. We met other families with kids and adults living with aniridia. We had high hopes. There was mild to severe cases, but they lived "normal" lives. They attended school, college, had jobs, were married. I was happy!

Routine ultrasound in May was one month later. We had just had one in December, and all was clear. My perfect world just ended. I was told Grace had a tumor in her kidney. I told the doctor he was wrong. My baby was fine, she is only 20 months old, and what 20 month old gets cancer? Where the hell is the sense in that? "Look at her she is happy and healthy" is what I told the radiologist. I could not accept those words. MRI later that day confirmed Grace had Wilms tumor. Just five months ago her ultrasound was clear, and within that time this tumor had taken up one-third of her kidney right in the center. Tailspin is putting it mildly as the next two weeks were a blur of meeting oncologist, surgeon, dr. after dr. etc... Grace had her right kidney removed two weeks after

Forward newsletter
submissions, stories,
pictures or ideas to:

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diagnosis and then followed with chemo.

Grace's second birthday will be the most memorable, as I was so grateful she made it to her second birthday and it happens to fall on the other most memorable time in our lives. 9/11. Not sure what to make of it, but hard to mourn for others when I'm celebrating literally my daughter's life. Whew! Two years of trials and tribulations. My daughter has aniridia and is a cancer survivor. Never would I imagine she was a WAGR child. She still wasn't like those stories I read and then decided they were too sad for me, and quit reading them. We lost touch with the WAGR group and focused on Grace's VI issues. (Denial?) Hmmmm.....

Grace started preschool in the VI class and remained there for 3 years. She developed well and loved school and was ready for kindergarten. All routine ultrasounds came back negative and my perfect world was back! Grace was being mainstreamed with VI support, mobility support, speech and language support, but she was going into a "normal" class. My daughter was not WAGR! (Denial?)

After first grade and a bazillion IEP's we saw Grace was really struggling and was not comprehending like the kids her age. It's because she's visually impaired right? Ha Ha. My reality finally came to the forefront of my brain when I had to accept that my daughter was different. She wasn't just visually impaired. She has WAGR. Oh my gosh, I finally said it! My daughter is like the stories I read. I just didn't know it because I denied, and refused to see it. No one knows what WAGR is. Even at her school I have to educate everyone involved in helping Grace. I am happy to report that Grace is in a center's program at her school and doing fabulous! She loves school and everything she gets out of it. We met up with the WAGR group last year at WAGR weekend and I felt at home. My daughter fit in with all of the kids we met. She is like them, and you know what she is perfect just the same as they are. I am

blessed that God gave me Grace and have always said that she would teach me more than I will ever teach her. I found the WAGR network again at the time I needed because I was ready to accept and not deny. So thank you to all the families we met at our very first WAGR weekend and all your support.

Grace is an amazing young lady who loves music, takes dance classes, is in brownies, swims, and sings in choir. She annoys her two brothers to no end but can warm your heart just upon meeting her. She is healthy and happy and I have my perfect world!

Jennifer Westcott

How do I explain our special girl? Sarah Hanson was born into a Military family in San Antonio, Texas three days before Christmas in 1987. She has an older brother by two years and a sister four years older. Sarah was always a happy little girl growing up, except for the occasional temper tantrum. She has 11p- chromosome deletion with glaucoma and aniridia and mild MR. She has had cataracts removed from both her eyes; the first at four months and the second just a couple of years ago. Oh, and she has the most gorgeous red hair!

Being a military brat, Sarah has moved with her family seven times. She has lived in Florida, Virginia, Alabama, New Jersey, California, Germany and currently back in San Antonio. In Virginia we discovered Challenger Baseball and Sarah got hooked right away. She played on challenger leagues every year from



five years old to 18 years. Her dad has even had to create leagues at some military bases in order for her to play. If you have never seen this league, I highly recommend you check it out, it is amazing!

When Sarah was born there was no name for her condition such as WAGR, so my husband and I struggled to find answers on our own. Luckily the military has programs and wonderful health care that have provided for her. It is an interesting story of my discovery of WAGR and the group. About four years ago, I took Sarah to a routine doctor appointment at our base clinic. Sarah was still in high school and was learning how to use a cane. We spotted another mother with a daughter about Sarah's age also using a cane leaving the clinic. We spoke hellos and Sarah and I went in for her appointment. A couple of minutes later the lady came back into the clinic and introduced herself and her daughter and gave me her card. This nice lady said if I ever wanted someone to talk to feel free to call. I put the card in my purse and went in to the appointment with Sarah. After the appointment was over, I noticed the doctor had written WAGR syndrome on her form. This was the first I had ever heard of WAGR and had always been told she had 11p- and aniridia. When I got home that afternoon, I looked at the card in my purse and it said Kelly Trout, president WAGR syndrome association. You could have knocked me over with a feather!! Anyway, it has turned into a life changing event for all of us to finally have someone to call with questions, someone who actually knows what we are going through, as I am sure you all know.

