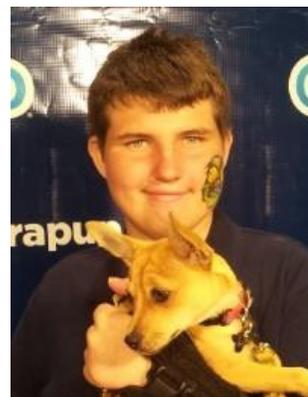


Klinefelter Syndrome *at a Glance*

Klinefelter syndrome (KS) is caused by a difference in the sex chromosomes (XXY). It affects males, occurring in 1 in 650 males. KS can cause a variety of problems including breast development, infertility, and hormonal changes. Some males with KS have learning and language problems.



Meet Graham on page 8.

Things to Think About

1. Medical / Dietary Needs

What you need to know

No special diet is required for KS although a well balanced diet is important.

It is important to ask the parents about the medical issues in their child and what medical management challenges they may face at school.

Many boys with Klinefelter are followed by an endocrinologist when they reach the age of puberty. They may be placed on testosterone treatments.

School age children with KS may have annual doctor and specialist visits to monitor medical conditions.

Physical characteristics and/or symptoms:

Not all people with Klinefelter syndrome have all of these characteristics.

Low testosterone is the reason for many issues in KS. The features vary widely between individuals with KS and many individuals with KS are diagnosed as adults because of infertility

and have few of the physical features.

- Tall stature
- Longer legs
- Shorter torso
- Broader hips
- Reduced facial and body hair
- Reduced muscle mass
- Absent, delayed or incomplete puberty
- Weak bones
- Hypogonadism (reduced or absence of hormone secretion of the testes)
- Fertility problems
- Increased risk diabetes and /or hypothyroidism
- Dental problems
- Scoliosis
- Swelling of breast tissue during puberty
- Cognitive, reading, and language difficulties
- Depression and/or social differences
- Adults with KS have increased risk of breast cancer and systemic lupus

What you can do

- A yearly check-up and studies as needed should occur in the child's Medical Home.
- Be aware of any changes in behavior or mood that seem out of line and notify the parents.
- Be aware of any academic changes. Contact parents when any differences are noticed.
- Provide and seek appropriate supports as needed, whether school based or more medically based:
 - Communication Supports
 - OT or PT for supporting motor development, coordination, tone and sensory needs
 - Supports for emotional or behavioral issues that might arise, as needed

2. Education Supports

What you need to know

It is important to have HIGH LEARNING EXPECTATIONS for children who have Klinefelter syndrome. Encourage use of the core educational curriculum and modify it in order to meet

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the individual needs of the child.

Many individuals with KS grow up to be successful in academics, careers, and personal lives. However, there are differences related to the presence of an extra “X” chromosome. This includes differences in cognitive, language and psychological growth. Individuals with KS typically don’t usually have an intellectual disability. Most have average to low average intelligence.

The following may occur in boys who have XXY

- Delayed speech
- Gross and fine motor delays
- Sensory integration difficulties
- Sensitivity to sound, touch, movement
- Low muscle tone
- ADHD
- Problems with spelling and math

Individuals with KS often have auditory processing and language based learning disabilities. These have been identified in 70-80% of children with KS.

Difficulties in the following areas may occur

- Expressive speech and language
- Receptive language
- Lower skills in auditory, memory, confrontation naming (word retrieval), and verbal fluency
- Specific reading disability ~50-75%
- Individuals may experience delays in meeting speech and language milestones.
- Difficulty in conversation, expressing personal thoughts, opinions and needs may occur.

If learning difficulties aren’t addressed it can lead to lower academic achievement, reduced self-esteem, and behavior problems.

- Boys with KS may have trouble using language to express their thought and needs
 - May have difficulty putting thoughts, ideas, emotions into words.
 - May find it hard to learn and remember words of common items.
- They may have difficulty processing what they hear
 - May appear to “tune out” or fidget because it takes longer to process information.

- May find it difficult to concentrate in a noisy class.
- Many boys have difficulty understanding what they read.
 - May read slower

What you can do

Provide and seek appropriate classroom supports as needed.

