



KBG
SYNDROME
ASSOCIATION



KBG SYNDROME SCHOOL GUIDE

NEED TO KNOW INFORMATION
FOR TEACHERS & SCHOOLS

WWW.KBGSYNDROME.ORG

KBG SYNDROME



INTRODUCTION TO KBG

KBG Syndrome is a rare genetic disorder that affects many body systems. It is often characterized in current literature by skeletal abnormalities, distinct facial features and intellectual delays. The specific symptoms experienced may vary from one individual to another. This syndrome can occur spontaneously, or randomly or be inherited by a parent.

[KBG Syndrome is a life changing genetic disorder that may interfere with a student's abilities to perform in the classroom. We hope this guide will help educators to:](#)

- Understand the basics of KBG Syndrome
- Learn recommended educational and classroom modifications that will help to ensure the best learning environment for students with KBG Syndrome

KBG QUICK FACTS

A Rare Genetic Syndrome caused by changes in the ANKRD11 gene. The name "KBG" comes from the initials of the last names of the first three individuals diagnosed.

There are currently under 1,000 known diagnosed cases worldwide. With the advances in technology, more are being found every day.

Individuals with KBG Syndrome are unique! While many share common characteristics, no two are exactly alike.

KBG Syndrome is caused by mutations or loss of material in the gene ANKRD11



Features of KBG Syndrome

Individuals with KBG Syndrome may experience:

Distinctive Facial Features

- Larger Upper teeth (Macrodonia)
- Distinctive facial features with a short, wide skull (brachycephaly)
- Wide Eyebrows that may grow together (synophrys)
- Prominent Nasal Bridge
- Thin Upper Lip
- A longer space between the bridge of the nose and upper lip (long philtrum)

Skeletal Abnormalities

- Cervical Ribs
- Delayed Bone Age
- Curved Pinky Fingers
- Flat Feet
- Short Stature

What Families are Saying

Kids with KBG Syndrome are:

- Very loving
- Funny
- Have the biggest most genuine heart
- Playful and Silly
- Very Brave
- Cute
- Have a great sense of humor
- Kind
- Stubborn or Strong Willed
- Spirited

Individuals with KBG Syndrome are unique! While many share common characteristics, no two are exactly alike.

LIVING WITH KBG SYNDROME

A diagnosis of KBG Syndrome can directly impact future care and quality of life of those it affects. This is why it is important to get an appropriate diagnosis, learn about any recommended treatments or screenings, be prepared for possible surgeries and know when to take precautions.

Even though living with KBG Syndrome requires multiple doctor appointments, regular monitoring, and special tests, individuals with KBG Syndrome can live a long, productive life.



The following health conditions have been reported in individuals with KBG Syndrome

- **Cardiology (Heart)**
 - Congenital Heart Defects: VSD, ASD, Aortic Dilatation
 - Dysautonomia
- **GI**
 - Constipation
 - Feeding Difficulties including the occasional need for feeding tubes or TPN
 - Failure to Thrive
 - Reflux
 - Cyclic Vomiting Syndrome
 - Dysmotility
- **Neurology**
 - Seizures (some cases refractory to treatment)
 - Hydrocephalus
 - Central Apnea
 - Cysts
 - Tremors
 - Hypotonia
- **Sleep**
 - Excessive sleep including Narcolepsy
 - Sleep Disorders
- **Nephrology**
 - Hydronephrosis
- **Eyes**
 - Strabismus
 - Nystagmus
- **Ears**
 - Hearing Loss (All types- Conductive, Mixed, Sensorineural)
 - Recurrent Otitis Media
- **Lungs**
 - Asthma/Reactive Airway
- **Dental**
 - Enamel Hypoplasia
 - Macrodonia
 - Premature teeth loss in adults
- **Endocrinology**
 - Hypothyroidism
 - Adrenal Insufficiency
 - Empty Sella
 - Premature Puberty

*This is not a complete list of all conditions that may affect an individual with KBG Syndrome.

SCHOOL AND KBG SYNDROME

Due to the number of body systems that can be affected by KBG Syndrome there may be barriers to learning and classroom participation. To help offset the difficulties children may face, appropriate interventions are needed.