So, over the 21 years we have struggled with the medical issues of WAGR and also anxiety, some obsessive behavior, temper tantrums, depression, weight problems, nail biting, painful periods etc. Medication has been a great help in these areas and Sarah has become a more self-confident out-going person most recently. She dreams big and never lets her handicap dampen her drive to be normal and to do the things that her older siblings have done. She

dreams of owning a red van one day. She will spend hours doing artwork in her room; projects like coloring little squares into amazing designs. Her brother and sister have some of her work framed on their walls.

We have struggled with Sarah's weight problems since she was about ten years old. At a routine appointment, about four years ago, the doctor told me that her body mass index (BMI) was over 30. It was a wake up call for me. This was borderline obesity and I knew I had to do something. Her weight problems were solved by lots of walking and also joining Weight Watchers. She lost 32 pounds total and is now at a healthy BMI. It was a very difficult first year, but once the weight came off it was much easier to maintain and control the food cravings. She eats much healthier now and we did it as a family which I think was the defining factor for success.

She completed 12 years of public school, special education and many, many IEPs, along with several years of job training in the school system, all with great success. She is currently working part time at the local department store on base. She is well liked by her coworkers and is very happy to have the job and responsibilities.

Sarah has a puppy who is a great friend and companion. Her name is Libby and Sarah walks and plays with her everyday, not to mention feeding her too.

Sarah goes to the gym with her mom three to four times a week and ran her first 5K race this past January. Her sister even spotlighted her and WAGR on her blog, <http://honeyandquilts.blogspot.com>, (under Blog Archive January). We plan to run another race in May, this time with her dog and the race proceeds will go to the local animal shelter.

One thing is for sure: Sarah has taught me to be a better person and to really put things into perspective. We have had many difficult times and I am sure there will be more to come, but we have certainly had wonderful fun times with this special girl with the funny sense of humor. We hope to meet everyone at a WAGR weekend one day.

Catherine Hanson, daughter Sarah, WAGR, age 21.

Group Homes

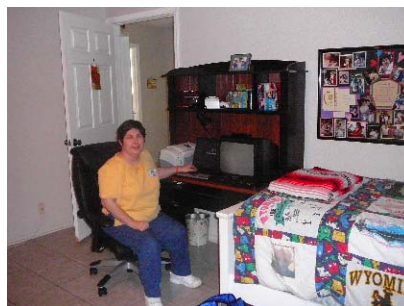
By Carol Larson

When talking about Group Homes one needs to remember the Four Facts.

One, a group home is not a State Institution, the kind of place where you have no freedom. In a group home, you have tons of freedom. In a group home you can talk to the staff about what you need or what you want and they will listen and take care of it for you.

Two, in a group home it's a partnership between the staff and the clients (the person with a disability). It's a partnership in which the staff and the clients talk about stuff that they need or want to do. Communication is definitely the key.

Three, the experience is what you make it. For example I have two friends who lived in a group home and didn't have a good experience. One of them, Susan, thought she was able to live on her own,



and she didn't have a very good experience. The other, Joel, broke all the rules of the group home. For breaking the rules he got kicked out two times. If you as the client are able to live on your own, then you may be too independent for a group home. In my case, I think of it as a stepping stone toward living on my own, so I am having a good experience so far.

This leads us to fact four, behavior issues. Sometimes there are staff members who think that when a client is having an attitude problem they are having behavior issues. That may not always be the case. The client just might be upset about something, or just needs to take a chill pill. Did I mention communication is the key here?

"C3" Day another HUGE Success in Riverview, MI

One Friday every month is designated Casual Clothes for a Cause Day (C3 Day) in the Riverview Community School District. Staff members pay at least \$5 to "dress down" or wear jeans on this day. Money raised is donated to pre-selected charities.

The generous staff in the Riverview Community School District raised over \$750 dollars for the IWSA in October and \$2,400 to date.

**Way to go "Pirates"
Thank you again for your
generosity & support!**

Tax Deductible Donation

The IWSA is a 501(C)(3) charitable organization.

We rely on contributions from private and public sponsors.

Please consider donating today.

