



Getting Good Medical Care For Your Child with WAGR Syndrome A Checklist For Parents

Children with WAGR syndrome may be born with physical, mental and medical challenges. WAGR syndrome is a very rare condition. Because it is so rare, few physicians are familiar with caring for an individual with WAGR syndrome. This purpose of this article is to provide parents with a checklist for insuring that their children with WAGR syndrome get the medical care they need to survive and to grow.

PLEASE NOTE: All children with WAGR syndrome are not the same. The recommendations below are not appropriate for every child with this disorder. Please discuss these recommendations with your child's physician, and together you can determine which tests and referrals are right for your child.

The article, "Health Supervision for Children with WAGR Syndrome" has been prepared to assist physicians who are caring for children with WAGR syndrome . You may want to print it out and give it to your child's doctors.

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

CHECKLIST FOR YOUR CHILD: BIRTH TO 1 MONTH: NEWBORNS

At the Pediatrician's Office:

- Ask the doctor to discuss the diagnosis of WAGR syndrome with you, and to explain the results of the genetic testing. Try to have someone with you at this visit, such as your spouse or a trusted friend or family member.
- Ask the doctor to review your child's physical examination with you. Have the doctor discuss these findings with both you and your spouse, if possible.
- Ask the doctor for copies of all lab test results, and copies of reports from any specialists. Keep these copies in a medical file for your child at home. In time, you will find this file to be very helpful.
- Ask lots of questions. You may want to write your questions down before the visit, and take them with you.

Talk with your Doctor About:

- A "Medical Home."**

Will your pediatrician help you to manage and coordinate the various medical specialists?

Wilms tumor.

When will the first ultrasound be done?

When and where will our 3-month ultrasounds be done?

When will we get the results?

Ask the doctor to teach you how to check your child's tummy in-between ultrasound exams.

Referrals you may be given: Newborns

-Genetics. It's important for a geneticist to confirm the diagnosis, to order further testing if necessary, and to follow your child's progress as he grows.

-Pediatric Ophthalmology. Children with Aniridia should be seen by an eye doctor who specializes in children, if possible.

-Urology (If your child has external genital abnormalities, like hypospadias or undescended testicles)

CHECKLIST FOR YOUR CHILD: 1 MONTH TO 1 YEAR: INFANTS

At the Pediatrician's Office:

A physical examination, including a thorough tummy check, will be done at each office visit.

Laboratory tests:

-a complete blood count, if necessary

-a urine test to check for blood in the urine.

Ask lots of questions.

Ask the Doctor About:

The results of any genetic tests done since the last visit.

The results of any consultations with other specialists.

Copies of any reports from other medical specialists.

Copies of any laboratory test results.

Keep these in your child's Medical File at home.

Your child's history of ear infections, and problems like pneumonia.

If there have been many of these, should there be any changes in the treatment plan?

The plan to monitor for Wilm's tumor.

Are ultrasounds scheduled every 3 months?

Checking for lumps in your child's tummy.

The doctor can teach you how to do this at home.

Your child's growth and development. Children with WAGR syndrome are at risk for developmental delay, so should be referred to Early Intervention Services early in infancy.

Referrals you may be given: INFANTS

·Genetics (to confirm the diagnosis, and to order additional tests, if needed)

·Urology (specialist for genital and/or urinary tract problems)

·Hematology/Oncology (cancer specialists, for treatment of Wilm's tumor, if it occurs)

·Early Intervention Services (Therapy to maximize mental and physical development)

·Pediatric Ophthalmology (eye doctor for children)

CHECKLIST FOR YOUR CHILD: 1 TO 5 YEARS: EARLY CHILDHOOD

At the Pediatrician's Office:

A physical examination, including a thorough tummy check, will be done at each office visit.

Laboratory tests, including:

-a complete blood count

-a "lipid profile" (complete test for cholesterol)

-a urine test to check for blood and/or protein in the urine.

Ask the Doctor About:

A "Medical Home."

Is your pediatrician helping you to coordinate and manage all the medical specialists?

Are the specialists sending written reports back to your pediatrician?

Copies of any reports from other medical specialists.

Copies of any laboratory test results.

Keep these for your child's Medical File at home.

Your child's behavior.

Discuss symptoms of autism and other behavior disorders.

