Need-to-Know Information for the

TEACHER

Raising awareness of Marfan syndrome and related conditions

THE MARFAN FOUNDATION

CONTENTS

1 INTRODUCTION

Marfan syndrome is a life-threatening genetic condition that causes some physical problems that can interfere with a student's ability to perform in the classroom without modifications. These physical limitations cause many students living with the condition to feel isolated and alone. We hope this guide enables educators to:

- Understand the basics of Marfan syndrome and its impact on a student
- Learn recommended educational and classroom accommodations to ensure the best learning environment for students with Marfan syndrome
- Create a supportive classroom environment for students with Marfan syndrome
- Identify ways to incorporate Marfan syndrome and/or genetic diversity into lesson plans

Many students with Marfan syndrome suffer from low self-esteem because they look different and are unable to participate in many of the activities of their peers, reinforcing intense feelings of isolation. Physical limitations usually mean that physical education requirements need to be modified.

What is Marfan syndrome?

Marfan syndrome is a life-threatening genetic condition of the body's connective tissue. Knowing the signs of Marfan syndrome, getting a proper diagnosis, and receiving the necessary treatment can enable people with Marfan syndrome to live a long and full life.

Our community of experts estimates that nearly half of the people who have Marfan syndrome don't know it. Without proper diagnosis and treatment, they are at high risk for an early sudden death. Marfan syndrome affects our connective tissue, which helps to hold the body's cells and tissues together. It also regulates how our bodies grow.

There are also several conditions related to Marfan syndrome that cause people to struggle with the same or similar physical problems, and anyone affected by these conditions also needs an early and accurate diagnosis.

What are the features of Marfan syndrome?

Some features of Marfan syndrome are easier to see than others. These include:

- Long arms, legs, and fingers
- Tall and thin body type
- Curved spine
- Sunken or protruding chest
- Flexible joints
- Flat feet
- Crowded teeth
- Unexplained stretch marks on the skin

Harder-to-detect signs include:

- Heart problems, especially related to the aorta, the large blood vessel that carries blood away from the heart

Other signs include:

- Sudden collapse of a lung
- Eye problems, including severe nearsightedness, dislocated lens, detached retina, early glaucoma, and early cataracts.

What causes Marfan syndrome?

Marfan syndrome is caused by a change (mutation) in the gene that tells the body how to make fibrillin-1, a protein that is an important part of connective tissue. This mutation creates different Marfan syndrome features and causes medical problems.

Who has Marfan syndrome?

About 1 in 5,000 people have Marfan syndrome. This includes men and women of all races and ethnic groups. People can inherit Marfan syndrome; that is, they get the mutation from a parent who has Marfan syndrome. This happens in about 3 out of 4 people with Marfan syndrome. Other people have a spontaneous mutation, meaning that they are the first in their family to have Marfan syndrome. People with Marfan syndrome have a 50 percent chance of passing the mutation on each time they have a child.

People are born with Marfan syndrome, but they may not notice any features until later in life. However, Marfan syndrome features can appear at any age, including in infants and young children. Marfan syndrome features and medical problems can get worse as people age.

How is Marfan syndrome diagnosed?

A Marfan syndrome diagnosis can often be made after exams of several parts of the body by doctors experienced with connective tissue conditions, including:

- A detailed medical and family history, including information about any family member who may have the condition or who had an early, unexplained, heart-related death.
- A complete physical examination and tests to identify Marfan features that are not visible during the physical exam.

Some of the features of Marfan syndrome can be found in conditions related to Marfan syndrome; therefore, genetic testing may be helpful when a diagnosis cannot be determined through an exam by doctors.

It is possible for someone to have one or more features of Marfan syndrome, but not enough to have a Marfan syndrome diagnosis. Additional exams by other doctors and genetic testing help determine if the diagnosis is related to Marfan syndrome.

What is life like for someone with Marfan syndrome?

Advances in medical care help people live longer and enjoy a good quality of life if they are diagnosed and treated. Most people with Marfan syndrome can work, go to school, and enjoy active hobbies. It is very important that people with Marfan syndrome get treatment and follow medical advice; otherwise, heart problems can cause sudden death. With an early diagnosis, helpful medical treatment can begin early in life. People with Marfan syndrome also need to adapt their physical activity to stay safe. In general, they should not play competitive team sports such as football, soccer, or basketball. In addition, they should not lift heavy objects when at work, home, or the gym.