IWSA Officers & Board Members:

President - Catherine Luis
Vice President - Tammie Hefty
Secretary - Julie Dell
Treasurer - Jeffrey Hefty

Board Chairperson - Annie Prusakiewicz
Member - Tom Cox
Member - Shari Krantz

NIH UPDATE



Dear WAGR Families,

Greetings from the National Institutes of Health in Bethesda, Maryland! The WAGR syndrome studies are going very well, and we would like to update you about what is currently happening.

First, we would like to introduce ourselves to anyone who is new to our research. Joan Han, MD, principal investigator on the WAGR syndrome study, is a board-certified pediatric endocrinologist. Dr. Han attended college and medical school at Harvard University. She trained in pediatrics at Boston Children's Hospital and in pediatric endocrinology at Nemours Children's Clinic in Jacksonville, Florida and at the NIH. Kristen Danley, BS, is a research assistant working with Dr. Han as coordinator of the WAGR syndrome study. She graduated from Bucknell University with a biology degree.

The current WAGR Syndrome research we are conducting at the National Institutes of Health consists of Phase I and Phase II studies. Both are recruiting participants and are designed to study how genotypes (the specific genes deleted) are associated with phenotypes (the clinical symptoms present) for each individual with WAGR. For both phases, we request that you complete the various consent forms and

informational sheets for your child. We ask that your child's doctors send us copies of any past medical records (we pay for all shipping costs). We also request a comprehensive medical history, which involves completing a form by writing, typing, or speaking on the phone with us.

Phase I is an outpatient study, consisting of a fasting blood draw, collection of medical records and a complete medical history for the participant. One or both parents may choose to participate by having their blood drawn as well, but this is entirely optional. WAGR individuals of any age may enroll in Phase I.

The Phase II study, launched in November 2008, is entitled "WAGR Syndrome and Other 11p Contiguous Gene Deletions: Clinical Characterization and Correlation with Genotype." Children age six and older and adults with WAGR syndrome or a diagnosed 11p deletion can participate. The study involves a one-week visit (arriving Sunday evening and leaving Saturday) to the NIH in Bethesda, MD during which participants stay on the NIH campus. We are also recruiting children and adults with isolated aniridia to complete a similar visit for the study.

During the week, participants will receive a comprehensive evaluation of a variety of symptoms associated with WAGR syndrome. The evaluation includes:

- testing of how much energy they use
- testing of how they process glucose
- evaluation of hearing and vision
- evaluation of behavior and motor skills
- kidney screening
- head and body imaging
- dental and facial measurements
- testing of sensation thresholds
- measurements of body composition (fat mass and non-fat mass)

If you would like to review the full list of tests, please contact us and

we will send it to you as well as discuss it in detail with you over the phone.

After receiving feedback from families who have completed Phase II at the NIH, we have learned that completing all the scheduled tests can be difficult and we wanted to reassure families that lightening the schedule by eliminating some tests, prolonging their stay at the NIH to spread out the tests, or splitting the testing among multiple visits are all options they can choose.

Since the Phase II study began, we have had several families complete the week-long visit and have approximately twenty families enrolled. We are currently recruiting for visits during August and Fall 2009 as well as Winter 2009/10. We are learning so much from each visit, and we are very excited to meet each participating family.

If you are interested in participating in the Phase I and/or Phase II studies or have other questions about NIH research, please feel free to contact Kristen or Joan.

Kristen Danley (301) 402-6762
danleykr@mail.nih.gov
Dr. Joan Han (301) 435-7820
hanjo@mail.nih.gov

Go to NIH, participate in the research, and you'll be very happy that you chose to do so. It just may be the greatest decision you could make for your child with WAGR syndrome. Your hosts, Dr. Joan Han and Kristen Danley, will make certain that you are heard, so bring your questions. They will spend the entire week finding answers for you.

Bryan and I set off for NIH on Sunday, November 30th, after a hectic Thanksgiving Holiday. I wasn't well prepared, and didn't know what to expect, but I had a lot of questions. I felt that this was a great opportunity to get some answers. I was also glad that I had my GPS, because the Sunday traffic was heavy in and around Bethesda. We arrived at 3:00 p.m., got out of the car, showed our IDs, and received our badges. The security was very impressive.

They checked everything in my car and then continued to direct us to The Children's Inn.