1. Difficulties that may face students

- Speech Delays/Impairment. Students may require more time or alternative methods of instruction.
- Fatigue- Due to hypotonia or medications
- Impaired vision
- Shortness of breath from heart and/or lung involvement
- Difficulty holding a pencil because of impaired strength or coordination, may be worse as the day progresses—may require physical and/or occupational therapy
- Headaches
- ADD/ADHD. Management of these conditions in KBG children requires the same testing, strategies, and accommodations as for children in the general population.
- Students with KBG syndrome may spend a lot of time with doctors, in hospitals, and getting medical tests. In some cases, they may miss school for surgery, therapies, or other treatments. Some students take this in stride, while others are stressed and frightened.
- Learning Difficulties. Some students with KBG Syndrome may take longer to learn, require additional supports or time. The earlier students receive GOOD intervention that is tailored to their needs the higher the chances they will succeed.



SCHOOL AND MEDICAL CONSIDERATIONS

Due to the variable presentation of individuals with KBG Syndrome these may not apply to all students. This is a general list to consider of medical issues that may be present, the impact on the student and the accommodations that may be utilized in a 504/IEP.

What impact do medical issues have on a student and what are suggested accommodations?

Heart & Blood Vessels

Medical Problems

- Aortic root enlargement
- Congenital Heart Conditions
- Dysautonomia

Impact on Student:

- Restrictions on lifting (backpacks, textbooks, boxes, etc.)
- Fatigue
- Medication regimen may require taking doses at various times throughout the day

Accommodations

- Take all medical complaints seriously
- Provide unlimited access to the nurse's office
- Adjust school schedule to allow for rest during the day while still meeting academic requirements



Bones & Muscles

Medical Problems

- Muscle issues (hypotonia or hypertonia)
- Curved back
- Flat feet
- Loose and flexible joints
- Pain (may be chronic)

Impact on Student:

- May not be able to sit comfortably for long periods of time
- Difficulty walking long distances
- May not be able to participate in regular physical education or field day activities
- Muscle fatigue
- Severely affected students may require use of a wheelchair
- Difficulty with penmanship and/or writing for long periods of time

Accommodations

- May need to provide a special desk/chair
- Allow access to nurse for pain management
- Handwriting modifications or use of other devices
- Wheelchair accessibility if required
- Modify Physical Education Requirements

Eyes

Medical Problems

- Strabismus (misalignment of the eyes)
- Near/far sightedness
- Swollen optic nerves

Impact on Student

- Difficulty reading for long periods of time
- Difficulty seeing the chalk board/smart board/projection screen, etc

Accommodations:

- Provide seating toward the front of the classroom
- Provide computers with settings to accommodate visual disability

Nervous System

Medical Problems

- Seizures
- Headaches/Migraines
- Hydrocephalus
- Tremors

Impact on Student

- May not be able to participate in regular physical education or field day activities
- May have difficulty completing assignments on time
- Needs access to emergency medications at all times

Accommodations

- Provide access to nurse as needed
- Allow for extended time to complete school work
- Adjust school schedule to provide more free time for resting during the day without relaxing academic requirements

Lungs

Medical Problems

- Asthma
- Sleep Apnea

Impact on Student

- May not be able to participate in regular physical education or field day activities

Accommodations:

- Ability to carry inhalers on person or in close proximity
- Needs access to Nurse station at all times

GI System

Medical Problems

- Constipation
- Feeding Difficulties including the occasional need for feeding tubes or TPN
- Failure to Thrive
- Reflux
- Cyclic Vomiting Syndrome
- Dysmotility

Impact on Student

- May need extra time with nurse for feeds
- May need extended lunch time or periods for snacks
- May need alternative bathroom arrangements or accessibility

Accommodations

- Provide access to nurse as needed for meds or feeds
- Allow for alternative lunchroom arrangements, additional time etc
- Allow for private bathroom or changing room as needed

SCHOOL AND SOCIAL CONSIDERATIONS

What impact do social issues have on a student and what are suggested accommodations?

