

Wilms' Tumor Syndromes



PLAY PICMONIC

WAGR Complex

WA-Group

WAGR Complex stands for Wilms' tumor, Aniridia, Genito-urinary malformations, and intellectual disability (formerly known as mental retardation).

Intellectual Disability (Mental Retardation)

Book Covered in Tar

Intellectual disability in WAGR syndrome can be seen in the borderline-severe form.

Aniridia (Absence of Iris)

Absent-iris

Aniridia is a panocular disorder affecting the cornea, iris, intraocular pressure, lens, fovea, and optic nerve. Patients can present with nystagmus and visual acuity impairment. Milder findings of aniridia can manifest as subtle iris structure changes, no vision impairment, and normal foveal structure. Other symptoms are strabismus and occasionally microphthalmia.

Genitourinary Malformations

Genitourinary Malformed

Genitourinary malformations affect those of the male sex more than those of the female sex. These malformations present with undescended testes in those of the male sex and as undeveloped clumps of tissue called streak gonads, functional ovaries, and bicornate uterus in those of the female sex.

Denys-Drash Syndrome

Denim-Crash

Denys-Drash Syndrome involves kidneys and genitalia. The kidney disorder can begin in the first few months of life. 90% of patients can develop Wilms tumor.

Diffuse Mesangial Sclerosis

D-fuse Maze-angel Skull-roses

Glomerulopathy in Denys-Drash Syndrome can present with Diffuse Mesangial Sclerosis. This presentation will result in nephrotic syndrome within the first year of life.

Gonadal Dysgenesis

Gonad-gopher Disc-genie

Denys-Drash syndrome patients are commonly seen with 46XY and ambiguous or female external genitalia and streak (undeveloped) gonads or testes. Kidney disease developed at the age of 4.

Beckwith-Wiedemann Syndrome

Beckham-with-Wide-man

Beckwith-Wiedemann Syndrome is a growth disorder that presents with organomegaly, macroglossia, omphalocele, and hemihyperplasia.

Organomegaly

Big-organ

Organomegaly can be present with one or more enlargements of intra-abdominal organs, such as adrenal glands, kidneys, liver, spleen, and or pancreas.

Macroglossia

Macaroni-glass Tongue

Macroglossia is defined as an abnormal enlargement of the tongue. This condition is present with protruding enlarged tongue surpassing the alveolar ridge in a resting position. This enlargement can disturb the child patient's ability to speak, eat, or breathe.

Omphalocele

O-full-of-eels

The most common syndrome that is related to omphalocele is Beckwith-Wiedemann syndrome. Omphalocele is characterized by a persistent herniation of abdominal organs via the umbilicus.

Hemihyperplasia

Half-hiker-plates

Hemihyperplasia is characterized by asymmetric overgrowth, which presents with all or part of one side of the body. Examples may include the involvement of a single limb, an entire half of the body, one side of the face, or combinations thereof.