



Wyoming Department of Education

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MEMORANDUM NO. 2013-063

TO: School District Superintendents
School Principals
School Nurses
Special Education Directors

FROM: Bruce Hayes
School Safety Consultant

DATE: May 20, 2013

SUBJECT: Turner Syndrome

STUDENT HEALTH UPDATE

Turner Syndrome is a condition affecting an average of about 1-in-2000 female students. It can cause a number of physical problems including abnormally short stature, gonadal dysgenesis, cardiac malformations, inner ear defects, obesity, scoliosis, kidney malformations, a variety of endocrine problems, and other issues including auto-immune diseases.

Additional potential problems can impact educational issues including difficulty with math, word problems, image perception and shape, and distance. Interpreting social clues and sensing others feelings can also be problems as well as having difficulty in adapting to new situations.

If you believe you may have a student like this in your school, you may need to consult with his or her teacher, tactfully consult with the parents, and ultimately arrange for a karyotype to be done for a definitive diagnosis. A delayed diagnosis may lead to future problems and missed treatment opportunities for those afflicted with Turner Syndrome.

More information can be found in the enclosed attachments and the website www.turnersyndromefoundation.org. If you have any questions or concerns, please feel free to contact Bruce Hayes, at 307-777-6198 or bruce.hayes@wyo.gov.

Attachments

A Guide for Teachers: Turner Syndrome in a Classroom Setting



TURNER
SYNDROME
FOUNDATION
INC.

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(800) 594-4585
TurnerSyndromeFoundation.org



Turner Syndrome affects 1 in 2,000 girls, and many will have special learning needs

We can help.

The goal of the Turner Syndrome Foundation (TSF) is to support research initiatives and develop educational programs which will increase professional awareness and enhance medical care of those affected by Turner Syndrome.

Early diagnosis and comprehensive treatments over the lifespan may lead to a brighter and healthier future for all young girls and women with Turner Syndrome. *TSF is a 501(c)3 nonprofit organization EIN# 27-1409942.*

Possible Indications for this lifetime condition:

- * Short stature
- * Delayed pubertal development
- * Wide short neck
- * Wide shield chest
- * Wide spaced nipples
- * Low set ears
- * Low set hairline
- * Vision issues
- * Pigmented moles and dry skin
- * Otitis media and hearing loss
- * Heart abnormalities
- * Kidney issues and infections
- * Bladder issues and infections
- * Lymphedema
- * Hypertension
- * Scoliosis
- * Short 4th metacarpal
- * Malformed bones
- * Dysmorphic facial features
- * High arched palate and dental problems
- * Upturned finger and toenails
- * Weight Issues (under and over)
- * Auto-Immune disease(s)
- * Cognitive strengths and weaknesses
- * Verbal strengths
- * Executive function weaknesses
- * Social Issues
- * Anxiety and depression

Early diagnosis and treatment.



Education and support.

Turner syndrome is a random mutation of chromosomes that affects only females. There is an absence of all or part of the second X chromosomes in some or all of the cells in the body. If a child does not exhibit classic TS characteristics, but is **short** and falling below the curve on the growth chart, has **delayed pubertal development**, presents **verbally gifted**, or expresses **academic** or **social challenges**, she may have TS. Diagnosis is made with a **Karyotype blood test**. An endocrinologist will monitor the patient, screen for other conditions and provide specialized medical care, and treatments. Early interventions can promote a positive self-esteem and improved prognosis.

Medical evidence indicates that early diagnosis and treatment is essential.

A Guide for Teachers: Turner Syndrome in a Classroom Setting

In 1 out of every 160 classrooms sits a girl with Turner Syndrome

TEACHERS:

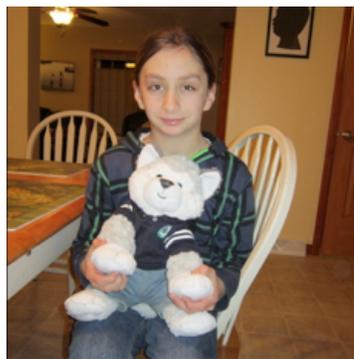
A pathway to a lifetime of success begins with a collaborative effort and support of parent and teachers.

ADMINISTRATORS:

A well-informed faculty can make a difference in a young child's life.

SCHOOL NURSES:

Girls at 5% or below normal height along with other possible indications are recommended to be screened for Turner Syndrome.