Due to the fact that it was Sunday, the parking was quite easy, however, later on in the week the parking can become somewhat more difficult. I never used my car while at NIH so parking was not an issue for me. Once we were at the Inn, we were welcomed and shown to our room, and then their very friendly staff gave us a tour. Bryan and I were given our own keys and he was then shown to his own personal mailbox, where he received something special every day! At the end of each day he asked if it was time to "check my mail!" What a great idea! We had a room right next to the "Tree-house Playroom", which soon became Bryan's favorite place to be. There was a pretend kitchen and he made me "tea" every night before bedtime. The room we stayed in was absolutely immaculate. I was very pleased with the TV, which allowed me to catch up on the news and relax without having to go out into a "community room." Bryan has medicine he needs to take, and they provided us with labels and a key to the special refrigerator where that could be kept safe. This was also needed later when we had to do our 48-hour urine collection.

The inn itself is incredible! It's very cozy, the fireplace is inviting, and the large screen TVs are in cozy nooks where you could snuggle up on a couch and watch a movie with your family if you chose to do so. The kitchens were large and the pantries had some staple items. You need to make or provide your own meals. The way our schedule worked, I found it easier to eat at the hospital. We never prepared a meal. One night, a volunteer group did come in and cooked a meal for everyone. The other nights we ate at the hospital and just had a snack at the Inn. There is a vending machine if you want candy or chips. I also brought my cooler stocked with Bryan's favorite drinks and snacks.

We were tired every night after the

long days of tests. The Inn is located just across the street but below the NIH. We took the shuttle once or twice, but I found it easier and somewhat more convenient to walk. Thankfully, I had brought Bryan's wheelchair/stroller, because I would not have been able to walk with him without it. It was cold and it would have taken Bryan much too long to walk along the pathways. If you go when the weather is nice, I would recommend that you walk.

Shortly after we arrived, we went to the hospital to register. You must do this on Sunday because you have to be ready for "work" early Monday morning. We then took a peek at the pediatric ward where Bryan would be spending most of time, including two nights of our stay. I stayed with him at his bedside on those occasions and it was very comfortable. The pediatric ward has a play/craft room that Bryan absolutely loved! The



hospital itself has a cafeteria, but I only went there once. I personally preferred the gourmet coffee shop in the lobby, which opened at 7:00 a.m. Monday thru Friday. They served sandwiches, salads and an array of tempting snacks.

Bryan was amazing. I have to admit that I had my doubts about both their expectations of him and the amount of cooperation that would be needed to participate in many of the tests. To my surprise, however, he did everything that they asked! Some of the tests were too difficult because of his age and limited understanding, but he always tried. The taste test was absolutely hilarious. Bryan ate everything and said everything was "great", regardless of what it

may have tasted like. He couldn't do the smell test, however, because it was a "scratch & sniff" type procedure. Kristen scratched a paper and held it up for Bryan to smell. He kept responding, "paper!" I still don't know if he can smell anything.

If you have any specific areas of concern, talk to Dr. Han before you go to NIH. I wanted an MRI of Bryan's brain and a thorough scan of his testicles. She accommodated me and even sent the reports directly to Bryan's urologist at Johns Hopkins.

By the time Saturday rolled around, we were ready to return home. I was disappointed because the coffee shop wasn't open on Saturday, but we plugged along and managed to get through the day despite my lack of caffeine. Bryan reluctantly turned in his mail key after emptying his mailbox, and I came home with a stack of reports, with several still pending. Initially, I was confused with all of the information, but when I got home and began to really delve into it, I was in awe of what we had accomplished. We found out that Bryan probably does NOT have Autism, but rather an Auditory Processing Disorder (APD). I had never heard of that condition before, and once it was explained to me I realized that so much of it fit! Much of Bryan's behaviors and mannerisms now finally made sense. I was surprisingly relieved.

I carry that stack of reports to every doctor Bryan ever needs to see. I cannot emphasize how valuable all of this information has been and continues to be for all of us. I am forever grateful for the time, patience, and especially the wonderful opportunity given to us by Dr. Han and everyone at the NIH. We look forward to seeing everyone at WAGR Weekend in July.

Patty Desmond

We joined up with the WAGR group over nine years ago. It was wonderful to have information that, despite our searching, had never been available to us before. Immediately we were welcomed

into a family of friends. Never did I imagine this group would have leaders that could give such time and persistence towards getting the medical world to realize the urgency of helping our WAGR children. I can never thank them enough for their dedication! That is why there was no question as to whether we would take part in the NIH study. I believe that this study is the answer to many of our prayers. It has already been a blessing to us.

