Turner Topic

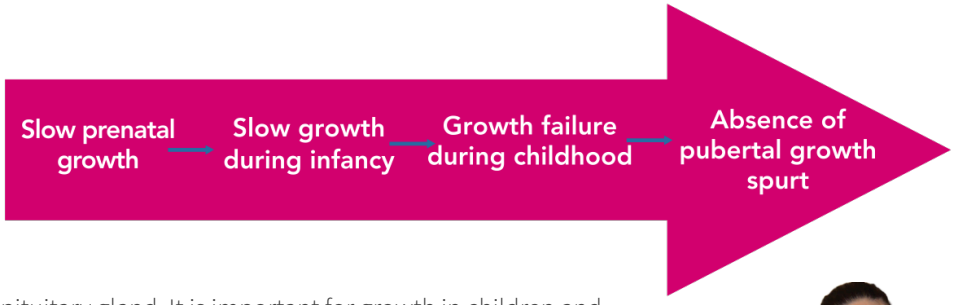
Turner Syndrome Society of the United States

A Patient Guide to Growth in Turner Syndrome

Growth in Turner Syndrome

Short stature is one of the most common and easily recognizable features of TS, with an average adult height being 4'8". Most girls with TS produce growth hormone, but their bodies don't respond to respond to it effectively. Growth failure in TS often begins before birth and growth is slow during infancy and early childhood. About 75% of girls with TS fall below the 5th percentile in height by the age of 3.5 years of age. Girls with a mosaic form of TS vary more in their growth, but 50% still fall below the 5% around 2 years of age. Many girls with TS have a delayed bone age, which means that their skeletal maturation is slower than their actual age. Because puberty speeds up bone growth and maturation, it is recommended that puberty in TS be initiated around 11-12 years of age. Physical growth stops when the growth plates of the bones fuse together, which happens at a bone age of about 15 years. Most girls with TS, who are not treated with any hormones including those who have spontaneous puberty, will not have a pubertal growth spurt but may continue to grow at a slow rate until they are in their late teens.

The typical growth pattern in Turner syndrome is characterized by:



What is Growth Hormone?

Human Growth Hormone (HGH) is produced by a part of the brain called the pituitary gland. It is important for growth in children and adolescents, but also helps regulate body composition, body fluids, muscle and bone growth, sugar and fat metabolism, and possibly heart function. A biosynthetic form of HGH (made in a lab, but with a similar structure to naturally-made GH) is available for medical use and is FDA approved for treatment in patients with HGH deficiency, or short stature from other causes.

Growth Hormone Therapy in Turner Syndrome

Growth hormone therapy is an optional treatment to increase height in girls with TS. The goals of growth-promoting therapies are to attain a normal height for age as early as possible, progress through puberty at a normal age, and attain a normal adult height. GH treatment is associated with height gains of 2 to 3.5 inches over treatment periods ranging from 5.5 to 7.5 years in several medical studies. Height gain of about 1 cm per year is a reasonable expectation of GH therapy. Some studies have shown height gains exceeding this, with height improvement sometimes reaching up to 4 to 5 inches when compared to the predicted adult height at the beginning of GH treatment. Growth hormone therapy should be directed by a **pediatric endocrinologist** and the child monitored at intervals of 3–6 months.

The benefits of growth hormone therapy

Growth hormone may be combined with another medication called Oxandrolone if adult height outcome is likely to be unsatisfactory with growth hormone alone. Studies have shown slight increases in growth response (1 to 2 inches). There is a chance of unwanted effects such as of delayed breast development and an increase in the size of the clitoris, voice-deepening, male-pattern hair growth, and acne. When correctly dosed, these complications can be avoided. Oxandrolone should not be used until around 9–10 years of age. It should be started at a dose of 0.03 mg/kg/day and maintained at no

Turner Topic

Turner Syndrome Society of the United States

Is Growth Hormone Therapy Safe?

In TS, safety of GH treatment in long-term medical clinical studies has generally been reassuring with respect to blood pressure (BP) and risk factors for heart-related disease, blood sugar and fat metabolism, body composition, bone mineralization (essential for its hardness and strength), body proportions (proper relationship of parts), and occurrence of ear infections and hearing loss.

Studies indicate that girls with TS taking growth hormone appear to be at increased risk of:

- Intracranial hypertension (pressure built-up in the head); rare occurrences
- Slipped capital femoral epiphysis (hip pain and limp); rare occurrences
- Development or progression of scoliosis (abnormal curvature of the back)
- May be at greater risk of pancreatitis; very low risk

Recommendations from International Experts on TS

- Begin GH around 4–6 years of age, or earlier if growth failure is already noticeable.
- Continue therapy until satisfied with final height or until little growth potential remains
- GH therapy dosages for TS in North America is generally initiated at a dose of 0.350-0.375 milligram/kilogram /week, in Europe at 1.3-1.4 mg/m2 /day, and in Australasia at 4.5-9.5 mg/m2/week
- Dosage is divided into 7 doses (one for each day of the week)
- GH is given in the fatty tissue under the skin as an injection/shot.
- It is not recommended to a add very-low-dose of estrogen replacement **before** puberty age to promote growth

How Much Does Growth Hormone Therapy Cost?

Costs vary because the dose is generally based on the weight of the child, how the body utilizes the hormone, and insurance coverages. The TSSUS office or your doctor can help you understand options available to you and many pharmaceutical companies offer patient assistance programs to help obtain approval for coverage or financial assistance.

The newly-updated Clinical Care Guidelines for Girls and Women with TS can be found on the TSSUS website: www.turnersyndrome.org

What Results Should We Expect?

- The results of GH vary for each girl
- On average, girls with TS who are taking growth hormone may expect an average final height gain of 2.8 inches. However, results can range from no gain to over 4.7 inches. This depends on age when GH was started, dosage, when estrogen therapy is started, and how the body responds to growth hormone.
- Many girls with TS will require treatment with estrogen to help induce normal female changes of puberty.
- Estrogen has effects on bone and increasing growth rate, but also causes the closing of the growth plates at the end of the bones in order to complete growth.
- The effects of estrogen therapy can have an impact on the effects of growth hormone response.
- Current recommendations suggest replacing estrogen at the usual age when puberty naturally occurs.

For More Information, Contact Us:

Turner Syndrome Society of the United States

11250 West Road, Suite G

Houston, TX 77065

tssus@turnersyndrome.org

www.turnersyndrome.org

1.800.365.9944