

Wilms Tumor in WAGR Spectrum

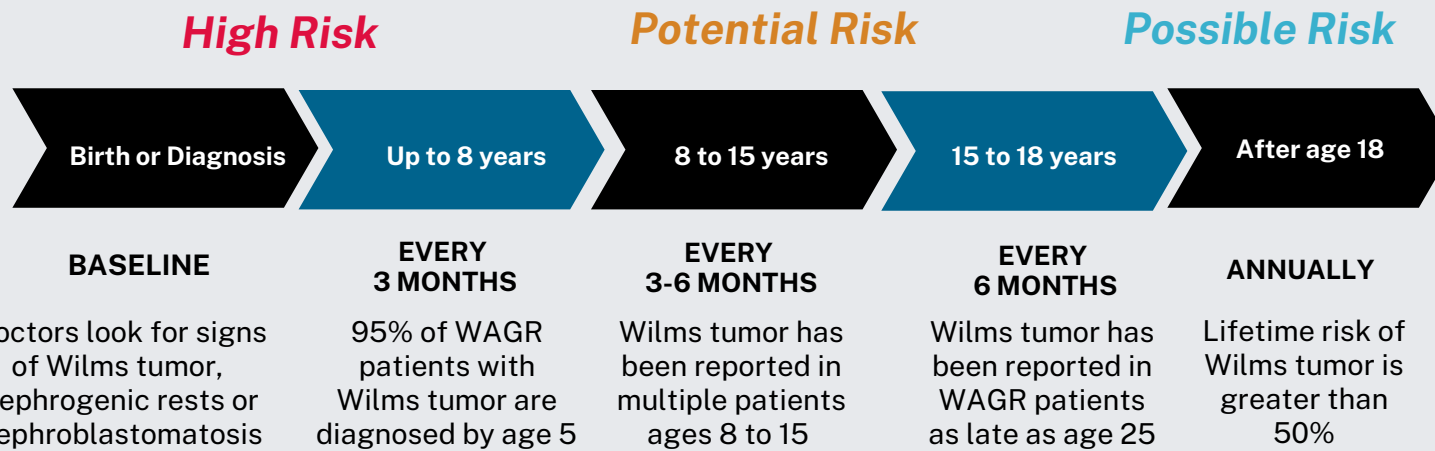
About 50% of people with WAGR spectrum develop Wilms tumor

Regular screening is important for early detection and treatment. Most cases occur before age 8, but can occur after age 8, and in very rare cases, in adulthood.



RECOMMENDED SCREENING*

ULTRASOUND OF KIDNEYS



Wilms tumor: a tumor (cancer) of the kidney

Nephrogenic rest: a cluster of abnormal cells in the kidney. Not cancer, but can become cancer

Nephroblastomatosis: two or more areas of nephrogenic rests (clusters of abnormal cells) in one or both kidneys

