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***Toward the cure of
Blue Cone Monochromacy***

BEST PRACTICES FOR SCHOOL INTEGRATION OF STUDENTS WITH BLUE CONE MONOCHROMACY (BCM)

By BCM Families Foundation - October 2015 - last revision July 2020

This document contains notes and advice collected during many years of firsthand experience by men who have BCM and by families with children who have BCM, most of whom live in USA and in Europe. It has been written for teachers of Primary and Secondary/Elementary, Middle and High Schools. We hope this document will help students with BCM by giving their teachers useful information about the visual impairments caused by BCM, plus by recommending proven strategies to better integrate the BCM student.

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“Here is my secret. It is very simple. It is only with the heart that one can see rightly; What is essential is invisible to the eye.”
The Little Prince - Antoine de Saint Exupéry

Blue Cone Monochromacy (BCM) is a rare genetic disease of the retina that affects males almost exclusively. Its prevalence in the general population is 1 in 100,000. There are less than 15,000 people in Europe and the United States who have BCM. It should not be confused with achromatopsia, which is inherited in an autosomal recessive manner and affects 1 in 30,000.

BCM Overview:

- It is hereditary and occurs due to a partial malfunction of the cones of the retina. The disease is a genetic disease linked to the X chromosome;
- It causes loss of visual acuity with myopia (nearsightedness), hemeralopia (blindness from light glare, also called photophobia), colorblindness and infantile nystagmus (shaky eyes) that can last well into adulthood.

To learn more, go to www.bcmfamilies.org

Symptomatology

Reduced visual acuity

Because of the disease, your student who has BCM has reduced visual acuity. Because he is nearsighted, he needs to get very close to see and read writing. When he reads a book, his face will be so close to it that he looks like he is smelling the book - consider this normal for him. Similarly, without a visual aid, he may be unable to see a film or movie in a theater or to follow a lesson when he is too far from the board or teacher. The BCM student will not recognize people from far away and may get lost in unfamiliar locations.

Aversion to light

Light can be very annoying and even blinding for someone who has BCM. In the presence of strong light, a normal-sighted person is not dazzled and blinded by glare; but for the male with BCM, the same light is blinding, like being dazzled by a lighthouse beacon glaring directly into his eyes.

Inability to distinguish colors

For the male with BCM, color vision is severely impaired since birth. The remaining capacity is allowed by the blue cones and rods, so it is difficult for BCM males to distinguish colors. Males who have BCM are monochromats and see in black, white, and shades of gray, although some reduced-color recognition (especially in the blue – green spectrum) is not unusual.

Nystagmus

Some BCM males have involuntary movement of the eye, which increases when the person is stressed or overly-tired. The eye movement can be side-to-side or up-and-down.

STRATEGIES IN SCHOOLS

Poor visual acuity

Visual acuity is the ability to distinguish fine details and see in the distance. It is much reduced in males with BCM and is called 'low vision' in this case.

Visual acuity varies among individuals who have BCM from 20/200 to 20/60. Some BCM males are registered as legally blind. Important: visual acuity can become even more reduced in the presence of strong light.

How to assist your BCM student to function well in class despite his poor acuity

- Assign him sit in the front row, away from strong light. If he is too far from the lesson, he will not see anything, distractions will increase, and he may only be able to use (with great fatigue) his hearing.
- Write on the board (blackboard, whiteboard, PowerPoint presentation, etc.) with large easily recognizable fonts. Use strong contrast between the fonts and the background. The BCM student may have difficulty reading the board even from the front row.
- Students with BCM may be hesitant to continually ask what is written on the board. Therefore, the teacher should monitor the student's ability to see the board and consider providing additional material, for example, handouts with white lettering against a dark background or a device to connect to an electronic white board so the student can sit anywhere in class and still access the information.
- A student with BCM uses great effort to read books with too small lettering so is in danger of fatigue. The reading speed and comprehension of a text may be reduced. To cope with this, choose ebooks or Kindle books used on an iPad or Tablet, so that the font may be enlarged. Also use digitalized dictionaries and encyclopedias. Large print versions of books may be available from other support resources.
- Avoid indicating parts of a page of a book from distance as you teach. Consider providing your BCM student with his own individual copy of any materials/books used in front-of-class demonstrations.
- Large print worksheets, tests and quizzes (magnification 100%), with high quality and high contrast, should be provided for your BCM student. Your BCM student must have his own individual copy and not be expected to share a copy with another student or a group.
- You may need to give your BCM student extra time to do class assignments or exams, tests and quizzes. You should allow additional time during all standardized testing.

