

What is Isolated Hemihypertrophy?

Hemihypertrophy is a genetic disorder characterized by overgrowth of one side of his or her body in comparison with the other. The overgrowth may affect only one part of the body, such as the legs. However, it may involve several different areas of the body including the arms, face, and tongue. Hemihypertrophy is “isolated” when it occurs by itself. But, it can be one of several characteristics of Beckwith-Wiedemann Syndrome (BWS) or another genetic syndrome. Sometimes hemihypertrophy is not apparent at birth but can become clear as he or she grows.

Children with Hemihypertrophy may have a range of the following features:

- Overgrowth of one side of the body (*hemihypertrophy*).
- Uneven leg length. This may require a shoe lift.
- Large tongue (*macroglossia*). This can cause speech and feeding problems.
- Large abdominal organs on one side of the body. Such as the kidney, liver, adrenal glands, and pancreas.
- Increased risk of developing certain cancers during childhood. This includes Wilm’s tumor, tumors of the liver (hepatoblastoma), and other more rare tumors.

What causes Hemihypertrophy?

- **The change is not caused by anything that a mother does during her pregnancy. There is nothing a parent can do to prevent the genetic change.**
- Most children diagnosed with hemihypertrophy are the first in their family with the diagnosis. Most of the time hemihypertrophy happens because of a brand new genetic change in the child. In most cases, the specific genetic change is not identified.
- Very rarely, the genetic change is passed from parent to child. In these cases, there is up to a 50% chance for each pregnancy that the affected parent will pass on the abnormal gene to their child.
- Because the genetics of hemihypertrophy can be complex, we recommend that parents have genetic counseling to better understand their individual case. Genetic testing may identify the cause of hemihypertrophy and provide information about future pregnancies.
- The mechanism of hemihypertrophy is not completely understood. Scientists believe the genetic change causes overgrowth because the genes that decrease growth are turned off or because there are more genes that increase growth than there should be.

How is Hemihypertrophy diagnosed?

Physical features: Some features are found more often in children with hemihypertrophy than in the general population. When a child has a group of features frequently associated with hemihypertrophy, he or she may be sent for genetic testing.

Genetic testing: The genetic test requires a blood sample. This sample will be sent to a lab in order to look for genetic changes associated with hemihypertrophy. It is important to know that a normal test result does not rule out hemihypertrophy because some genetic changes are still unknown. Children who do not have the genetic change for hemihypertrophy will have the same medical management and cancer screening as children who do.

How is Hemihypertrophy treated?

Every child is different! Treatment and outcomes vary and depend on how severe the symptoms are.

- The treatment of hemihypertrophy is directed toward the specific symptoms that a child develops.
- Medical providers from different specialties may be involved in your child's care. This depends on his or her specific signs and symptoms. For example; your child may meet with surgeons, kidney doctors, dental specialists, speech specialists, genetic specialists, pediatric oncologists, and orthopedic specialists.

Important information to know

Children with hemihypertrophy usually have normal intelligence and a normal lifespan. They are capable of leading successful, happy, and fulfilling lives. Most children with hemihypertrophy grow up to be healthy adults. By adolescence, growth normalizes and the risk for childhood cancer decreases. Rarely, some adults have intellectual or physical delays. Most physical features decrease with time. Some of the features of hemihypertrophy, such as leg length differences or enlarged tongue, may need surgery to correct.

Is my child at risk for cancer?

Since hemihypertrophy is an overgrowth syndrome, children are at risk for developing certain childhood cancers. The two most common forms of cancer are Wilm's tumor (kidney tumor) and hepatoblastoma (liver tumor).

- Wilm's tumor is the most common cancer in children with hemihypertrophy. The risk of developing Wilm's tumor decreases by age eight, but rarely it can develop at a later age.
- Hepatoblastoma is the second most common form of cancer that can develop. In most cases, this cancer develops by the age of two.
- Other forms of cancer have been found in children with hemihypertrophy, but are very rare.

Will my child be tested for cancer?

Children with hemihypertrophy have about a 5 to 10% chance of developing a cancer in early childhood. For this reason, routine screening is very important! Screening can help us find cancer early. Wilm's tumor and Hepatoblastoma are very treatable forms of cancer if caught early. **Cancers treated at an earlier stage will usually need less treatment and have a higher rate of survival.**

1. Abdominal Ultrasound:

Abdominal ultrasounds are safe and painless. They do not expose your child to radiation. We recommend children with hemihypertrophy have an abdominal ultrasound every 3 months until age eight. This test is done to look for abdominal tumors, including Wilm's tumor.

2. Measurement of alpha-fetoprotein (AFP)

We recommend a blood test, AFP (alpha feta protein), every 6 weeks until age four. AFP is a hormone secreted by certain tumors. The AFP test is an easy and effective way to screen for hepatoblastoma.

3. Routine Follow Up

We recommend that all children with hemihypertrophy be seen by a pediatric specialist every 6 months until age eight, and then yearly after that. A pediatric specialist can talk to you about your child's health and examine your child for other signs of cancer.

For General Health Information:

Call or visit the Family Resource Center (a free health information library) located on the 3rd floor of St. Louis Children's Hospital. Call 314.454.2350. Email at frc@bjc.org