Possible Indications:

Infants: small size, puffy hands and feet, extra neck skin folds, heart abnormalities, feeding difficulties

Children: small size in relation to peers, below the "normal" growth chart for both height and weight, ear infections, hearing problems, learning difficulties

Teens: short stature, delayed puberty with no breast tissue development or menses, social issues

Adults: short stature, menstrual cycle irregularities, infertility, hearing issues, heart problems, hypertension, hypothyroidism, type II diabetes, osteoporosis

Cognitive Profile

- Physical and cognitive variability
- Spectrum disorder
- Verbal IQ is significantly higher than performance IQ. This correlates with a nonverbal learning disability (NLD/NVLD)
- Most girls will express NLD/NVLD, few do not

Diagnosing Cognitive Issues

- Neuropsychological evaluation should include Wechsler Intelligent Scale for Children WISC-IV
- All girls with Turner Syndrome will qualify for a 504 plan for accommodations as a result of a medical diagnosis
- Most girls will benefit from having an I.E.P. in place for instruction, especially before heading to post-secondary education

Cognitive Functioning

- Turner Syndrome is not associated with mental retardation
- Impaired working memory; Impaired visual-spatial abilities; Impaired executive functions; Impaired attention and focus

Cognitive Strengths

- At or above average with peers in word recognition, reading and spelling; some are avid readers
- Relative strength in basic arithmetic, number comprehension and productivity, comparing numbers, and estimating accurately

Cognitive Weaknesses

- Organize, plan, and implement multi-step problem solving
- Managing materials
- Following directions
- Initiating tasks (low motivation or arousal)
- Cognitive flexibility
- ADD/ADHD
- Math learning disorder
- Verbal fluency issues

Social Functioning Issues

- Difficulty reading social cues and facial expressions
- Immature behavior
- Relates with younger or older peers
- Physical size (height/weight)
- Primed to be ostracized and/or bullied
- Emotionally sensitive

Recommendations

Younger Children

- Prepare child for daily routine and inform of changes
- Provide parents with information for their added support at home
- Introduce new activities one step at a time
- Introduce to positive peer role models
- Small group or in-class support

Recommendations

Older Children

- Provide & update planner
- Provide copy of notes
- Organizational guides & instruction
- Preferential seating
- Added test-time, modified work
- Teach memory & concentration strategies
- Technology: calculator & word processor for writing
- Reinforce verbal strengths with class discussions
- Show sensitivity to feelings of being "overloaded"
- Provide opportunities of strengths to shine in classroom
- Repeated verbal instruction and detailed explanations,

Recommendations

Older Children Cont'd.

- Provide non-threatening interactions with peers
- Counselor and social worker to provide added support as needed
- Inclusion in intramural and extra-curricular activities encouraged by faculty
- Focus on child's strengths
- Work to improve expression of weaknesses

Optimal Learning Environment

- Inclusive learning
- Pleasant instructor
- Positive reinforcement
- Proactive measures
- Willingness to verbally cue student, repeatedly
- Coaching the student towards preparedness and academic success with short and long term goals
- Flexibility
- Open communications with student and parents

Parents

- Share diagnosis with school nurse and Director of Special Education
- Advocate for testing, services and interventions as needed
- Keep open lines of

School Nurses and Educators' role in identifying students with Turner Syndrome

By Rosemary Scales, MS, BSN

Have you seen female students who are clearly shorter than their peers? Have frequent ear infections? Have frequent urinary tract infections? Have had a history of heart surgery at an early age? Seem to have difficulty with math after 4th grade? Get lost in the building? You may know a Turner Syndrome (TS) young lady. She may not have even been diagnosed with TS yet. **YOU** could be instrumental in obtaining the diagnosis that is so important to find as early as possible.

Turner Syndrome is a chromosomal deletion (full or partial) that causes short stature and gonadal dysgenesis in 1 out of 2000 to 2500 females. Most girls diagnosed with TS will have these 2 characteristics as well as a number of other characteristics. Since they exhibit varying characteristics, the diagnosis may be difficult to determine at an early age. The following list contains the physical characteristics in Turner Syndrome that may/may not be exhibited.

| | |
|-------------------|--|
| FACIES | Narrow maxilla, wide mandible, flattened cranial base, marked reduction in posterior cranial base length, narrow and high-arched palate, small chin (micrognathia), chin sits farther back than normal (retrognathia) |
| SKIN/NAILS | Broad chest with widely spaced nipples (shield chest), occasional inverted nipples, short webbed neck, nuchal folds as an infant, low posterior hairline, occasional start of hair growth pointing upwards, low and rotated ears, occasional deformed auricles, lymphedema of hands and feet as an infant (improves with age), increased body hair growth, bushy eyebrows, hyperconvex uplifted or underdeveloped/ hypoplastic nails, sometimes absent toenails, increase in pigmented moles, telangiectasis ("birthmark") |
| DENTAL | Defective dental development (distal molar occlusion, anterior and lateral open bite, lateral cross bite, early eruption of secondary teeth, simple crown morphology, thinner enamel, less dentine, and short roots), are at greater risk for root resorption. |
| OPHTHALMIC | Ptosis, strabismus, amblyopia, myopia, and/or epicanthus, occasional cataracts |
| AUDITORY | Inner ear defects resulting in recurrent ear infections and hearing problems, sensorineural hearing loss, sinusitis and mastoiditis may occur. |
| CARDIAC | Left-sided cardiac malformations, i.e., coarctation of the aorta, bicuspid aortic valve, stenosis of aortic isthmus, aortic dilatation/aneurysm, hypoplastic left heart, anomalous pulmonary venous return, may be a generalized dilatation of major vessels, including brachial and carotid arteries as well as the aorta, high blood pressure (in the absence of cardiac or renal problems). As adults, they can experience aortic root dissection resulting in aortic rupture. (7, p. 242) Cardiovascular abnormalities represent the single highest cause of death. |

