

Muscular Dystrophy: A Walk in Their Shoes

by Sherri Garcia

Seabury Hall, Makawao, Hawaii

In collaboration with the Centers for Disease Control and Prevention's
Katie Kolor and Aileen Kenneson

This lesson focuses on empathy. Students in the classroom will be exposed to what it feels like to have muscular dystrophy through a modeling activity. They can then gain knowledge about the symptoms, prognosis, and treatment of muscular dystrophy through teacher presentation as well as Internet research. Finally, they explore ethical dilemmas associated with the muscular dystrophy and discuss various hypothetical situations. This lesson would fit best in a biology classroom directly after sex-linked traits are studied.

Disclaimer: The findings and conclusions in this report are those of the author(s) and do not necessarily represent the views of the Centers for Disease Control and Prevention.

Muscular Dystrophy: A Walk in Their Shoes

by Sherri Garcia
Seabury Hall
Makawao, Hawaii

Summary

This lesson focuses on empathy. Students in the classroom will be exposed to one aspect of muscular dystrophy and experience some of what it feels like to have muscular dystrophy through a modeling activity. They can then gain knowledge about symptoms, prognosis, and treatments of muscular dystrophy through teacher presentation as well as Internet research. Finally, they explore ethical dilemmas associated with the muscular dystrophy and discuss various hypothetical situations. This lesson would fit best in a biology classroom directly after sex-linked traits are studied.

Learning Outcomes

- Students will understand the difference between sympathy and empathy and will take part in activities designed to help them develop empathy towards people with muscular dystrophy.
- Students will gain an understanding of the symptoms, prognosis, and treatments of Duchenne/Becker muscular dystrophy.
- Students will develop sensitivity with regard to the social and ethical situations associated with muscular dystrophy.

Materials

1. Wrap around three to five pound leg weights
2. Computer/LCD projector/Flash Player and QuickTime
3. Internet access for students to complete Web Walk
4. Poster board or butcher paper
5. Drawing paper/art supplies

Total Duration

3 hours, 15 minutes

Procedures

Teacher Preparation

The teacher will purchase or prepare leg weights. The teacher might want to review Web sites such as the National Institute of Neurological Disorders and Stroke (NINDS) muscular dystrophy information page and others to become familiar with muscular dystrophy, as well as how muscular dystrophy is inherited. Teacher should also review the PowerPoint slides and accompanying notes. All the student materials should also be downloaded, printed, and copied. Note to teachers: the muscular dystrophies are a group of genetic diseases. One of the most common forms of muscular dystrophy in children is Duchenne muscular dystrophy (DMD). A milder form of the disease is Becker muscular dystrophy, and the combined spectrum of these two types is known as Duchenne/Becker muscular dystrophy (DBMD). Most of the information in this lesson refers to DBMD. For more information about DBMD and other forms of muscular dystrophy, please refer to the Web resources listed below.

Web Resources

Title: Muscular Dystrophy Information Page: National Institute of Neurological Disorders and Stroke

URL: <http://www.ninds.nih.gov/disorders/md/md.htm>

Description: This Web site defines muscular dystrophy and discusses treatments, prognosis, and current research.

Title: Parent Project Muscular Dystrophy

URL: <http://www.parentprojectmd.org/>

Description: This Web site defines muscular dystrophy and discusses treatments, prognosis, and current research. It also discusses educational, emotional, and legislative issues.

Title: Duchenne/Becker Muscular Dystrophy

URL: <http://www.ygyh.org/dmd/whatisit.htm>

Description: This Web site gives an extensive overview on inheritance, symptoms, incidence, testing and screening, cause, and treatments of Duchenne/Becker muscular dystrophy.

Title: Parent Project Muscular Dystrophy: Treatment/Care

URL: http://www.parentprojectmd.org/site/PageServer?pagename=tc_supplements

Description: This Web site discusses the use of prednisone, a catabolic steroid, in the treatment of muscular dystrophy. It also has a section on anabolic steroids.

Introduction

Duration: 15 minutes

This introductory activity should be done at the end of the period, the day prior to beginning the actual lesson.