What emergencies could arise for someone with Marfan syndrome?

People with Marfan syndrome are at increased risk for emergencies involving the aorta, eyes, and lungs. These include:

Aortic Dissection

Aortic dissection is a tear between the layers of the aorta, the large blood vessel that carries blood away from the heart. An aortic dissection can be fatal if not treated immediately. Emergency care is needed immediately. Aortic dissection is rare in children with Marfan syndrome, but symptoms include:

- Pain in the center of their chest, abdomen (stomach), or back; may be "severe," "sharp," "tearing," or "ripping" and may travel from the chest to the back and/or abdomen. Sometimes, the pain is less severe, but a person still has a feeling that "something is very wrong."
- Nausea
- Shortness of breath
- Fainting
- Loss of pulse
- Tickling, numbness, burning, prickling of the skin (parasthesia)
- Paralysis

Collapsed lung

Collapsed lung happens when air or gas collects in the space between the lungs and the chest and prevents the lung from inflating completely, symptoms include:

- Chest pain, sudden onset; sharp and may lead to feelings of tightness in the chest
- Shortness of breath
- Rapid heart rate
- Rapid breathing
- Cough
- Fatigue
- Skin may develop bluish color (cyanosis) due to decreased blood oxygen level

Detached Retina

A detached retina is a separation of the light-sensitive membrane in the back of the eye (the retina) from its supporting layers. Retinal detachment can cause permanent, severe vision loss or blindness if it's not treated. Symptoms include:

- Translucent specks of various size, shape and consistency in the eye
- Bright flashes of light, especially in the peripheral vision
- Blurred vision
- Shadow or blindness in a part of the visual field of one eye

What conditions are related to Marfan syndrome?

There are several conditions related to Marfan syndrome that cause people to struggle with the same or similar physical problems, and anyone affected by these conditions also needs an early and accurate diagnosis.

Related conditions include:

- **Familial thoracic aortic aneurysm and dissection (FTAAD).** Those affected have only one Marfan syndrome feature—aortic dilation—and are at risk for sudden death from aortic dissection and should follow Marfan syndrome heart care.
- **MASS Phenotype.** People with this condition have Marfan syndrome features of mitral valve prolapse, mild aortic dilation, skeletal features, and skin stretch marks. They should have an echocardiogram every 1–2 years until growth is completed and at least every 5 years thereafter.
- **Ectopia lentis syndrome** (dislocated lenses of the eye). People with this condition have dislocated eye lenses and Marfan syndrome skeletal features. Most people with this condition have a normal-sized aorta with a risk for developing aortic dilation later in life. Therefore, they should have an echocardiogram every few years.
- **Beals syndrome.** People with Beals syndrome have many skeletal features of Marfan syndrome. They may also have joint contractures (cannot fully stretch their knees, elbows, and other joints), oddly shaped ears, and aortic dilation. Those with aortic dilation should have an echocardiogram once a year.
- **Loeys-Dietz syndrome.** People with Loeys-Dietz have some skeletal features of Marfan syndrome and aneurysms in the aorta and other arteries. They also have features not seen in Marfan syndrome including a twisting aorta, widelyspaced eyes, and a bifid uvula (a split in the tissue that hangs at the back of the throat). Based on aortic size, they need heart surgery sooner than people with Marfan syndrome.
- **Vascular Ehlers Danlos syndrome (VEDS).** VEDS is a genetic connective tissue condition. It can affect many different parts of the body, including the arteries, hollow organs, skin, and lungs. These systems can be prone to tear, which can be life-threatening.
- **Other Ehlers-Danlos syndromes.** A group of genetic connective tissue conditions characterized by unstable, hyper-mobile joints; loose, “stretchy” skin; and tissue fragility.

Many of the related conditions are also genetic conditions that cause the aorta to enlarge, which requires regular care. Advances in diagnoses, treatments, care, and research about Marfan syndrome will likely advance the diagnoses, treatments, care, and research about related conditions—and vice versa.

