Reaching Out, The WAGR Network

Fewer than 1% of children diagnosed with Wilms tumor have Wilms tumor as part of the WAGR syndrome. Parents of children with WAGR have created a support group, called Reaching Out, The WAGR Network, and a web site **http://www.wagr.org**. The following is an interview with Catherine Luis, whose daughter, Irma, has WAGR syndrome. Catherine and several other parents created this important support group.

Catherine, you and Kelly Trout started a support group called "Reaching Out, The WAGR Network." What motivated the creation of the group and the web site?

The group actually began with a little newsletter that was started by June Kuntze in Minnesota who found a couple of other WAGR families. Another mom, Annie Prusakiewicz, in Michigan took over the newsletter and expanded it. Kelly started an email discussion group, and I set up the web site. At that time, we had all of seven families in the group! WAGR syndrome really is quite rare, so the web site really helped other families to find us. Currently we have 68 families in the group, in 11 different countries (the United States, Canada, Australia, England, France, Germany, Greece, New Zealand, the Philippines, South Africa and Croatia)! So really, a number of parents "started" this group. We have all suffered in some way from the lack of information about this disorder, so we are all very motivated to help each other to help our kids. One great thing is that in addition to parents, many other people are also accessing our web site; physicians, therapists and students are using it, too. We're continuing to work on it to keep it up-to-date and user friendly for anyone interested in information on WAGR syndrome.

Please let our readers know what WAGR syndrome is:

WAGR syndrome is a rare genetic disorder. "WAGR" is an acronym. It stands for Wilms tumor, Aniridia (having no iris, the colored part of the eye), Genital or urinary tract abnormalities (like undescended testicles), and mental Retardation. Children with WAGR syndrome usually have at least two of these conditions. WAGR syndrome is associated with a defect in a certain part of a chromosome technically identified as 11p13. No one knows what causes this genetic defect.

Most children with WAGR syndrome are diagnosed at birth, or shortly after, because the aniridia is usually obvious. But thorough genetic testing (including a special test called the FISH probe) is important, because the other signs may not be present or obvious. If a child has two of the characteristics, such as aniridia or genital abnormalities or developmental delay, then screening for the presence of a Wilms tumor is recommended.

What difficulties do children with WAGR syndrome face?

Well, most kids with WAGR syndrome have some degree of vision impairment, although they often have a lot of functional vision. Aniridia can be complicated by cataracts or glaucoma, and these can cause further vision loss, so it's important for them to be followed by a pediatric eye doctor. More than half of the children with WAGR syndrome will develop Wilms tumor, usually at an earlier age. This is another reason why close follow-up is important, because it allows for early diagnosis. Treatment for Wilms tumor is difficult not only for the child with WAGR syndrome, but for the entire family. The good news is that most of our children are long-term survivors.

Many kids with WAGR syndrome also have mental retardation or developmental delay. In some cases, there may be behavior disorders, such as Attention Deficit Disorder (ADD), Attention Deficit and Hyperactivity Disorder (ADHD), and Autism or the so-called Autism Spectrum disorders.

Do WAGR children need special education?

Special education is a big issue for our families. Early intervention services, which are designed to help infants and young children cope with vision impairment and developmental delay, can be extremely helpful. Appropriate special education throughout their school years can make a huge difference for these children.

Do these children develop kidney problems?

In the last couple of years, we have become aware of another risk for kids with WAGR syndrome. The National Wilms Tumor Study Group found that older children with WAGR syndrome may develop kidney failure. In our group, about 60% of those over age 12 have some degree of renal failure, and five of them have had kidney transplants.†

What resources do you provide to families with children diagnosed with WAGR?

The wagr.org web site includes a "Just Diagnosed" parent information packet designed to explain the

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disorder in detail. The web site also has lots of information about how to get good medical care, and access to government agencies and special education. We publish a quarterly newsletter that has medical news and profiles of families and kids. We have an email discussion group that is really lively; parents find it is a great place to talk about everything from coping with cancer to where to get good quality sunglasses for babies.

We also offer assistance to professionals. Physicians often contact us for information about genetic testing, and our web site has a checklist of Guidelines for Health Supervision of Children with WAGR Syndrome. We're also working on a booklet now, called, "WAGR Syndrome: A Guide for Parents and Professionals." We hope to complete it sometime in the coming year.

What do you think is the greatest asset of the network?

Families! Moms and dads, grandmas and aunts and friends, we are all inspired by our precious kids and their desire to overcome their tremendous challenges. We're driven to help them and to help each other. Without each family's willingness to share there would not be a network. "New" families (those who have a new baby, or those who've just found out about us) often say that connecting with others is what helps them most. Our other great strength is that there are so many of us now. By getting together we really can make a difference for our kids.

What more can be done to find out about WAGR? You have conducted some surveys of your families. If these were expanded to be more scientific, would this be valuable to your families?

WAGR syndrome is so rare that the primary information about it in the medical literature has been case studies (reports of a single case or two). When the families began to communicate with each other, we found that our children had many things in common that had never been reported. So we drew up a list of health questions for parents to complete. The results of this survey told us a lot about what kinds of conditions to look for as well as some important ways to help our children.

The survey taught us that gathering this information is not only valuable--it's vital. We are constantly looking for ways to support and expand the research that will help our kids.

Your fourth annual WAGR Weekend is scheduled for next June. Please tell us what activities are being planned.

Our WAGR Weekends are just fun, informal gatherings where we finally meet each other in person. It's wonderful to spend time with other families who know just what your life is like. And meeting each other's children is the greatest! Our kids with WAGR get to meet other kids who are "just like me," and their siblings get to see that they're not alone either. For many families, it's a rare opportunity to just relax. The 2004 WAGR Weekend will be June 25-27, in Gatlinburg, Tennessee.

Your daughter Irma is now in high school. How is she doing?

She's thriving! What I've learned is never to say never. She attends a vocational technical high school in New Jersey. This is a wonderful place where she continues honing her academic skills and is learning different job skills to prepare her for entering the workforce. She loves this school and has never been happier.

† **Reference:** Breslow NE, Norris R, et al: Characteristics and Outcomes of Children with the Wilms Tumor-Aniridia Syndrome: A Report from the National Wilms Tumor Study Group. J. Clin Oncol 21: 4579-4585, 2003.

Are There Any Internet Chat Rooms the NWTS Can Recommend?

We have been asked this question a number of times. We have also received recommendations from participants for rooms that they have found to be very helpful. Unfortunately our organization cannot make any recommendation, as this would be seen as an endorsement of the site and all of its contents. Some contributions to chat rooms can be misleading or incorrect, and sound advice about your health and medical care is important, so we cannot endorse a source of information that may steer you in the wrong direction.

We are not discouraging you from finding helpful and supportive chat rooms. Many people do find these to be important sources of information and advice. Any recommendations regarding medical treatment should be reviewed with your doctor first. If you are looking for reliable sources of information, please check the links posted on our web site **www.nwtsg.org**.