Organizing Thoughts:

The teacher will ask the students to write down four things they know about muscular dystrophy. Students will then be asked to read what they have written, and the teacher will write the students' statements on the board. Next, the students should write down four things they want to learn about muscular dystrophy and share these things with the class. Finally, the teacher should then ask leading questions to introduce ideas that might not have been suggested.

Walk a Mile in Their Shoes:

Students will then be asked to simulate one aspect of muscular dystrophy by wearing 3- to 5-pound weights on their legs for 24 hours, or until the class meets again the next day. If possible, students should wear the weights for all of their daily activities.

Step 2

Duration: 45 minutes

Following the removal of the leg weights, a discussion should take place at the next class meeting. The teacher should allow each student time to express his or her feelings. How did it feel to have the weights on your legs all day and all night? What things were more difficult? What things were the same? After each student has shared his or her experience, the teacher will give a brief introductory PowerPoint presentation explaining the basic symptoms, prognosis, and treatment of Duchenne muscular dystrophy.

Supplemental Document

Title: Muscular Dystrophy Basics

File Name: Muscular Dystrophy.ppt

Description: This PowerPoint presentation discusses basic symptoms, prognosis, and treatment of Duchenne muscular dystrophy.

Step 3

Duration: 45 minutes

Students will reinforce their understanding of muscular dystrophy by conducting research through a Web walk. The teacher should distribute the “WebWalk” worksheet listed below. The worksheet contains instructions and questions for students to research. Students will use the “Duchenne/Becker Muscular Dystrophy” Web site listed below for the Web walk.

Web Resource

Title: Duchenne/Becker Muscular Dystrophy

URL: <http://www.ygyh.org/dmd/whatisit.htm>

Description: This Web site gives an extensive overview on inheritance, symptoms, incidence, testing and screening, cause, and treatment of Duchenne/Becker muscular dystrophy.

Supplemental Documents

Title: Web walk

File Name: WebWalk.doc

Description: Exploration questions based on the Duchenne/Becker muscular dystrophy Web site.

Title: Answer sheet for the Web walk

File Name: Muscular dystrophy.doc

Description: Answers to the exploration questions based on the Duchenne/Becker muscular dystrophy Web site.

Step 4

Duration: 45 minutes

Disclaimer:

Teachers should note that muscular dystrophy can be a sensitive topic and these discussion questions could bring up difficult issues. The discussion should be conducted in a manner that respects the students' thoughts and views as well as understanding that some students could have family members with this or other conditions. These questions are meant to initiate discussion and do not have any right or wrong answers. These situations are all hypothetical and are not based on real people or events.

Now that students have a clear understanding of muscular dystrophy, the teacher can challenge the students to use the knowledge they have gained from the modeling activity and the research to discuss possible ethical dilemmas. The teacher will read a possible ethical dilemma associated with muscular dystrophy from the document, “Muscular Dystrophy Ethical Dilemmas,” to the students and will begin a discussion about each dilemma. Students can discuss their thoughts about each situation.

These discussions will help students understand the potential complexity associated with muscular dystrophy. They will have an opportunity to explore how they might feel in some of these situations.

Web Resources

Title: Parent Project Muscular Dystrophy-Treatment/Care

URL: http://www.parentprojectmd.org/site/PageServer?pagename=tc_supplements

Description: This Web site discusses the use of prednisone, a catabolic steroid, in treatment for muscular dystrophy. It also has a section on anabolic steroids.

Title: How Steroids Work and Steroid Abuse

URL: <http://www.drugabuse.gov/infofacts/Steroids.html>

Description: This Web site defines and discusses steroid use and abuse.

Supplemental Document

Title: Muscular Dystrophy Ethical Dilemmas

File Name: MuscularDystrophyEthicalDilemmas.doc

Description: List of possible ethical dilemmas associated with muscular dystrophy.

Conclusion:

Duration: 45 minutes

After conducting research through the Web walk and discussing the ethical dilemmas, students should have a better understanding of what muscular dystrophy is and possible challenges faced by families and individuals affected by muscular dystrophy. The lesson concludes by giving students a chance to express empathy.

