

# TURNER SYNDROME

## What is it?

Turner syndrome (TS) is a condition only affecting females as a result of an X chromosome abnormality. TS occurs in approximately 1 in 2,500 newborn females. While one X chromosome is normal, the other female X chromosome is missing or altered. TS is characterized by a variety of medical and developmental problems but the most consistent features affect bone development and growth resulting in short stature and lack of ovarian development.

Diagnosis can be made prenatally or in early childhood but over 1/3 of girls diagnosed are diagnosed in mid-childhood or adolescence. A blood test can confirm suspicion of the syndrome. The long term health outcomes are improved with an earlier diagnosis.

## What are the symptoms or complications?

Diagnosis can be made prenatally or during early childhood years. However, over 1/3 of diagnoses occur during adolescence. A blood test can confirm suspicion of the syndrome. Signs and symptoms may be subtle and develop slow over time, or they may be significant. They can occur in varying degrees based on the individual's genetic makeup.

- Short stature
- Swelling of hands and feet
- Lack of spontaneous puberty
- Kidney problems e.g. UTI's
- Heart issues e.g. congenital defects
- Type II Diabetes
- Hypertension
- Thyroid disease
- Lack of stamina
- Scoliosis
- Recurrent ear infections that may lead to hearing problems
- Webbed neck
- Droopy eyelids
- Strabismus
- Low set ears and hairline
- Poor vision
- Infertility

A child with TS will not only face medical problems but also learning disabilities. Students with TS often have a cognitive profile that includes normal intelligence and verbal capabilities but weaknesses in the areas of visual-spatial, executive, and social cognitive function. This includes variables in things like nonverbal communication, slowed response times and immaturity. Students with TS might have trouble recognizing social cues and things like facial expressions are easily misinterpreted. Students with TS may have problems with personal boundaries and struggle to keep order in their personal environment. While they can problem solve, the issues of spatial awareness and abstract concepts make subjects like geometry very difficult. Spatial awareness can also affect spelling, punctuation, mapping, understanding time, changing point of view, and even drawing and handwriting.

## What is the treatment?

While there is no cure for TS, there are treatments available that can help with the symptoms and complications. The primary treatment for nearly all girls with TS includes hormone therapies. Growth hormone therapy is usually given as an daily injection to increase height during early childhood to the teen years. Estrogen therapy is usually started around the age of 11 or 12 to begin puberty. A girl with TS may have numerous health care specialists involved with her care depending on what medical and developmental issues she might encounter.



Kennedy Krieger Institute

The Specialized Health Needs Interagency Collaboration (SHNIC) program is a collaborative partnership between the Kennedy Krieger Institute and the Maryland State Department of Education.

## Suggested school accommodations

Supporting students with this condition in the school require educators and parents/guardian to work as a team. Some accommodations to consider for a 504/IEP could include:

- PT/OT evaluations for gross and fine motor skills
- Executive function supports e.g. clear and concise information, behavioral supports, daily schedule, graphic organizers and checklists
- Maintain structure and daily routine
- Present information first in outline form
- Hearing assessment and preferential seating
- Schedule most difficult subjects early in the day
- Practice social skills through role-playing
- Check for understanding, repeat information
- Allow extra processing time
- Offer short bursts of information
- State the obvious when teaching
- Note difficulty with fine motor skills like buttons, buckles and ties
- Note difficulty with gross motor skills
- Consider physical layout of the school as related to gross motor skills
- Maintain a quiet, calm environment
- Encourage eye contact
- Note short term memory problems
- Monitor for fatigue and need for rest breaks
- Staff education and training as appropriate
- Emergency Evacuation Plan (EEP)

## Specific health issues for Individualized Healthcare Plan

- Diagnosis including student specific characteristics
- Current medication list
- Baseline cardiac assessment including blood pressure parameters; note date of last cardiac exam, if applicable
- Fluid intake goals
- Nutrition orders
- Education on growth hormone and replacement hormone therapy, if applicable
- Monitor for social isolation, depression and anxiety
- Communicate with school staff, parents/guardian, and provider any changes or concerns about the disease
- Emergency Care Plan(s) (ECP) related to medical needs in the school setting and staff education/training as appropriate for each

## Resources & Manuals

### Turner Syndrome Foundation

<http://www.turnersyndrome.org>

### Turner Syndrome Support Society : How to help your child survive and succeed at school: A Guide for Parents and Teachers

<http://tss.org.uk/downloads/TSS%20education%20booklet.pdf>