Wes is thirteen and although he had not had Wilms', he has AGR, as well as no lenses (infantile cataracts removed by four months old). I won't go into his story as I have written a couple of stories already. What I would like to say is about our experience at NIH. It was intense in that there was a schedule of tests that filled each day from early morning until 5 PM or so. Dr. Han and Kristen are wonderful as well as the nurses and other people that worked with Wes. They explain everything and answer any and all questions. When we got tired of the "heading to another test" or "sitting for hours waiting for him to come out of one" we could not complain. We were in such awe over how Wes handled it all! He was unbelievably patient, good natured and agreeable through all of the constant tests, questioning and prodding. It was as if we had all been given extra patience to see it through.

Already we have been given a wealth of information. Some things are very concrete such as protein in his urine (and we had been testing!) to why he can't seem to close his mouth when he eats. Some findings had specific recommendations and others gave us insight



and understanding for certain conditions. It was a relief to know we could do more for Wes.

I have listened and watched as families have and are going through such unbearable situations. Always knowing that life has been pretty easy for us by comparison but knowing that at any time we could be facing harder hurdles. I always knew of the possibility of FSGS and that Wes could get glaucoma and go blind at some point but those stay as possibilities not probabilities. I am very hopeful, that through the research study, Wes's life will be so much better as will all the WAGR kids. How very blessed we have been to see this happen in our children's lives! It is called a research study but the truth is we get immediate help for our children just by getting the results of all their tests-tests that might not have been done otherwise. The results of the research will be the bonus that will be there for all children, present and future.

Grandma Bev Schmezer

WAGR Syndrome is a very rare disorder. Physicians are generally not familiar with this condition, or with the features and complications of it. People with WAGR syndrome benefit greatly when family caregivers, medical professionals, teachers and therapists learn as much as possible about the disorder, and become active partners in their care. "WAGR" is an acronym. The letters stand for the most common features of this disorder.

W-Wilms' tumor

A-Aniridia

G-Genital and/or urinary tract abnormalities

R-mental retardation/developmental disabilities

People with WAGR Syndrome have many things in common, but they are also individuals. It is important to remember that a given individual with WAGR syndrome may or may not have or develop the same conditions.

WAGR Weekend 2009 Gaithersburg, Maryland July 10-12, 2009

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Tentative Itinerary:

Friday Night
Meet & Greet
Mom's night out

Saturday - NIH/NICHD
Research Update from Dr.
Joan Han & Kristen Danley
Lunch - Catered
Dinner - Catered pizza party,
Dad's night out
Free time - Playground, Water
park, Miniature Golf, Skate
Park

**Sunday - Morning tour of the
NIH**

Details and RSVP at the web-
site

<http://www.wagr.org/wagrweekends.html>

WAGR Weekend Family Financial
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able. For more information
please contact Tammie Hefty at
tammiejo@mhtc.net or
(608) 347-6356

Nephrology 101, What every parent MUST know

Dr. Jeffrey Kopp is a Staff Clinician at the National Institutes of Diabetes and Digestive and Kidney Diseases, National Institutes of Health, Bethesda, MD. His research interest lies in the syndrome of focal segmental glomerulosclerosis (FSGS). Each kidney has about 1 million glomeruli each of which is a cluster of blood capillaries; these glomeruli filter small molecules including toxins from the blood. FSGS is a set of diseases in which scarring (sclerosis) affects the glomeruli and does so in a manner that at the onset of disease affects parts (segments) of some glomeruli (focal distribution) while other glomeruli remain normal. FSGS has many causes, including genetic, viral (most notably HIV), and obesity, but most cases are at present labeled idiopathic, meaning the cause is unknown.

1. Do all individuals with WAGR syndrome need to be screened for kidney disease?

Yes. We recommend that all individuals with WAGR syndrome have regular screening to detect kidney disease.

2. My child never had Wilms tumor -- does he/she need to be screened for kidney disease?

Yes. Even if your child never had Wilms tumor, your child is still at risk for developing kidney problems.

3. What percent of individuals with WAGR syndrome develop kidney disease?

We do not know the exact incidence as this is something we are still in the process of researching. A recently published case series of 54 patients with WAGR syndrome reported that 60% of individuals older than age 12 years had decreased kidney function or kidney failure (Pediatrics 2005; 116:984-988).