| | |
|-------------------|---|
| ORTHOPEDIC | Cubitus valgus (deviation of the extended forearm to bend away from the body when the arms are at rest or an unusual carrying angle for the elbows), short fourth and/or fifth metacarpal, scoliosis and/or spongiotic bone structure, osteopenia, occasional arthritis, congenital hip dislocation in infants, knock-knees |
| RENAL | Kidney malformations –may be any one of 3 types, 1.) defect in collecting system resulting in partial or complete duplications 2.) positioning as in horseshoe, ectopic, or malrotated kidneys or 3.) variation in number and position of renal arteries |
| ENDOCRINE | Primary hypothyroidism, Hashimoto thyroiditis, glucose intolerance, diabetes mellitus |
| OTHER | Autoimmune diseases such as inflammatory bowel disease, Crohn’s disease, celiac disease and rheumatoid arthritis Obesity |

Early diagnosis resulting in early referral for treatment is imperative. Most of the characteristics mentioned above require treatment. Possible treatment includes growth hormone therapy, estrogen replacement as a teen, calcium supplements and possibly thyroid replacement therapy. Growth hormone therapy is the most vital medication to start as soon as possible so the girls do not have to “catch up” growth, but can keep growing at the same rate as their peers. These are coordinated by the pediatric endocrinologist. Dietary/exercise counseling may be necessary. Surgical repairs may be required for cardiac and renal anomalies as well as cosmetic procedures for a webbed neck and perhaps breast augmentation. Scoliosis treatment may also be required. Other recommended procedures include an ophthalmologic exam, an ENT/audiology exam, blood pressure monitoring, scoliosis exam, DEXA scan, an orthodontic evaluation, and comprehensive psychosocial and academic assessments. Speech therapy, educational interventions and a psychosocial plan may also be necessary. These multiple treatment needs make it obvious why early diagnosis is advantageous.

What should you do if you observe a young lady with these characteristics? Consulting with her teacher may be your first step. These young ladies may show evidence of nonverbal learning disorder, a combination of learning, academic, social and emotional issues. They may have early development of speech, vocabulary, reading and spelling skills. However, reading comprehension is weak. They may process at a very concrete level and interpret information literally. They have problems with image perception, sense of direction, estimation of size, shape and distance. They also may have difficulty arranging written material on a page, placing written responses, organizing information. Math is particularly challenging as they have difficulty with mechanical arithmetic, word problems and math skills that rely on spatial abilities or seeing the relation between concepts. They also experience difficulty interpreting social cues, sensing the feelings of others, difficulty adapting to new and complex situations. Again they may exhibit few or many of a long list of characteristics of nonverbal learning disorder. If their teacher has noticed these, this may help verify your suspicion of Turner Syndrome.

The next step may be to tactfully consult with her parents to see if she has been diagnosed with Turner Syndrome but that may not have been shared with the school personnel. If the diagnosis has not been made or an evaluation has not been done, a suggestion to have the primary care provider follow up on her short stature and other characteristics may be in order. If your student is exhibiting any of these characteristics, a karyotype should be done to provide a definitive diagnosis. The primary care provider would then refer her to a pediatric endocrinologist. At diagnosis, this would include more testing. Radiologic procedures necessary include an echocardiogram and/or a cardiac MR, a renal/pelvic ultrasound, a bone age and any other x-rays (i.e., scoliosis, etc.) indicated.

If a positive diagnosis is made, educating the teachers in her school is crucial so that the suitable accommodations can be made to improve her school experience. It would also be helpful to share the possible characteristics listed above with teachers who would be able to observe for these in the classroom in the event that the school nurse does not have an opportunity to see them.

Because few characteristics may be obvious, this diagnosis may be easily missed. A delayed diagnosis may lead to obstacles in health care and education while an earlier diagnosis will be beneficial when treatment, medications and school accommodations can be initiated in a timely fashion.

Submitted by: www.turnersyndromefoundation.org

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