, Students are asked to express what muscular dystrophy “feels” like by reflecting on knowledge of symptoms and emotions gained during the lesson and creating a drawing. For example, the drawing might reflect physical aspects such as joint pain or the weakening of the heart. It could also be in the form of an abstract piece that reflects emotions and feelings that a person with muscular dystrophy might experience. The drawing should be 11 inches x 14 inches or larger and mounted on foam board or a hard surface. The student should write a paragraph describing the artwork and attach it to the back of the drawing. Any drawing media may be used. Display the art pieces in the classroom or in an area for the entire school to see.

Supplemental Document

Title: Muscular Dystrophy through Art

File Name: Artpiecerubric.xls

Description: This Excel spreadsheet contains a simple rubric for grading the art piece and the paragraph explaining the art piece.

Assessment

Informal Cognitive assessment:

In step 3, students are asked to complete a Web walk using the Web site “Duchenne/Becker muscular dystrophy.” Answers are included in step 3.

Formal assessment: Final assessment is in the form of the art piece prepared by the students. Although this piece is very subjective, a simple rubric is included in the conclusion.

Modifications

Extension

True empathy can be gained by spending more time with people who have muscular dystrophy. Interested students can volunteer to be counselors at Muscular Dystrophy Association (MDA) summer camps. More information is available on the MDA Web site.

Web Resource

Title: Muscular Dystrophy Association

URL: www.mdaua.org

Description: This Web site offers information about the MDA organization, current news and research, special features, and summer camp.

Other Modifications

The teacher might wish to have groups of students prepare PowerPoint slides based on information from the Web walk. For example, one group might prepare a slide on the symptoms while another group prepares a slide on the mode of genetic transmission. The teacher would put the slides together and students would go up and present their slides as they come up. The teacher should be prepared to fill in any missing gaps.

Education Standards

National Science Education Standards

LIFE SCIENCE, CONTENT STANDARD C:

As a result of their activities in grades 9-12, all students should develop understanding of

- The cell
- **Molecular basis of heredity**
- Biological evolution
- Interdependence of organisms
- Matter, energy, and organization in living systems
- Behavior of organisms

SCIENCE IN PERSONAL AND SOCIAL PERSPECTIVES, CONTENT STANDARD F:

As a result of activities in grades 9-12, all students should develop understanding of

- **Personal and community health**
- Population growth
- Natural resources
- Environmental quality
- Natural and human-induced hazards
- Science and technology in local, national, and global challenges

Muscular Dystrophy Web Walk

Muscular Dystrophy
Sherri Garcia, CDC's 2005 Science Ambassador Program

You will be exploring muscular dystrophy using this Web walk. The following Web site offers a wealth of information through interactive animation. Flash Player and QuickTime are required to view this Web site. Please answer the questions using the following Web site: <http://www.ygyh.org/dmd/whatisit.htm>

Part 1: What causes it?

1. What causes the muscle weakness associated with Duchenne muscular dystrophy (DMD)?
2. Where is the genetic change that causes DMD located?
3. How does the loss of an exon affect the production of dystrophin?
4. Without the proper amount of dystrophin in the cell membrane, the cell membrane becomes weakened and ruptures. At this point, describe what happens in the cell and surrounding areas.
5. As people with DMD become older, the number of dying muscle cells rises faster than the repair capacity of the satellite cells. What fills the spaces left by the dying muscle cells?

Part 2: How is it inherited?

1. Why are boys more likely to get DMD than girls?
2. What does the word "carrier" mean?
3. How can a boy get DMD if his mother is not a carrier? How often does this happen?
4. A female carrier of DMD marries a man who does not have DMD. Show the Punnett square for their children. What is the chance that they will have a child with DMD?

Part 3: How is it diagnosed?

Creatine Phosphokinase (CPK) Assay:

1. What does CPK measure?
2. How much higher will CPK be in a boy with DMD compared to a boy without DMD?

DNA Testing:

3. Can a geneticist tell which exon is missing? If so, how? If not, why?
4. What is the final outcome of a PCR test if the exon is present? What if it is not present?
5. A gel can be run on a female to detect the presence or absence of the exon.. How will the band look if the exon is present? How will the band look if the exon is not present?