If your child has symptoms of a behavior or psychiatric disorder, request a referral for assistance with these.

Your child's activity level and sleep patterns.

How do these compare to typical children?

If there's a big difference, how do we go about investigating and treating the cause?

Your child's infections.

Does he have more of these than a typical child?

If so, should the treatment plan be adjusted to allow for prompt diagnosis and thorough treatment?

Wilms tumor. Peak time to develop Wilms is between ages 1 and 3 years.

Are ultrasounds scheduled every 3 months? Are you doing tummy checks in-between?

Your child's motor development.

Is he walking properly, moving normally?

Your child's weight.

If he or she is overweight, what can we do, and how should it be done?

Your child's social and intellectual function.

Are any additional services needed?

Referrals you may be given: EARLY CHILDHOOD

·Hematology/Oncology, if necessary (cancer specialists, if Wilm's tumor develops)

·Pediatric Neuropsychiatry (specialist in the diagnosis and management of behavior disorders)

·Pediatric Ophthalmology (eye doctor for children)

·Nephrology, if necessary (kidney specialist)

·Otolaryngology, if necessary (ear, nose and throat specialist)

·Orthopedics, if necessary (muscle and bone specialist)

- Dietary Consultation, if necessary (to help with diet, nutrition, and weight management)
- Pediatric Dentistry, if necessary
- Pediatric Gastroenterology, if necessary (specialist in problems like pancreatitis)

Recommended Follow-up for Wilm's Tumor Survivors

The tests below have been recommended by the National Wilms tumor Study Group (USA)

·Laboratory tests: CBC, WBC/differential, Liver Function Tests (AST, ALT, AlkPhos, Bilirubin)
Renal Function Tests (BUN, plasma Creatinine, GFR) Urinalysis, 24-hour urine collection.

·Blood pressure

·If child received Ifosfamide (Cisplatin): Blood and Urine pH, Electrolyte plasma and urine levels (K, P, bicarbonate and uric acid)

·If child received Adriamycin (doxorubicin): Echocardiogram and MUGA scan. Refer to cardiology as needed.

·If child received radiation or was treated for CCSK: Yearly skeletal survey and bone scan until child is fully grown, then every 5 years, indefinitely.

CHECKLIST FOR YOUR CHILD: FROM 5 TO 13 YEARS: LATE CHILDHOOD

At the Pediatrician's Office:

A physical examination, including a thorough tummy check, will be done at each office visit.

Laboratory tests, including;

-a complete blood count

-a "lipid profile" (complete test for cholesterol)

-a urine test to check for blood and/or protein in the urine.

Blood pressure check.

Ask the Doctor About:

A "Medical Home."

Is your pediatrician helping you to coordinate and manage all the medical specialists?

Are the specialists sending written reports back to your pediatrician?

Copies of any reports from other medical specialists.

Copies of any laboratory test results. Keep these in your child's medical file at home.

Tests for girls with WAGR.

If your daughter has streak ovaries, these should be monitored at least once a year with either pelvic ultrasound or MRI. If the status of her ovaries is unknown, then regular ultrasound or MRI monitoring should be done.

Kidney failure.

Children with WAGR have a very high risk for developing kidney failure, most often during their teen years. This is true *even if they never had Wilms tumor*

Appropriate tests for this include:

- Blood pressure
- Measurement of protein in the urine.
- Blood tests to measure kidney function.

High blood pressure, or more than a trace of protein in the urine, should prompt a referral to a nephrologist.

Your child's behavior.

Discuss symptoms of autism and other behavior disorders.

If your child has symptoms of a behavior disorder, request a referral for assistance with these.

Your child's activity level and sleep patterns.

Children with WAGR are at risk for obstructive sleep apnea.

How does your child's activity level and sleep patterns compare to typical children?

If there's a big difference, how do we go about investigating and treating the cause?

Wilms tumor

Wilms tumor has been diagnosed in people with WAGR as late as age 25.

Some doctors continue abdominal ultrasounds indefinitely, often at 6 to 12 month intervals. Blood pressure monitoring and checking for blood in the urine should also continue indefinitely.

Your child's weight.

If he or she is overweight, what can we do, and how should it be done?

School and classroom placement.

Are these appropriate and adequate?

Are any additional services needed?