- Allow the use of visual aids such as magnifiers. A sloping board/bookstand may also help reduce physical fatigue associated with long periods bent over a desk.
- In primary school, give your BCM student paper that has bigger spaces and darkened lines when he is learning to write. Use a pencil that has dark lead.
- During educational field trips, make sure the BCM student stays with the group and has a companion (“Buddy System”). Allow the BCM student to get as close as possible to exhibitions or activities. Consider providing your BCM student with a cell phone and the teacher’s telephone number on punch-one-number speed-dial in case the BCM student loses sight of the teacher or classmates. For primary school this may or may not be a possibility. A Health and Safety Assessment needs to be carried out prior to the trip.
- Always remember that the student with BCM has difficulty recognizing people at a distance or seeing events or demonstrations at a distance.
- Sports may be challenging for a student who has BCM. He will likely not see a ball, especially if it is small and fast. He may not be able to focus on balls or projectiles while running. Games such as baseball, softball or tennis can be dangerous for those with the BCM since the child may not see the approaching ball. Do not exclude him from games and activities, just adapt them to suit his needs.
- The student with BCM must have the option to substitute safe comfortable activities for those activities that he cannot do safely.
- Due to the frequency of high myopia in BCM males, the risk of retinal detachment is greater than normal. Recognize the need to protect the eyes of your BCM student from impact during Physical Education.

Aversion to light

Light causes glare and almost total loss of vision for your BCM student. Recognize that, because of his disease, your BCM student suffers aversion to light. He will have difficulty outside on sunny days, in a sunny classroom, or too near the window or too near an overhead projector. Some BCM students use colored contact lenses (magenta or brown) inside to protect their eyes from light and to enhance visual acuity. They may often squint or look down to avoid the light.

How to assist your BCM student to function well in class despite his extreme difficulties with glare

- Assign the BCM student a seat away from the classroom window or light sources that are too strong. Request curtains for the windows – specifically, consider black-out curtains. Adjust the lighting of the class so that it is not too bright.
- During school field trips, the BCM student may like to wear dark wrap-around sunglasses and a hat visor.

- During projection or use of overhead projectors, make sure the BCM student is able to follow, and/or provide him a hard copy of what is projected. Prevent him from sitting next to the headlamps' glare.
- The BCM student should be allowed to wear sunglasses at any time he deems it necessary. Ideally, he should keep his sunglasses in his pocket or on a cord around his neck.

Inability to distinguish colors

Males with BCM do not distinguish colors well. It is probable that your BCM student will be unable to do activities requiring color discrimination. In particular, your BCM student will not distinguish red from black.

How to assist your BCM student to function well in class despite his inability to see color

- Check teaching materials, including textbooks, for lessons that require color vision. Compensate in other ways for any information conveyed by color, for example, by giving the BCM student handouts of the same information but using patterns or different styles of fonts instead of colors.
- To test materials to determine whether a student with BCM will be able to follow it, you can make a photocopy in black and white of those pages to see if the meaning can be read the same way.
- Avoid using colored pie charts or colored bar graphs while teaching, or else provide the BCM student with an equivalent pie chart or bar graph that uses lines, dots, patterns or different distinguishable shades of gray instead of color.
- It is impossible for a student with BCM to identify corrections written with red pen on black fonts. Instead, highlight the corrections by circling or squaring any error.
- Sometimes mistakenly a student with BCM may write whole pages with red pen.
- During educational trips, recognize that your BCM student will have difficulty seeing signs (including neon signs) or numbers in the waiting rooms of a station or an airport, as well as signs on bus routes or at bus stops. In fact often these signs, in many countries, are red or orange over a black board. Many BCM students use a mobile phone to photograph and enlarge the signs or to enhance contrast - in this way they claim that they become able to read the signs.
- For younger children, avoid embarrassing your BCM student by asking him to name the colors or requiring him to do tasks in which he must recognize colors. Provide pencils or pens with the name of the color written on them. However, many BCM students enjoy art and coloring, despite their limited capacity for recognizing the colors they are using. Together we need to be conscious that parents and teachers don't end up limiting a child's creativity because he can't see colors the same way others see them.
- During Art and Art History class, assess how art can be seen in black, white and gray.