Muscle Biopsy:

6. What differences do you see between the muscle cells with DMD and those without DMD?

Prenatal Testing:

7. Is it common to screen for DMD in prenatal testing? If so, when is this test performed?

Part 4: How is DMD treated?

1. By what age do most boys with DMD start using a wheelchair?
2. What is a joint contracture?
3. At what time of day would the cardiac muscle and respiratory muscles be most affected?
4. What drugs can be used to slow the progression of DMD? What are some side effects of the drugs?

Part 5: What is it like to have muscular dystrophy?

1. What advice do the parents of children with muscular dystrophy give on answering their sons' questions about DMD?
2. What might be expected with regard to friendships as the condition progresses?
3. What are some issues that might arise at school?

Muscular Dystrophy Web Walk Answers

Muscular Dystrophy
Sherri Garcia, CDC's 2005 Science Ambassador Program

You will be exploring muscular dystrophy using this Web walk. The following Web site offers a wealth of information through interactive animation. Flash Player and Quick Times are required to view this Web site. Please answer the following questions using the following Web site: <http://www.ygyh.org/dmd/whatisit.htm>

Part 1: What causes it?

1. What causes the muscle weakness associated with Duchenne muscular dystrophy (DMD)?

The muscle cells begin to die.

2. Where is the genetic change that causes DMD located?

The gene is located on the X chromosome.

3. How does the loss of an exon affect the production of dystrophin?

A shortened form of dystrophin might result or the dystrophin production may stop completely.

4. Without the proper amount of dystrophin in the cell membrane, the cell membrane becomes weakened and ruptures. At this point, describe what happens in the cell and surrounding areas.

When the cell membrane becomes weakened or ruptures, the molecules can flow in and out of the cell. One of these molecules that can flow in and out is Calcium, which causes the muscle cell to contract near the area where the cell membrane is damaged.

6. As people with DMD become older, the number of dying muscle cells rises faster than the repair capacity of the satellite cells. What fills the spaces left by the dying muscle cells ?

Fat cells and connective tissue begin to fill the empty space.

Part 2: How is it inherited?

1. Why are boys more likely to get DMD than girls?

Boys carry an X and a Y chromosome. The disorder is carried on the X chromosome, which means that boys have only one copy of the gene available. Girls have two copies of the gene (one on each X chromosome). If one gene is affected, girls still have the other gene on the other X chromosome that can be used to make dystrophin.

2. What does the word “carrier” mean?

In general, a carrier is a person who does not have a given disorder; however, he or she can pass the disorder down to his or her children. In the case of X-linked recessive conditions like DBMD, only females are carriers, and may pass the disorder on to male children.

3. How can a boy get DMD if his mother is not a carrier? How often does this happen?

This happens when the gene mutates in the mother’s egg or his early embryonic development. It happens in about 1/3 of the Duchenne cases.

4. A female carrier of DMD marries a man who does not have DMD. Show the Punnett Square for their children. What is the chance that they will have a child with DMD?

| | | |
|---|--------|----|
| | X^D | X |
| X | XX^D | XX |
| Y | X^DY | XY |

The chances are one in four that this couple will have a child, a boy, with DMD.

Part 3: How is it diagnosed?

Creatine Phosphokinase (CPK) Assay:

1. What does CPK measure?

The test measures the enzyme creatine phosphokinase, which has leaked from the damaged muscle cell into the blood stream. People can have elevated CPK levels for many reasons, including DMD.

2. How much higher will CPK be in a boy with DMD compared to a boy without DMD?

A boy with DMD will have levels 50-100 times greater than a boy without DMD.

DNA Testing:

3. Can a geneticist tell which exon is missing? If so, how? If not, why?

Yes, a geneticist can tell which exon is missing. A rapid DNA test can be done. Exons that are present will show up as dark bands. A missing exon will leave a blank in the test.

4. What is the final outcome of a PCR test if the exon is present? What if it is not present?

When the exon is present, the two primers will attach to the exon and replicate. When it is absent, they fail to attach. The DNA will not be replicated if the exon is absent.