Referrals you may be given: Late Childhood

·Hematology/Oncology, if necessary (cancer specialists)

·Pediatric Neuropsychiatry (for diagnosis and management of behavior disorders)

·Pediatric Ophthalmology (children's eye doctor)

- Nephrology, if necessary (kidney specialist)
- Orthopedics, if necessary (muscle, bone, movement disorders)
- Gynecology (specialist care for females)
- Dietary Consultation , if necessary (nutrition and weight management)
- Pediatric Dentistry, if necessary
- Pediatric Gastroenterology, if necessary (specialist in problems like pancreatitis)

Recommended Follow-up for Wilm’s Tumor Survivors

The tests below have been recommended by the National Wilms tumor Study Group (USA)

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Renal Function Tests (BUN, plasma Creatinine, GFR) Urinalysis, 24-hour urine collection.

·Blood pressure

·If child received Ifosfamide (Cisplatin): Blood and Urine pH, Electrolyte plasma and urine levels (K, P, bicarbonate and uric acid)

·If child received Adriamycin (doxorubicin): Echocardiogram and MUGA scan. Refer to cardiology as needed.

·If child received radiation or was treated for CCSK: Yearly skeletal survey and bone scan until child is fully grown, then every 5 years, indefinitely.

CHECKLIST FOR YOUR CHILD: 13 TO 21 YEARS OR OLDER: ADOLESCENCE TO ADULTHOOD

At the Pediatrician’s Office:

- A physical examination, including a thorough check of the abdomen, will be done at each office visit.
- Laboratory tests, including;**

- a complete blood count
- a "lipid profile" (complete test for cholesterol)
- a urine test to check for blood and/or protein in the urine.

Blood pressure check.

Ask the Doctor About:

A "Medical Home."

Is your pediatrician helping you to coordinate and manage all the medical specialists?
Are the specialists sending written reports back to your pediatrician?

Copies of any reports from other medical specialists.

Copies of any laboratory test results.

Keep these in your child's medical file at home.

Tests for girls with WAGR.

If your daughter has streak ovaries, these should be monitored at least once a year with either pelvic ultrasound or MRI. If the status of her ovaries is unknown, then regular ultrasound or MRI monitoring should be done.

Kidney failure.

Children with WAGR have a very high risk for developing kidney failure, most often during their teen years. This is true *even if they never had Wilms tumor*

Appropriate tests for this include:

- Blood pressure
- Measurement of protein in the urine.
- Blood tests to measure kidney function.

High blood pressure, or more than a trace of protein in the urine, should prompt a referral to a nephrologist.

Your child's behavior.

Discuss symptoms of autism and other behavior disorders.

If your child has symptoms of a behavior disorder, request a referral for assistance with these.

Adolescence to Adulthood(continued)

Your child's activity level and sleep patterns.

Children with WAGR are at risk for obstructive sleep apnea.

How does your child's activity level and sleep patterns compare to typical children his/her age?

If there's a big difference, how do we go about investigating and treating the cause?

Wilms tumor

Wilms tumor has been diagnosed in people with WAGR as late as age 25.

Some doctors continue abdominal ultrasounds indefinitely, usually at 6 to 12 month intervals.

Blood pressure monitoring and checking for blood in the urine should also continue indefinitely.

Your child's weight.

If he or she is overweight, what can we do, and how should it be done?

School and classroom placement.

Are these appropriate and adequate?

Are any additional services needed?

Transitioning to adulthood issues.

You may want to begin discussing things like vocational training, sexuality, guardianship, group homes/independent living options, and transferring to adult medical care.

Referrals you may be given: Adolescence to Adulthood

·Hematology/Oncology, if necessary (cancer specialists)

·Pediatric Neuropsychiatry (for diagnosis and management of behavior disorders)

·Pediatric Ophthalmology (children's eye doctor)

·Nephrology, if necessary (kidney specialist)

·Orthopedics, if necessary (muscle, bone, movement disorders)

·Gynecology (specialist care for females)

·Dietary Consultation , if necessary (nutrition and weight management)

·Pediatric Dentistry, if necessary

·Pediatric Gastroenterology, if necessary (specialist in problems like pancreatitis)

If your child is a survivor of Wilms Tumor, see the Recommendations for follow-up of survivors, above