Academic Supports for reading comprehension, math, language

- Help with organization
- Present information in concrete manner
- Provide routine, structure, and consistency
- Use manipulative materials to demonstrate concepts
- Simplify verbal information and explain concepts clearly
- Provide visual cues and instructions
- Repeat information and use positive reinforcement.
- Provide quiet learning environment, background noise may be distracting
- Help prioritize work and activities
- Allow a “timeout” from concentration
- Reach good study skills
- Use technology when appropriate

Communication Supports

- Appropriate and timely intervention by a speech and language pathologist can help keep speech and language skills on track.
- Promote language understanding by using simple short sentences, visual prompts, and pictures.
- Use a child’s experiences and interests to engage child in learning
- Allow extra time, repeat directions, provide lesson summaries, and record lessons so child can listen again.
- Have child repeat directions
- Creating situations in which student’s can practice their skills in natural settings is suggested for therapy.
 - Allow opportunities to use descriptive expression throughout the day

Promote language development

- Providing ample time for responding
- Increasing the child’s self confidence by calling on them when they know answer
- Encouraging a child to repeat the questions before responding

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- Allowing a child time to rehearse and respond

OT or PT

- Supporting motor development, coordination, tone and sensory needs

Supports for auditory processing

- Instruction needs to be repeated
- Poor concentration
 - Provide non-distracting environment

Supports for emotional or behavioral issues

- Monitor and evaluate behavior and emotions.

3. Behavioral and Sensory Support

What you need to know

Many social and behavioral symptoms in KS may be related to language and learning difficulties. Language difficulties may inhibit social interactions as the boys may have difficulty expressing feelings.

Boys with KS may be:

- Quieter/shy
- Less assertive or self-confident, or immature
- Sensitive
- Delayed in social skills
- Anxious or depressed
- Restless
- Less physically active
- More helpful and eager to please

Teenage years

- Teenage boys may notice differences between them and other boys.

- They are at risk for depression, substance abuse, behavior problems.
- They may feel withdrawn and/or sad.
- They are no more likely to have serious psychiatric disorders or get in trouble with the law.

What you can do

- Support healthy emotional and behavioral development
 - Teach self-talk to help child develop self-control. Use specific, short phrases such as “stop and think.”
 - Give directions one-step at a time. Wait for the child to do the first step in the directions before telling the child the second step
 - Assist with social skill development
 - Teach basic rules of social behavior
 - Model, rehearse, and practice and provide feedback
 - Support development of self-esteem
 - Find out what activities are meaningful to the child and help them join in. For example, encourage child to participate in sports, clubs or other structured activities
 - Make sure language supports are in place to help with social development
- Monitor emotional and behavioral health and refer as necessary.
 - Learn how to tell when child is getting frustrated and help out early
 - If the child has anxiety and depression:
 - Medication and or counseling may be helpful

4. Physical Activity, Trips, Events

What you need to know

- Exercise is important and should be encouraged, especially those exercises that build muscle mass and motor skills.
- Coordination may be a problem and make individuals a target for teasing.
- Lack of stamina and energy levels can be problem
- If you live in New England (USA) and qualify, Northeast Passage offers Therapeutic Recreation and Adaptive Sports programming (www.nepassage.org).

What you can do

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- Encourage exercise
- Encourage practice to help build coordination.
- Teach boys who have KS how to pace themselves and learn when to rest.

5. School Absences & Fatigue

What you need to know

- Children with Klinefelter Syndrome should not have excess absences or fatigue.

6. Emergency Planning

What you need to know

- Develop an emergency plan if necessary, depending on the needs of individual children.

7. Resources

The American Association for Klinefelter Syndrome Information and Support

The American Association for Klinefelter Syndrome Information and Support (AAKSIS) is a national volunteer association with the mission of education support, research, and understanding of 47 XXY and its variants, collectively known as Klinefelter syndrome.