5. A gel can be run on a female to detect the presence or absence of the exon. How will the band look if the exon is present? How will the band look if the exon is absent?

If the exon is present, the band will be dark in color. If the exon is absent, the band will be lighter in color. In addition, the PCR machine will produce twice as much DNA from a woman with the exon compared with the woman without the exon.

Muscle Biopsy:

8. What differences do you see between the muscle cells with DMD and those without DMD?

The muscle with DMD shows a wide range of cell sizes and shapes. The image shows fat, connective tissue, and dark colored bodies due to excess calcium ions,. Possibly there are some macrophages engulfing the muscle cells. The muscle cells without DMD are very similar to each other in size and color with consistent cell membranes.

Prenatal Testing:

9. Is it common to screen for DMD in prenatal testing? If screening is performed, when is the test done?

It is not common to screen for DMD prenatally. However, if there is family history of the disease and the DMD mutation is known, then prenatal diagnosis can be done through amniocentesis or CVS screening.

Part 4: How is it treated?

1. By what age do most boys with Duchenne muscular dystrophy start using a wheelchair?

Boys who are not treated with steroids might choose to use a wheelchair by 10 or 11 years of age and almost always use a wheelchair by age 12. Boys that are treated with steroids may be able to avoid wheelchair use until 13 or 14.

2. What is a joint contracture?

A joint contracture is the tightening of the muscles around the joints so that the joint is kept in a semi-flexed position and is unable to extend.

3. At what time of day would the cardiac muscle and respiratory muscles be most affected?

These muscles seem to be most affected during the night. Boys might need a respirator to help them breathe at night and the disease could progress so they would need one all of the time.

4. What drugs can be used to slow the progression of DMD? ? What are some side effects of the drugs?

The steroid prednisone has been used to slow progression of weakness. Some of the side effects include weight gain, mood changes, early puberty, bone brittleness, and cataracts.

Part 5: What is it like to have muscular dystrophy?

1. What advice do the parents of children with muscular dystrophy give on answering their sons' questions about DMD?

Parents suggest that you answer questions honestly, but always keep in mind how much the boy can handle at the time.

2. What might be expected with regard to boy's friendships as the condition progresses?

Invitations to friends' houses might decrease due to fear of how to handle the difficulties associated with the disease. The parents of a child with muscular dystrophy can help with this problem by asking other children over to their house instead.

3. What are some issues that might arise at school?

Classes that involve a great deal of standing, such as chorus, might fatigue the children with muscular dystrophy. The simple task of writing can become difficult and the child's sentences will become shorter. Field trips will need to take into consideration the needs of the child with muscular dystrophy.

Reference:

Cold Spring Harbor Laboratory. Dolan DNA Learning Center: Duchenne/Becker Muscular Dystrophy [online]. 2002. [cited 2005 July 8]. Available from URL: <http://www.ygyh.org/dmd/whatisit.htm>

Muscular Dystrophy Ethical Dilemmas

Muscular Dystrophy
Sherri Garcia, CDC's 2005 Science Ambassador Program

Disclaimer:

Teachers should note that muscular dystrophy can be a sensitive topic and these discussion questions could bring up difficult issues. The discussion should be conducted in a manner that respects the students' thoughts and views as well as understanding that some students could have family members with this or other conditions. These questions are meant to initiate discussion and do not have any right or wrong answers. These situations are all hypothetical and are not based on real people or events.