Some students with KBG Syndrome look different from their peers. They may have glasses, hearing aids, feeding tubes or other use of medical devices. They may be quite thin and have difficulty putting on weight or be heavier due to special medications they need for medical conditions.

As a result of these situations a person with KBG Syndrome may be self conscious of how they look or become the target of bullies. Special accommodations for them can sometimes make things more difficult as well.



One of the easiest things you can do to make a lasting impact is to educate classmates about KBG Syndrome. A presentation in class or school wide may be helpful. There are several opportunities throughout the year that are great for this (KBG Days, Rare Disease Day etc) Teachers can try to include the student or family if they wish to.

SCHOOL AND PSYCHOLOGICAL CONSIDERATIONS

What impact do psychological issues have on a student and what are suggested accommodations?

Students with KBG Syndrome have a higher rate of neuropsychiatric disorders. It is important to remember that not all students will be diagnosed with one of these disorders and some are more mildly affected.

These may include: obsessive-compulsive, tic, depressive and attention deficit and hyperactivity disorders, autistic-like features in both children and adults.

Some individuals struggle with anxiety and shyness that may be related to difficulties in understanding social situations. While others are reported to be very outgoing and social with an enhanced ability to make friends. Some may experience mild/moderate difficulties in regulating emotions with outbursts of anger, inconsolable disturbance and occasional aggressive explosions have also been described



The next few pages will detail a few of the more common neuropsychiatric disorders that students with KBG Syndrome may be diagnosed with, the signs/symptoms you may see, impact on the student in the classroom and recommended accommodations.

These disorders are all documented in KBG Syndrome and should be given appropriate consideration and management.

Anxiety

One of the more common struggles for KBG Syndrome students. May present differently from student to student, typically is "silent." Many children struggle internally and do not seek out help. May appear quiet or disengaged while some may "act out" or be considered a "difficult child." With appropriate accommodations and patience children can reduce anxiety and be successful students in the classroom. There is no "one size fixes all" Applying supports to the entire class often helps avoid embarrassment.

Signs:

- Difficulty focusing or mind going blank
- Tantrums or meltdowns
- Excessive worry.
- Fatigue from sleep disturbance
- Irritability (sometimes referred to as "acting out")
- Difficulty separating from care givers
- Avoidance behaviors
- Psychosomatic complaints (headaches, stomachaches, muscle pains, etc.)



Impact on student

- Interfere with social interactions at school
- Negatively affects functioning
- Creates health concerns "worry themselves sick"

Accommodations

- Extra time and warnings before transitions
- Give the child advance notice what they will be reading aloud in class or Encouraging group and interactive work when needing children to read aloud or work math problems on the board
- Extended time for tests
- Breaking down assignments into smaller pieces
- Identify one adult at school to seek help from when feeling anxious (school counselor, if available)
- Buddy system: Pair student with a peer to assist with transitions to lunch and recess (these less structured situations can trigger anxious feelings)
- Help after illness: Missed work can spike anxious feelings. Providing class notes and exempting students from missed homework can help.
- "Cool down passes" to take a break from the classroom. This should be clearly explained to the student. Examples might include a walk down the hallway, getting water, standing outside the classroom door for a few minutes, completing coloring pages in the back of the room, or using a mindfulness app with headphones.
- Preferential group (teacher or adult child knows well) for field trips

Check out www.kbgsyndrome.org for more resources on working with anxiety.

ADD/ADHD

ADD/ADHD develops when there is central nervous system impairment during development. A child may struggle to maintain attention throughout the day or appear excitable or hyperactive. They may also display impulsive behaviors. It is important to manage ADD/ADHD proactively as it can significantly impact learning and socialization as well as lead to other disorders like anxiety.