4. What is the cause of kidney disease in WAGR syndrome?

When patients with WAGR develop kidney disease, they most commonly

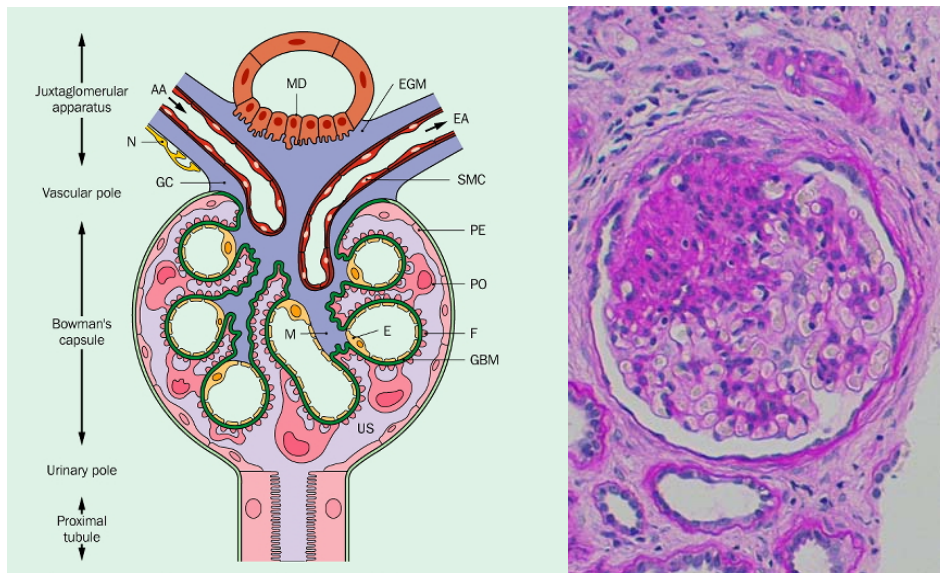


Figure. The left panel shows a schematic of a glomerulus. Blood enters the glomerulus via the afferent arteriole (AA), flows through the open capillary loops in the center of the glomerulus, and flows out via the efferent arteriole (EA). Fluid, but not blood cells or proteins, passes through the glomerular sieve and into the proximal tubule, much like water passing through a colander as vegetables are washed in the sink. The fluid, after passing through the tubule, will eventually become urine. The podocytes (PO) are cells that participate in maintaining the glomerular barrier and that are damaged in focal segmental glomerulosclerosis. (Used with permission, from Johnson and Feehally, Comprehensive Clinical Nephrology, Second Edition, Mosby 2000). The right panel shows a glomerulus from a patient with focal segmental glomerulosclerosis. There is an area of scarring (sclerosis), the red portion at the upper left glomerular segment, while the remainder of the glomerulus shows open capillary loops where blood circulates. Photograph courtesy of Dr. James Balow, NIDDK, NIH.

have FSGS. As noted above FSGS is a syndrome and WAGR patients may have several factors that promote this kind of kidney scarring.

- First, the WT1 gene is critical for normal function of the podocyte (meaning "foot cell"), which is a glomerular cell type. WAGR patients have only one normal copy of the WT1 gene, located on the other copy of chromosome 11, and in many cases this appears insufficient for the podocyte to function normally.

- Second, WAGR patients are prone to obesity, which puts a metabolic stress on the kidneys by compelling the kidneys to over-function to cope with the increased body size. An analogy would be that a car engine that is driven at high speed, or high rpms, for a long period of time will last less long than an engine that is driven moderately.

- Third, WAGR patients who have had a Wilms tumor will have just one remaining kidney, and that kidney then is called upon to do the work of two kidneys - again, this represents stress to the kidney. By itself it is unlikely to be problem (remember that people can donate one kidney safely to another person) but it can make other kidney diseases worse.

- Fourth, if hypertension develops, this may cause FSGS or may accelerate the progressive scarring of FSGS that was initiated by another factor.

- Fifth, WAGR patients who have had radiation that extended to the normal kidney may have radiation damage.

WAGR patients who develop diabetes are at risk to develop diabetic kidney disease, which is a different disease from FSGS, but has in common glomerular scarring (glomerulosclerosis). The

best way to prevent diabetic kidney disease is control of blood glucose to normal levels.

5. At what age should the testing begin for WAGR kidney disease?

We recommend that testing for kidney problems begin at birth. During infancy and early childhood, individuals with WAGR syndrome are at high risk for developing Wilms tumor. During late childhood and early adolescence, the risk for kidney disease increases. Therefore, lifelong monitoring is recommended.

6. What tests can be done to detect early kidney disease?

For Wilms tumor screening, children should undergo every-three-month kidney ultrasound and urinalysis from birth to age 5 years, and then the same tests every 6 months for the remainder of childhood.

For FSGS screening, children should have annual measurement of blood pressure and measurement of urine protein. We are not sure at what age this annual testing should begin, whether it should be age 5 or 10 years or some other age.