This resource will focus primarily on Marfan syndrome, but the issues of management of many of the related conditions included here are similar.

Further, since the diagnosis of the condition is so complex, and some features become more pronounced with time, a younger student may not have enough apparent features yet to be formally diagnosed with Marfan syndrome. This concept is some-times referred to as “emerging Marfan syndrome.” It is prudent to follow over time a student who has some Marfan features, but who has not been diagnosed.

2 SPECIAL CONSIDERATION FOR STUDENTS LIVING WITH MARFAN SYNDROME

Barriers to learning and classroom participation arise from the multiple body systems affected by Marfan syndrome. Appropriate interventions are needed according to the difficulties each child may face. These include:

- Low vision, mild to severe—may require teacher for the visually impaired (TVI) and orientation and mobility (O&M) specialist
- Fatigue (from medications)
- Shortness of breath from heart and/or lung involvement
- Difficulty holding a pencil because of loose hand ligaments, may be worse as the day progresses—may require physical and/or occupational therapy
- Chronic pain (particularly back and joint pain)
- Headaches
- Physical activity places great strain on the heart and blood vessels; therefore, students with Marfan syndrome should not participate in certain competitive or contact sports and physical education teachers should consult with a child's physician on a customized plan using The Marfan Foundation's physical activity guidelines as a resource.
- Restrictions on the amount of weight to be lifted. Management may require a second set of books to be left at home.
- Some doctors feel that there is a higher rate of ADD or ADHD in children with Marfan syndrome. Management of these conditions in Marfan children requires the same testing, strategies, and accommodations as for children in the general population.

Students with Marfan syndrome usually spend a lot of time with doctors, in hospitals, and getting medical tests. In some cases, they may miss school for surgery, physical rehabilitation, or other treatments. Some students take this in stride, while others are stressed and frightened.

What are Individualized Education Plans (IEP) & 504 Plans?

Marfan syndrome does not cause cognitive disabilities, but some students experience learning disabilities, emotional trauma, and mental instability secondary to the condition. These effects can relate directly to dealing with difficult physical traits, operations, and pain, or they may occur as side effects of medication.

Therefore, a student with Marfan syndrome may require an Individualized Education Plan (IEP) or a 504 Plan.

An Individualized Education Plan (IEP) is required by the Individuals with Disabilities Education Act (IDEA) so that eligible students can receive special education and related services. It requires a thorough evaluation, provides the widest range of services and accommodations, and entitles parents to be active participants. However, the eligibility requirements are many and, in some cases, carries stigma because of the special education component.

The 504 Plan was created by Section 504 of the Rehabilitation Act of 1973 for students who require school accommodations, but who are not eligible for special education. It is easier to qualify for a 504 than an IEP, but the 504 offers fewer services. There are also fewer legal protections to ensure compliance. Unlike the IEP, a 504 Plan may be adopted and changed with little to no parental involvement.

Both types of plans may include accommodations ranging from an extra set of textbooks to leave at home to wheelchair ramps. They should be reviewed and updated annually.

The purposes of instituting an IEP or 504 Plan are to:

- Enable the student to be independent in school
- Support the student's acceptance and self-esteem
- Allow the student to adapt and cope to school within his/her limitations
- Keep the student safe from predictable physical injury
- Facilitate communication between the student, parents/guardians and school staff about health needs and accommodations in school

Teachers, administrators, and parents should work together as a team to determine which plan is better for a student and work together to develop it.

The plan works best when each member of the team includes all other members in general correspondence. However, any member can update parents about the student without including other team members. This is to allow for candid, open, and unfiltered communication about the student as often as possible. Team meetings should be held as often as necessary in response to any changes in the student's condition or specific events at the school.

Your school district should have readily available a template for developing an IEP or a 504 Plan. There are many common issues that affect students with Marfan syndrome that should be considered in the development of either type of plan.

What are Individualized Health Plans (IHP) & Emergency Care Plans (ECP)?

In addition to an Individualized Education Plan (IEP) or 504 Plan, a student with Marfan syndrome should have an Individualized Health Plan (IHP) and an Emergency Care Plan (ECP), coordinated by the school nurse. Classroom teachers should be aware of these plans, as well.