Screening Guidelines for Wilms tumor*

(A) Summary of General Recommendations for Screening Programs			
General Recommendations for Screening Programs in Patient Populations			
<p>Children with Predisposition to Wilms Tumor</p> <p>(2016 AACR Childhood Cancer Workshop¹)</p>	<ul style="list-style-type: none"> • ‘These recommendations were designed to offer screening in cases with a 1% or greater risk when early detection is minimally invasive and significantly improves outcome.’ • ‘We acknowledge that uniform recommendations may result in some patients being screened more frequently and for a longer duration than some clinicians have previously determined to be necessary. Therefore, these recommendations should be discussed with each family...’ • ‘Surveillance can be further tailored on the basis of the disorder and knowledge regarding the specific characteristics of the tumors that occur in the syndrome.’ 	<p>Lifetime risk for WT cannot be estimated in WAGR Spectrum (>1% risk is conceivable)</p> <p>>50% of patients with WAGR develop WT, nephroblastomatosis, and/or NR during their lifetime</p> <p>Risk Stratifications by Age for WAGR Spectrum:</p> <ul style="list-style-type: none"> High risk (<8 years) Potential risk (8-15 years) Possible risk (>15 years) 	
<p>Early Identification and Intervention of Chronic Kidney Disease</p> <p>(2019 KDIGO controversies conference²)</p>	<ul style="list-style-type: none"> • Conclusion 1: ‘Persons with hypertension, diabetes, or cardiovascular disease should be screened for CKD.’ • Conclusion 2: ‘CKD screening and treatment programs should also be implemented in other high-risk individuals and populations based on comorbidities, environmental exposures, or genetic factors.’ • Conclusion 3: ‘The initiation, frequency, and cessation of CKD screening should be individualized based on kidney and CVD risk profiles and individual preferences.’ 	<ul style="list-style-type: none"> • Adverse cardiometabolic profiles (CVD and Obesity) and CKD are prevalent in patients with and without history of WT • There is a high risk for abnormal kidney consequences due to 11p13 deletions and health issues in patients with WAGR Spectrum 	
(B) Ultrasonography Screening Recommendations for Patients with WAGR Spectrum			
Birth or Diagnosis	Full Abd US Pelvic US	Initiate WAGR Spectrum Screening Program: (1) Evaluate baseline kidney and organ status; (2) Diagnose CAKUT and/or internal GU anomalies; (3) Look for signs of NR, nephroblastomatosis, or WT growth and/or potential early UTI to determine follow-up screening required for patient	
Patient Age Groups	Interval and Screening Techniques	Tumor Screening Management Aspects	General Kidney Health Aspects
<8 years of age	3-month RUS*	Monitor high risk for WT, nephroblastomatosis, and/or NR; Manage risks for other adverse kidney issues	<p>GOAL: EARLY DETECTION AND TREATMENT</p> <p>(1)Diagnose and manage CAKUT (2)Screen/Work-Up potential UTIs, CKD, or other signs of kidney damage (3)Monitor CKD status and progression to determine and evaluate treatment (4)Prevent or mitigate cardiometabolic consequences that contribute to severe kidney failure and ESRD</p>
8 th birthday	Full Abd US Pelvic US	Evaluate for other abdominal and/or internal GU tumors or abnormalities; Determine follow-up interval for RUS	
8 years - 15 years	3- or 6-month RUS*	Evaluate kidney health status and RUS follow-up interval; Monitor risk for CKD and/or potential risk for WT, Nephroblastomatosis and/or NR	
15 years - 18 years	6-month RUS*	Evaluate kidney health status and RUS follow-up interval; Monitor risk for CKD and/or possible risk for WT, Nephroblastomatosis and/or NR	
18 th birthday	Full Abd US Pelvic US	Evaluate for other abdominal and/or internal GU tumors or abnormalities; Determine follow-up interval for RUS	
>18 years of age	Annual RUS*	Monitor overall kidney health status and CKD signs; Manage possible lifetime WT risk	
Frequency Intervals Suggested and Considerations for Implementation			Purpose of Screening Interval
1-month and/or 2-month interval	Patients with history of recent UTI or recurrent UTIs to monitor treatment as determined by medical care team Provides shorter duration to evaluate clinical issues or concern for CAKUT, WT, CKD (may provide underlying etiology for clinical signs)		More frequent for patients with clinical signs to enable earlier detection than 3-month interval
3-month interval	Necessary for patients designated as ‘high risk’ for WT based on age (<8 years) and/or clinical characteristics (history of previous WT or NR); Can also assist monitoring CAKUT issues and provide early signs of CKD development and/or progression; Preferences of care team and patient/family should be considered Interval period for patients between 8-18 years that do not meet consideration for 3-month frequency		Routine WT screening and UTI monitoring
6-month interval	Consider implementing more frequent interval for those with clinical issues identified at annual screen and/or if preferences of care team and patient/family direct more frequent screening intervals		Routine WT, UTI, CKD monitoring or screening
Annual interval	Necessary minimum interval to screen for CKD and WT in patients >18 years with stable cardiometabolic and CKD health status; Preferences of care team and patient/family should be considered		Routine CKD monitoring and long-term WT screening

Risk

- More than 50% of individuals develop Wilms tumor, nephroblastomatosis and/or Nephrogenic Rests
- Highest risk occurs before 8 years of age
- Risk is lower after age 8, but continues into adulthood

Screening

Birth to age 8 years Ultrasound every 3 months

8-15 years Ultrasound every 3 to 6 months

15-18 years Ultrasound every 6 months

After age 18 Ultrasound every year

* Duffy KA, Trout KL, Gunckle JM, Krantz SM, Morris J, Kalish JM. Results From the WAGR Syndrome Patient Registry: Characterization of WAGR Spectrum and Recommendations for Care Management. *Front Pediatr.* 2021;9:733018. Published 2021 Dec 14. doi:10.3389/fped.2021.733018