There are three ways to measure urine protein:

- Dipstick test, done as part of a routine urinalysis - this can be useful but is not very sensitive (in other words, it can miss small amounts of protein in the urine that can still be a sign of kidney disease).
- Measurement of the urine protein/creatinine ratio, which is more sensitive and can detect early kidney disease.
- Measurement of the urine albumin/creatinine ratio (albumin is plasma protein). This is a standard test to detect early diabetic kidney disease, but its role in other kidney disease is less certain. The NIH studies will examine whether detection of microalbuminuria (small amounts of albumin) is a useful test to detect early kidney disease.

7. What are the early signs and symptoms of kidney disease?

Early kidney disease typically has no symptoms, and thus regular laboratory testing is important.

8. How is WAGR kidney disease treated?

There are three therapies that slow progression of other scarring kidney diseases, and although they have not been studied specifically in WAGR patients, these therapies are likely to be effective.

- First, control of blood pressure to the age-appropriate normal level.
- Second, use particular blood pressure medicines, termed ACE inhibitors and ARBs (angiotension receptor blockers). These medications have three beneficial effects: lower systemic blood pressure, lower blood pressure within the glomerulus, and prevent or slow the process of glomerular scarring (glomerular sclerosis) by turning of signaling molecules that regulate production of collagen (the major scar protein).
- Third, limit dietary sodium (salt) intake, as this lowers blood pressure and also increases the effectiveness of ACE inhibitors and ARBs to reduce kidney scarring.

9. Is the recommended treatment different for individuals with WAGR syndrome compared to other people with kidney failure? If so, why?

Treatment of progressive disease due to WAGR is similar treatment of progressive kidney disease of other causes. When the kidneys fail, the options are chronic dialysis (either hemodialysis or peritoneal dialysis) or kidney transplant - and kidney transplant is the preferred treatment.

10. My child is enrolled in Phase I (outpatient portion) of the NIH WAGR Study, if he/she is diagnosed with hypertension, proteinuria or both can we contact you for recommendations?

Yes. Please contact the WAGR Syndrome Research Study Coordinator (301-402-6762, wagr_study@mail.nih.gov) with

any questions or concerns. We request that you obtain a copy of the blood pressure readings and lab results from your child's doctor so that we can better assist you.

11. My child is enrolled in Phase II (inpatient portion) of the NIH WAGR Study, if he/she is diagnosed with hypertension, proteinuria or both can we contact you for recommendations?

Yes. Please contact the WAGR syndrome Research Study Coordinator (301-402-6762, wagr_study@mail.nih.gov) with any questions or concerns. We request that you obtain a copy of the blood pressure readings and lab results from your child's doctor so that we can better assist you.



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**Education Questions?
Families & Professionals
please Contact:**

**Rhonda Sena, M.Ed
Educational Consultant**

CasaSena5@yahoo.com

Behavior Disorders: Coping with explosive or aggressive behaviors

Kelly Trout, RN, BSN
Rhonda Sena, M.Ed

Some individuals with WAGR syndrome have behavior disorders, such as autism, autism spectrum disorder (ASD), attention deficit hyperactivity disorder (ADD/ADHD), anxiety disorder, and/or obsessive-compulsive disorder (OCD). These conditions can cause big challenges for a person and his family. Some of the most difficult of these challenges are tantrums, “rages,” meltdowns, and destructive or aggressive behavior. In a series of articles, we’d like to offer some suggestions for dealing with these behaviors.

The All-important First Step: Diagnosis

It’s hard to fix a problem if you don’t really know what’s causing it. So it’s critical to make sure you have a diagnosis first. Don’t be tempted to skip this step, hoping that your child’s explosive outbursts will stop, lessen, or will just go away on their own. Like most conditions, these behaviors respond best when they are recognized early and treated appropriately. It’s also common for parents to be told that their child’s explosive or aggressive behaviors are simply “the way he is,” and that little or nothing can be done. The truth is, a wide variety of positive, potentially successful options for treatment exist.

It is very hard to be told that your child has a behavior disorder, but if you suspect there’s a problem, getting a diagnosis can open the door to services, solutions, and safety for your child and for your family.

**Health Questions?
Families & Physicians please
Contact:
Kelly Trout RN, BSN
IWSA Health Consultant &
Liaison to our Medical
Advisory Board.
KellyTrout@sbcglobal.net**

Making sure the diagnosis is accurate is also critical. On the surface, challenging behaviors may look similar in children with ADD/ADHD, autism, anxiety, and obsessive-compulsive disorder. But effective treatment can be quite different for each. In fact, sometimes what works for one disorder can make another disorder worse.