The Individualized Health Plan (IHP) outlines the healthcare services a student needs during the school day. Prepared by the school nurse in collaboration with the student, family, teachers, school officials, and healthcare providers, it includes a thorough assessment of the student's physical and mental health and educational activity, along with guidance for school personnel. It should be reviewed and revised as needed at least once a year.

The Emergency Care Plan (ECP) is a shorter document (typically one page) that contains information about the student's condition and provides contact information for family, physicians, hospitals, and an ambulance service in the event of a medical emergency. Teachers, as well as the school nurse, principal, and other adults in the student's life should have a copy of it readily available.

Note: Excessive activity increases heart rate and blood pressure increases stress on the heart and blood vessels. This contributes to enlargement of the aortic root, the most serious and life-threatening complication of Marfan syndrome. Aortic emergencies are relatively rare in school-aged students, but chest pain should always be taken seriously.

What is the impact of medical problems on the student and what are the suggested accommodations?

Overall, students with Marfan syndrome may require a modified curriculum and/or alternative to physical education class. It is also important for school officials and teachers to understand and tolerate extended absence for medical treatments and/or surgery. Collaboration with the entire educational team and family is important to ensure the student does not fall behind. Here is a breakdown of the different parts of the body affected by Marfan syndrome and suggested accommodations.

Heart and blood vessels

Medical problems

- Irregular/erratic heartbeat
- Mitral valve prolapse
- Aortic root enlargement/aortic aneurysm
- Aortic dissection (relatively rare in school-aged students, but it is a medical emergency and should be taken seriously if symptoms occur)

Impact on the Student

- Fatigue
- Restrictions on lifting (backpacks, textbooks, boxes, etc.)
- Restriction on activities, i.e., those that can increase heart rate or raise blood pressure
- 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
- Adjust class schedule and/or group classes together to limit movement between classes
- Provide additional time to get to classes
- Provide extra set of books for home use and/or second locker to accommodate lifting/carrying restrictions
- Modify physical education curriculum or offer alternative to physical education class

Bones and joints

Medical Problems

- Tall stature, with disproportionately long arms and legs (students may be significantly taller than their peers)
- Loose and flexible joints
- Underdeveloped muscles
- Pain (often chronic)
- Chest bone that sinks in or sticks out
- Curved back
- Flat feet

Impact on the Student

- Difficulty fitting in standard size desks/chairs
- 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
- Inability to carry heavy books long distances
- Difficulty with penmanship and/or writing for long periods of time
- Prone to joint injuries
- Muscle fatigue
- Body image issues
- Severely affected students may require use of a wheelchair (rare)

Accommodations

- Provide a special desk and/or chair
- Allow student to stand if unable to sit comfortably
- Provide additional time to get to classes
- Schedule classes in rooms near each other
- Assign homeroom and locker near to classes students will attend; alternatively, assign a second locker in another part of the building
- Allow access to nurse for pain management, as necessary
- Modify curriculum or offer alternative to physical education class
- Provide an extra set of books at home and a set of books in each classroom in order to cut down on the student having to carry books for several classes
- Grade handwriting with leniency

- Provide additional time for handwritten tests
- Offer options for handwritten assignments, such as an aide who can write for the student or a laptop or tablet
- Wheelchair accessibility if required
- Provide a separate changing room for physical education class

Eyes

Medical Problems

- Severe near-sightedness
- Dislocated lens
- Risk of detached retinas (less common, but should be considered an emergency if it occurs)

Impact on the Student

- Vision may fluctuate
- Difficulty reading for long periods of time
- Difficulty reading small or light colored fonts
- Difficulty seeing the chalk board/smart board/projection screen, etc.