- 1. The creatine-kinase screening test for muscular dystrophy has just become available for newborn screening in your local hospital. You remember that someone talked about your Great-Uncle Harry dying at an early age from a muscle-related problem. Coincidentally, your older sister just had a baby boy. The hospital has the creatine-kinase test ready to give your new nephew. Do you feel that it would be best for your new nephew to be screened for DMD?**
- 2. Your family just found out that your little brother, who is three years old, has Duchenne muscular dystrophy. Your dad did some research on the Internet and found out that your mom might have passed on a copy of the changed gene that is associated with muscular dystrophy to your brother. He feels that she did something to cause the condition in their child. What do you think about this?**
- 3. You have four brothers, one of whom died at 15 years of age from muscular dystrophy, and three sisters. Your oldest sister was married last year and is now in the kitchen talking to your mother about how wonderful her life was as a child in a large family. She wants to continue the large family tradition. Your mother suggests genetic counseling before pregnancy. How do you feel?**
- 4. James, your best friend in high school, has a little brother with muscular dystrophy. The caregiving responsibilities for James have been divided between members of the family. You win two awesome tickets to a rock concert through a radio contest and want James to go along with you. Unfortunately, you can only give one day's notice and the concert is on the evening that James is scheduled to stay in with his brother. His parents insist that it is his responsibility and everyone else in the family has already made plans to be out. Do you agree with his parents? How do you think James' brother feels?**
- 5. Mary is a stellar student with straight A's across the board. She will most likely be accepted to one of the top tier Ivy Leagues at the end of her senior year. Her younger brother has muscular dystrophy as well as developmental delays. He admires Mary a great deal and asks to be on the electric car team with Mary during her senior year. Mary is embarrassed and considers quitting the team. Do you feel that her action is justified?**
- 6. Joe is only 5 years old. He is starting to notice that he is different than his other friends. He can't run as fast as his friends or pull himself up on the monkey**

bars at school. He has started asking his parents questions. His parents have told him a little about his condition. Joe's father is a matter-of-fact sort of person and has decided to tell Joe the complete prognosis of his condition. Would you do the same?

7. Sandy had an uncle who died of muscular dystrophy when she was a child. After being married for several years, she and her husband decided to start a family. She told him about her uncle and they both decided that she should go through genetic counseling to find out if she is a carrier for DMD. She found out that she is a carrier. She wants to get insurance, but she is afraid that her insurance premium might be higher because of her condition. Would it be fair for her to have a higher premium?

8. Successful gene therapy for Duchenne muscular dystrophy requires the delivery and long-term expression of the dystrophin gene to large numbers of cells throughout the muscle tissue. Once scientists have figured out all the technical difficulties for treating serious disorders like muscular dystrophy, it is possible that they will use the same technique for less serious diseases or even things like raising IQ. Should the scientist move ahead with this cure for DMD?

9. Lance's brother, a muscular dystrophy patient, uses steroids to stabilize the loss of muscle. Lance has a big wrestling match coming up next month and he is considering using his brother's steroids for muscle enhancement. What do you think? (Note to teacher: After students make their decision, you may want to discuss the difference between catabolic steroids, which break down muscle and are used to treat muscular dystrophy, and anabolic steroids used by athletes to build muscle.)

10. You have just started your first real job. You are now an adult; living on your own, doing as you want, and paying taxes! Patients with muscular dystrophy in your state could have a better quality of life if there were a treatment facility. The state does not have funds, but a proposition has been made to add \$100 dollars to every person's state income tax to fund the project. How do you feel about this?

Muscular Dystrophy through Art Rubric

Student Name: _____

| CATEGORY | 4 | 3 | 2 | 1 |
|--------------------------|--|--|---|---|
| Writing Process | Student devotes a lot of time and effort to the writing process (prewriting, drafting, reviewing, and editing). Student works hard to express the feelings in the art piece. | Student devotes sufficient time and effort to the writing process (prewriting, drafting, reviewing, and editing). Student works and gets the job done. | Student devotes some time and effort to the writing process but it is not very thorough. Student does enough to get by. | Student devotes little time and effort to the writing process. Student does not seem to care. |
| Spelling and Punctuation | There are no spelling or punctuation errors in the final draft. | There is one spelling or punctuation error in the final draft. | There are 2-3 spelling and punctuation errors in the final draft. | The final draft has more than 3 spelling and punctuation errors. |
| Focus on Assigned Topic | The entire writing is related to the assigned topic and allows the reader to understand much more about the topic. | Most of the writing is related to the assigned topic. The story wanders off at one point, but the reader can still learn something about the topic. | Some of the writing is related to the assigned topic, but a reader would not learn much about the topic. | No attempt has been made to relate the writing to the assigned topic. |
| Illustrations | The art piece is detailed and creative. It relates to the topic. | The art piece is somewhat detailed. It relates to the topic. | The art piece relates to the topic. | The art piece does not relate to the topic. |