



Renal Failure and WAGR Syndrome

Renal (kidney) failure is a common feature of WAGR syndrome. Up to 60% of individuals with WAGR syndrome may develop renal failure at some point in their lives, most often during adolescence or early adulthood.

Who Is At Risk?

Currently there is no method for predicting which individuals with WAGR syndrome will develop renal failure. Anyone with WAGR syndrome is at risk for this complication, including those who have never had Wilm's tumor.

What Causes Renal Failure in People with WAGR Syndrome?

The type of renal failure associated with WAGR syndrome is called "Focal Segmental Glomerulosclerosis," or FSGS. This type of renal failure can occur with a number of other medical conditions, such as diabetes. It is not known why people with WAGR syndrome develop FSGS, but it is thought to be the result of the genetic defect that causes WAGR syndrome.

FSGS refers to a condition in which the glomeruli, the tiny tubes which filter the blood to make urine, become scarred. Instead of filtering properly, these damaged glomeruli allow protein to appear in the urine. Over time, the kidneys lose their ability to function properly, and kidney failure is the result.

How Is FSGS Treated?

The course and outcome of FSGS varies from one patient to another. Some patients with FSGS respond well to medication, and their kidney function may remain relatively stable for long periods of time. Other patients may go quickly from diagnosis to the need for dialysis or kidney transplant.

Many patients with FSGS respond well to treatment with a class of drugs called angiotensin converting enzyme (ACE) inhibitors. ACE inhibitors are not a cure, but they do appear to slow the progression of the disease in some patients. The same may be true for patients with WAGR Syndrome and FSGS.

Signs of renal failure may include hypertension (high blood pressure) hyperlipidemia (very high cholesterol) and/or proteinuria (protein in the urine)

Early diagnosis of renal failure is important, because appropriate treatment may help prolong the time between diagnosis of the condition, and the need for dialysis or kidney transplant. Guidelines for testing and monitoring at each stage of life are available here:

[Health Supervision for Individuals with WAGR Syndrome>](#)

To learn more about the function of the kidneys, FSGS, or living with chronic renal failure, please visit the following websites:

<http://www.nephron.com>

<http://www.renalnet.com>

<http://www.kidney.org>

See also: **Characteristics and outcomes of children with the wilms tumor-aniridia syndrome: a report from the National Wilm's Tumor Study Group** J Clin Oncol. 2003 Dec 15;21(24):4579-85)

Pubmed link>

http://www.ncbi.nlm.nih.gov/entrez/query.fcgi?cmd=Retrieve&db=pubmed&dopt=Abstract&list_uids=14673045&query_hl=1