Accommodations

- Provide large print books
- Use a dark, clear font on school materials (homework assignments, tests, etc.)
- Provide seating in the front of the classroom
- Provide computers with settings to accommodate visual disability

Lungs

Medical Problems

- Asthma
- Sleep apnea
- Collapsed lung; this should be treated as a medical emergency
- Indented chest bone and curved back can reduce lung function, causing shortness of breath and fatigue

Impact on the Student

- May need additional time to get from class to class
- May not be able to participate in regular physical education or field day activities
- May experience mental or physical fatigue

Accommodations

- Provide additional time to get to classes, and schedule classes in nearby classrooms
- Allow access to nurse for medication or rest as necessary
- Provide access to emergency inhaler if needed for asthma
- Modify physical education class or provide alternative to physical education class
- Adjust school schedule to allow for more free time for resting without relaxing academic requirements

Nervous System

Medical Problems

- Dural ectasia (widening or ballooning of the dural sac surrounding the spinal cord), which can cause headaches and back, abdominal, or leg pain

Impact on the Student

- Pain (often chronic) may interfere with ability to focus or sit for long periods of time
- May not be able to participate in regular physical education or field day activities
- May have difficulty completing assignments on time

Accommodations

- Provide access to nurse as needed
- Adjust school schedule to provide more free time for resting during the day without relaxing academic requirements
- Allow for extended time to complete school work
- Allow student to take medication or take any other prescribed steps to relieve discomfort

What social and psychological problems do students with Marfan syndrome face?

Students with Marfan syndrome often look different from their peers. They are often taller than average and very skinny and lanky. They may wear thick glasses or have stretch marks. They may have a chest that sinks in or sticks out. They may have a curved spine that requires a back brace and foot problems that require orthotics.

In addition, students with Marfan syndrome are frequently quite thin and are unable to put on weight. This sometimes leads to suspicion of an eating condition. However, no matter how much a person with Marfan syndrome eats, they may remain thin throughout childhood.

As a result of these physical differences, students with Marfan syndrome may be highly self-conscious and become the target of bullies. Special accommodations made for them sometimes make the situation even more challenging.

Educating classmates about Marfan syndrome can reduce the stigma and prevent bullying. A classroom or school-wide presentation about Marfan syndrome can be helpful. Teachers can try to include the student with Marfan syndrome, as well as his or her family, in the presentation.

3 PHYSICAL EDUCATION & ACTIVITY GUIDELINES

Regular exercise improves both physical and emotional well-being and can be incorporated safely into the routine of students with Marfan syndrome. A student with Marfan syndrome should have a physical education and activity program that is individualized to ensure safety.

Physical activity should be modified to help eliminate undue stress on the aorta, avoid chest or eye trauma, and avoid potential damage to loose ligaments or joints.

The goal of physical activity guidelines is to help students achieve the benefits of safe levels of exercise and, at the same time, ensure that they don't add to medical problems related to Marfan syndrome.

What should I know about physical activity for students with Marfan syndrome?

In general, most students living with Marfan syndrome should exercise regularly through low-intensity, low-impact activities adapted to meet their specific needs. They should avoid contact sports because of the risk of damaging the aorta and injuring the eyes. Strenuous activities, such as competitive sports and weightlifting, also should be avoided because of the stress placed on the aorta.

However, it's important to keep in mind that every activity can be done at different intensity levels, and no recommendation holds true in all circumstances. For example, shooting baskets in the driveway is different from playing a full-court basketball game, and bicycling ten miles in one hour on a level course is different from competing in a triathlon.

It is essential for each parent of a student with Marfan syndrome to discuss physical activities and specific activity levels with their student's physician so that exercise can be incorporated safely into physical education at school and in their regular healthcare routine.

What if Marfan syndrome is not formally diagnosed but suspected?

Sometimes Marfan syndrome or a related condition is suspected, but has not been firmly diagnosed. In other cases, a diagnosis of Marfan syndrome has been made, but the individual currently doesn't have aortic enlargement. In these instances, determining whether or not to follow the physical activity guidelines is particularly confusing.

Several factors are taken into account in determining which activities are safe and which are not. These are: how strongly a diagnosis is suspected; whether or not there is family history of Marfan syndrome/related condition or a family history of early cardiac death; the age of the person; and level of activity planned. The individual's particular eye, skeletal, heart, aortic, and lung condition are important to consider when deciding on safe levels of physical activity.

It is best for parents to speak with their child's cardiologist (heart doctor), medical geneticist, or specific medical specialist to determine what is considered safe.

What are the different types of exercise and competition?

Exercise can be classified by several characteristics.