<http://www.aaksis.org/>

National Institute of Child Health and Human Development

More in depth condition information for Klinefelter Syndrome is available at the National Institute of Child Health and Human Development

<http://www.nichd.nih.gov/health/topics/klinefelter/conditioninfo/Pages/symptoms.aspx>

Klinefelter Syndrome Article

The cognitive phenotype in Klinefelter syndrome: a review of the literature including genetic and hormonal factors. <http://www.ncbi.nlm.nih.gov/pubmed/20014369>

Klinefelter Syndrome Association UK

The Klinefelter Syndrome Association offers support and information to all affected by, or having an interest in. They have information available which can assist employers, teachers, social workers, and medical professionals. <http://www.ksa-uk.net/>

Klinefelter Syndrome Information and Support

This website offers a wealth of information on Klinefelter syndrome, including current research studies, support group information, informative resources, and symptoms and educational information.

http://klinefeltersyndrome.org/Symptoms_Main_Page.html

Genetics Home Reference

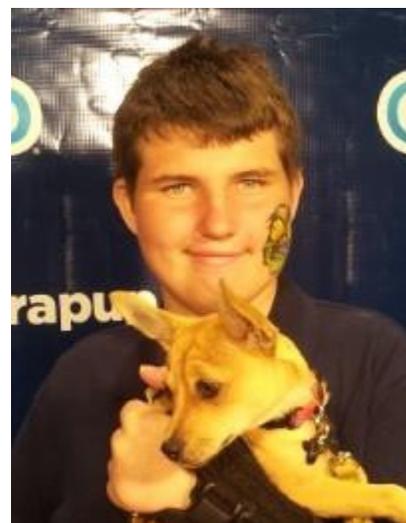
Consumer-friendly information about human genetics from the U.S. National Library of Medicine <https://ghr.nlm.nih.gov/condition/klinefelter-syndrome>

8. Meet a young man with Klinefelter Syndrome

Graham, Mystery Writer

At age 21, Graham loves to spend time on his favorite pastime of writing mysteries and screen plays. The topics vary but they often involve science-fiction. The stories have intrigue- like explorers going into a cave and finding gold but then someone is kidnapped and, and, and....they always have a cliff hanger to leave the reader wondering! He also recently started painting and he is exploring this interest and hopes to continue it as he transitions into the developmental disabilities services in his state.

Graham is outgoing, approachable and not shy about asking questions! He is athletic and likes to play basketball. He has a girlfriend and Lori describes the two as “fantastic friends.” In addition to his artistic endeavors, he loves to play Xbox games and loves social events like parties.



Graham is the 4th out of 5 children and lives in Massachusetts. As a young child he was involved in Early Intervention following his diagnosis at age 2 of Klinefelter (47XXY). His mother advocated for genetic testing when she noticed that he was small for his age, his speech was difficult to understand, and he had some behaviors that she thought were unusual.

Throughout school he has received related services such as Speech and PT. Although his mother wanted him to be included in regular classes, the school system was in favor of him being in special education classes. After much advocating, this year Graham began attending a residential school during the week and lives at home on the weekends. His mother explains that this situation imitates college life in a way.

Lori explains that, like a person on the autism spectrum, Klinefelter Syndrome encompasses a wide variety of abilities and challenges. Some of his challenges include difficulty with executive functions and at times making decisions that are not safe or wise. He has difficulty generalizing concepts. His natural curiosity may sometimes cause him to, for example, run off to explore. Medically, he has had teeth removed and has some joint pain and low tone. Weight control has been an issue as has some difficulty moving his ankle joints through a full range. A decision that might occur in families is whether or not to have testosterone shots.

“He is pretty independent and laid back,” explains Lori. Graham is close to everyone in his family and loves them very much and as a young adult transitioning, he will have lots of support in making choices about the next steps in his life.

Lori’s Advice to Parents:

- Because of the awareness level around autism, it might be helpful to advocate for your child describing this condition as “similar to autism” if it truly is.
- Don’t put limits on your child because of a diagnosis. There is no cap! Look for their strengths and build on those- like Graham’s artistry and writing skills.
- See them as unique and advocate for them.
- Educate yourself. Use the AXYS website located at www.genetic.org.
- Watch for bullying. Because your child may not notice teasing or recognize bullying and be more gullible, make sure to be aware of potential issues.

Lori’s Advice to Teachers:

- Many of the ideas above also apply to school personnel.
- They may have cognitive, language and some behaviors that are similar to students who have autism. You may need to advocate for supports and services that are used for children on the autism spectrum.