Nystagmus

The rhythmic involuntary movement and swinging of the pupils is present in many children with BCM. The faster the movement of the eyes the worse the student's vision will be. Stress increases the speed of movement of the eyes. Some people are able to block the nystagmus by seeking a "null point" with a particular inclination of the head. Twisting to "find the null point" may result in torticollis, requiring Physical Therapy.

How to assist your BCM student to function well in class despite his nystagmus

- Be aware that your student needs to manage his level of stress and fatigue.
- Allow rest periods, possibly a few minutes after each task that requires greater effort in visual concentration.
- Using a ruler can help the BCM student with reading, keeping the line of text easily fixed.

Psychological Aspects

At times your BCM student will be teased because he looks different or is not able to do the same things as others. Be aware that some BCM students develop anger/frustration issues tied to social exclusion due to their visual impairment. There is also a possibility that the BCM student may become depressed from being different, unable to do all the things his sighted peers enjoy. In most cases, children with BCM try to mask their disability and try to appear normal, developing their sense of autonomy and compensatory strategies. This costs them a lot of effort, and often there is a sense of fatigue.

How to assist your BCM student to function well in class despite any psychological pressures due to his BCM

- Promote social inclusion, by developing and strengthening autonomy, self-esteem, self-efficacy and full participation in all class activities.
- It is wise to support your BCM student as much, but as subtly, as possible. Integrate any adaptations with as little fanfare as possible.
- Recognize and understand the needs of the child without his having to continually declare that he does not see well.
- Avoid voicing aloud that your student doesn't see well or doesn't see colors. If it is necessary to comment, consider using more positive language. For example, instead of saying "he can't see it from there", instead use "he would benefit from a closer look".
- Try to give him visual aids and learning materials without much class attention, to avoid resentment or ridicule.

- A teenager might be very embarrassed by the teacher telling him in front of his friends to wear his hat or sunglasses or to use visual aids.
- Recognize the moments of fatigue and allow recovery time.
- Some younger BCM students, with the permission and support of their parents, have presented information on their condition to their classmates to help educate and foster further understanding of children with visual impairments.
- Your BCM student needs an Individual Education Plan (IEP).
- Vocational counselling should be appropriately provided to BCM students, to help them identify studies and professions suitable for their skills and abilities to adapt, as well as to make them aware of any financial assistance available to them in reaching their career goals.

Important “Don'ts”

- Don't assume BCM is a deteriorating disease. BCM is a very stable disease, present since birth, not changing over time. Therefore it is not parallel to many other degenerative eye diseases that worsen over time.
- Don't assume your BCM student will use a white cane or Braille. Your BCM student, his parents and his medical advisors may, or may not, choose for him to learn to use a white cane or Braille. Orientation and Mobility Instruction (OMI) may be helpful for a BCM student, assisting him to navigate uneven, unfamiliar terrain without falling plus identifying him as visually impaired to drivers in parking lots and as he crosses the street. Braille may be important to lessen his reading fatigue. However, OMI and Braille are considered optional tools for Blue Cone Monochromacy.
- Also, do not limit your BCM student. Remember that this condition ONLY affects his vision. In every other aspect he is a normal child requiring support, positivity and encouragement during his time in education.

Important “Do”

- Do ask your BCM student directly about what he needs and how to best assist him with any vision challenges he experiences. The child is the expert on BCM.