- Aerobic activities are conducted at an intensity that permits oxygen to be used to generate energy. There is a balance between the needs of the muscles and the ability of the body to provide oxygen to the muscles. If the person exercising can carry on a conversation while exercising, it is at an aerobic level.
- In an anaerobic activity, there is insufficient oxygen and cells have to rely on internal sources, which become depleted quickly, leading to fatigue. Anaerobic activity is usually of higher intensity, and is thus more stressful to tissues and the cardiovascular system.
- Isokinetic exercise is when a muscle contracts through much of its full range of motion, such as the arm muscles when throwing a ball and the leg muscles when running.
- Isometric exercise is when a muscle is contracting without moving, such as when straining to lift a heavy weight or pushing a heavy piece of furniture. An increase in blood pressure, which stresses the heart and aorta, is greater with isometric exercise.

Most exercises and athletic activities involve a combination of isokinetic and isometric muscle work and aerobic and anaerobic energy use. The proportion of work and energy is determined by the nature of the activity, how strenuously a person is participating and, in team sports, even the position being played. Sports are classified based on the risk of collision (contact) and how strenuous they are.

What are the classifications of sports and activities?

The following table is modified from a classification devised by the American Academy of Pediatrics. Please note that many sports can fall within several categories, depending on the intensity of participation. It is essential that parents talk to their student's doctor about the sports and activities that are safe, and how to monitor exertion levels so that exercise remains safe throughout involvement.

To maximize safety of low intensity, non-contact activities, it is important to take necessary precautions, such as not carrying a heavy bag of golf clubs and avoiding intense competitive efforts.

Contact/collision high potential: Strenuous	Basketball, boxing, field hockey, football, ice hockey, lacrosse, martial arts, rodeo, skiing (water), soccer, wrestling
Contact limited: Strenuous	Baseball, bicycling (intense), gymnastics, horseback riding, skating (ice & roller), skiing (downhill & cross-country), softball, squash, volleyball
Noncontact: Strenuous	Aerobic dancing (high impact), crew, running (fast), weightlifting
Noncontact: Moderately strenuous	Aerobic dancing (low impact), badminton, bicycling (leisurely), jogging, swimming (leisurely), table tennis, tennis
Noncontact: Non-strenuous	Golf, bowling, walking

How does a student's medication impact physical activity?

Before beginning or increasing any exercise program, it is important for the student's doctor to assess the student's current level of physical fitness, health, and medications. The advice offered here is general, and is not meant to substitute for the recommendations of the student's personal physician.

Many students with Marfan syndrome take a beta-blocker medication to reduce stress on the aorta. This medication lowers the pulse at rest and during exercise, and makes it somewhat more difficult to achieve a given level of physical fitness for the amount of physical work performed.

They do not, however, allow a person with Marfan syndrome or other aortic aneurysm syndrome to perform very strenuous exercises or play contact sports. Some patients with Marfan syndrome take medications called angiotensin receptor blockers (like losartan) or angiotensin converting enzyme inhibitors. These medications do not protect the aorta from strenuous exercise.

Students who have artificial heart valves usually take an anticoagulant medication, warfarin (Coumadin®). This medication interferes with blood clotting and increases the chances of bruising and internal hemorrhages. Students taking this medication should avoid contact sports and any activity with a moderate risk of a blow to the head or abdomen.

What are some guidelines and modifications that permit safer exercise for students with Marfan syndrome?

Physical activity modifications for students with Marfan syndrome include the following:

- Favor non-competitive, isokinetic activity performed at a non-strenuous aerobic pace. Especially suited are sports in which the student is free to rest whenever he/she feels tired and in which there is a minimal chance of sudden stops, rapid changes in direction, or contact with other players, equipment, or the ground. Some beneficial activities are brisk walking, leisurely bicycling, slow jogging, shooting baskets, slow-paced tennis, and use of 1-3 pound hand weights.
- Choose an activity the student can enjoy that can be performed three or four times per week for 20–30 minutes. If time is a major constraint, three 10-minute sessions are nearly as effective as one 30-minute session.
- Stay at an aerobic level of work (about 50 percent of capacity). If the student is on a beta-blocker, he/she should keep a pulse under 100 beats per minute. If the student is not on a beta-blocker, keep a pulse at less than 110.
- Avoid activities that involve isometric work, such as weightlifting, climbing steep inclines, and doing pull-ups. When using a stationary cycle or a step-climber, keep the tension low. Multiple repetitions with low resistance or low weight are safer than a few repetitions with a larger weight.
- Do not test limits. This is particularly difficult for students during physical fitness tests in school and for students who once were competitive athletes.
- Wear protective gear. For example, high-quality helmets should always be worn while bicycling.