Keep in mind that accurate diagnosis is often a process of elimination. This can be very frustrating, but it’s more than worth the time and effort. You can facilitate the process by educating yourself about the signs and symptoms of behavioral and psychiatric disorders. Certain other conditions associated with WAGR syndrome, such as Sensory Integration Disorder and Central Auditory Processing Disorder (CAPD) can also result in behavior problems. The more you know about these conditions, the more you can help your child’s doctor or therapist pinpoint the problem. For more information, check out the Behavior page at <http://www.wagr.org>

Diagnostic Evaluation

The best method of evaluating a child for behavioral or psychiatric disorders involves a “multidisciplinary team” approach. Such a team may include a psychologist, neurologist, neuropsychologist, psychiatrist, speech therapist or other specialists, as well as the parents and teachers or other caregivers. Some universities and regional or teaching medical centers offer team evaluations, and your pediatrician may be able to refer you to one of these. When this is not possible, specialists can be consulted separately, but a “team approach” can still be achieved by insuring that everyone communicates with each other.

Now What?

Once you’ve got a diagnosis, you can begin to look for the information, professionals, and programs that can help.

Behavioral therapy is available for some conditions, in some communities, and your child’s psychiatrist or psychologist may refer you to such a program. On the other hand, you may have to find this kind of help yourself.

Here again, learning about the disorder yourself can alert you to what type of therapy might help and where you might find it.

If your child is school age, his behavior disorder will very likely impact his education. A Functional Behavior Assessment (FBA) will help you and the IEP team develop interventions to address problem behaviors at school. Talk with the school and his teacher about including this assessment as part of his next IEP review or request it immediately if behavior is escalating. Behavior interventions that are specific to his diagnosis need to be included in his education plan.

Finally, many national, regional and local support groups sponsor behavior seminars and training specifically for parents. These seminars and training sessions can be tremendously practical and helpful. Disability organizations, autism support groups, and mental health groups are great resources. Call and ask what they may have available for parents in your area.

Remember The Truth

It’s incredibly difficult to deal with a child whose behavior is destructive, aggressive, embarrassing. Tantrums and rages can happen anywhere, anytime, and can make parents feel desperate, ashamed, and reluctant to leave the house or take the child out in public. Remember that you and your child are not alone. Lots of WAGR parents share your challenges, and know just what your life is like. Remember, too, that your child’s behavior disorder is a condition associated with WAGR syndrome, and not a reflection on you. It’s especially tough to remember these things when strangers (even family members) make rude comments or criticize your parenting abilities. Hold the truth close, gain strength from your fellow parents, and take pride in the progress that you and your child are making.

Upcoming articles:
**Patterns and Triggers,
Behavior Modification
Therapy, Medications**

The **IWSA** serves families all over the world in countries, including: Australia, Bavaria, Belgium, Brazil, Canada, Croatia, England, France, Germany, Greece, Ireland, Israel, Italy, Kosovo, New Zealand, Peru, Philippines, Poland, Portugal, Puerto Rico, Romania, Saudi Arabia, Scotland, South Africa, Switzerland, Tanzania, Turkey and The United States of America

You can support the IWSA in many ways. Financial donations both large and small are always welcome. The IWSA is a 501(c)(3) corporation, so all donations are tax deductible.

Spread the IWSA mission to your family and friends and ask for their support. It is a proven fact that the number one reason that people do not donate to a specific organization is because they were never asked. Friends and families are also more likely to donate to causes directly related to people they know than to ones they don't. More than 83% of all charitable gifts are still given by individuals.

Pay Pal is up and running on the wagr.org website. Pay Pal is a highly recognized way of collecting funds and is accepted in more than 190 countries. With this option, it will be easier to make a donation from inside and outside the United States.

Individuals who have Facebook accounts can also create awareness for the IWSA by searching for WAGR Syndrome – Deletion 11p. You can make donations to our cause via Network for Good (networkforgood.org). Simply search causes for WAGR Syndrome and ask your Facebook friends to do the same.

Other ways to help us fund the organization could include Gorilla



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Thank you for your continued interest and involvement with our organization. Together with your support we can continue to promote international knowledge and awareness of WAGR Syndrome and its complications and treatments, to stimulate research and to reach out to those affected by WAGR Syndrome in an effort to improve their lives.

Catherine Luis continuously updates the wagr.org website. So please check it periodically for updates and current information. 1800flowers.com has been added and allows you to purchase flowers with a percentage of the sale donated to the IWSA.

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