Signs:

- Being easily distracted
- Impulsive Behaviors
- Trouble paying attention to details or maintaining focus
- Trouble following directions
- Parents and teachers might feel like a child isn't listening to them even when they are speaking to them.
- Talking out of turn
- Poor fine motor skills

Impact on the Student

- Negatively affect educational progress
- Lead to anxiety disorders
- Impact executive functioning
- Difficulties interacting with peers

Accommodations

- Preferential seating. ADHD students should sit in the front row (or close to the teacher), away from windows and other distractions.
- Oral and written instructions. Oral instructions should be reiterated and written down for the student.
- Extended time on tests. Eliminate timed tests and/or provide extended time on tests.
- Supervised organization. Many students with ADHD benefit from a supervised daily cleanout of the backpack and desk to learn organizational strategies. Use of a homework planner checked by the teacher each day and color-coded folders can also be beneficial.
- Manipulatives for understanding. Number lines, math manipulatives, word banks and color-coded spelling words (to help focus on difficult words) are useful for both classwork and tests.
- Provide checklists. Detailed lists for homework assignments, organization, study skills, and checklists of frequently made mistakes help ADHD students stay on task and avoid repetitive mistakes.
- Whiteboard calendar for homework. Using a large, visible calendar to transfer long-term assignments and projects to can enable the student to better track tasks.
- Parent-teacher communication log. A daily communication log helps parents and teachers communicate and work together for the benefit of the student.
- Clutter free workspace. ADHD students need a clutter-free and distraction-free workspace near an adult who can help him/her remained focused and also eliminate screens and other distractions.

Autism

A complex disorder that can be challenging to diagnose. Despite the recommendations and guidelines it can be a complicated process. It is known that Autism tends to affect more boys than girls and can be found in all races and nationalities. It is known that individuals with KBG Syndrome have an increased rate of diagnosis and is being looked at more closely by researchers. There is a spectrum within Autism Disorders and not all students may present in a similar way, the list of signs is not a complete list and any concerns over a possible diagnosis should be discussed with a doctor. Mild cases may be even more difficult to diagnose.

Signs:

- May express repetitive and irrelevant phrases
- Their movement may lack fluidity
- May have fixations
- May seem socially disinterested or lack social perceptions
- Toe Walking
- May have a tendency to be overly emotional and dramatic
- May have a fascination with lights, spinning in circles, and experiencing any number of sensory issues.
- Difficulty recognizing nonverbal cues
- Appears aggressive, passive, or disruptive in a group context
- Failure to initiate or respond to social interactions
- Resists cuddling or other affection
- Specific food preferences
- Behaviors that can potentially result in self harm such as head banging, or biting
- Lack of flexibility: prefers specific routines and exhibits distress at small changes

Impact on the Student

Despite increasing awareness most of the public is still unaware of how autism impacts students. Contrary to belief not all students will avoid eye contact. They can smile, laugh and enjoy interacting with people.

- Impact executive functioning
- Difficulties interacting with peers
- Feeding Difficulties
- Risk taking behavior

Accommodations

- Low-Distraction Work Areas
- Printed Schedules of Activities & Events
- Visual Cues for Common Requests
- Earplugs, especially in hallways, lunch rooms or other noisy environments
- Sensory Retreat Spaces
- Exercise Breaks
- Classroom Aides
- Planning and practicing of communication strategies and social routines
- Using visual markers, tape, etc. to designate boundaries
- Designating a "safe" or "quiet" place in classroom
- Providing task analysis; breaking down goals into small steps
- Using manipulative materials to increase participation in learning experience
- Modifying lessons to emphasize essential concepts for master
- Providing task analysis; breaking down goals into small steps
- Providing peer modeling from socially competent peers





*The best views come after the
hardest climbs.*

Sources:

<https://www.psycom.net/>

KBG Syndrome Publications:

- Obsessive Compulsive Symptoms and Psychopathological Profile in Children and Adolescents with KBG Syndrome
- Intellectual Profiles in KBG-Syndrome: A Wechsler Based Case-Control Study
- Exploring the behavioral and cognitive phenotype of KBG syndrome
- Kbg Syndrome and the Establishment of its Neuropsychological Phenotype
- Clinical and Genetic Aspects of KBG Syndrome
- Mutations in ANKRD11 Cause KBG Syndrome, Characterized by Intellectual Disability, Skeletal Malformations, and Macrodonia
- KBG Syndrome- Gene Reviews
- Novel Mutations and Unreported Clinical Features in KBG Syndrome

To learn more visit us at www.kbgsyndrome.org or email us at kbgsyndrome@gmail.com