How can parents and teachers guide a child to safe physical activity and exercise?

Adults who are newly diagnosed are usually able to reconcile the need to modify their exercise; however, modifying activity is a greater concern to parents who have a child who is newly diagnosed.

Sports are a big part of childhood in many families. Being part of a team helps develop social skills and self-esteem. It is understandably frustrating or upsetting to children who suddenly have physical activity restrictions (and for their parents), particularly if the child already has a passion or talent for a particular sport.

The general guidelines for students with Marfan syndrome are to avoid competitive and contact sports that would put added stress on the aorta, cause chest or eye trauma, or be potentially damaging to loose ligaments and joints. However, there are also concerns that go beyond the potential physical dangers.

Consider youth soccer, which is not an intensely competitive sport; it's more recreational and is not regarded as dangerous for children with Marfan syndrome because aortic dissection in a young child with Marfan syndrome is very rare. However, youth soccer leads to more competitive soccer in middle school and beyond. Asking a child to give up a sport after he or she has been involved for many years impacts their social circle and their self-esteem, and removes from their life an activity for which they have developed a passion and talent.

When children are diagnosed at a very young age, parents and teachers are encouraged to provide guidance for activities that are appropriate for the long-term. Golf, bowling, archery, piano, art, and music are just a few alternatives that can provide an outlet for creativity and competition while still providing the interaction and socialization a student needs.

When a diagnosis is made when someone is on an athletic scholarship in college, the new physical activity restrictions can be particularly devastating and life-changing. Yet, the alternative can be deadly.

If the student is having difficulty adjusting to the restrictions or has become depressed about necessary lifestyle changes, speaking with a therapist may be helpful. The Marfan Foundation also offers opportunities for parents of affected children to speak with other parents with in-person and online support groups. We also offer specialized programs for children, teens, and young adults at our annual family conference.

What are the suggested accommodations for students with Marfan syndrome?

To ensure the safest environment for a student with Marfan syndrome that allows them to participate to the greatest/safest extent possible, the physical education teacher should partner with the student's medical team (especially, their heart doctor) and parents.

It is not possible to create a single exercise program that is safe for all students who have Marfan syndrome. Each student's physician should provide the physical education teacher with information about safe activity levels for that student. The physical education teacher can then design activities within these levels. It may be helpful for the physical education teacher to provide the physician with a list of planned activities in the physical education curriculum.

In addition, a student may need assistance in developing a realistic self-concept of abilities and limitations. It is important to recognize that the student's level of comprehension can be misjudged, especially because many students with Marfan syndrome are treated as if they are older because of their increased height.

General accommodations and suggestions

- Encourage the concept of the "personal best" to minimize competition between students and limit peer pressure • Instruct the student in safe levels of intensity and duration
- Be receptive to a student with Marfan syndrome who reports certain symptoms, such as chest pain and difficulty breathing
- Provide adequate time for gradual warm-up and cool-down
- Monitor the student's level of exertion more closely in extreme weather conditions because heat and cold may add additional stress and may affect the student's endurance and exertion level
- A child with Marfan syndrome should be encouraged to take part in noncompetitive activities performed at a mild to moderate level, e.g., they aren't out of breath and their pulse stays at a certain level

Physical education teachers who are monitoring the exertion level of a student with Marfan syndrome should be aware of the student's medication. Some medications slow the heart rate and therefore measuring heart rate is not a true indicator of exertion level.

Students with Marfan syndrome often have muscular underdevelopment and joint laxity or tightness and may lack bulk and muscle tone.

Muscle strengthening can be helpful for these students. Focus on activities that increase strength of both muscles and ligaments. However, the student should only do exercises with a weight that enables 15-20 repetitions comfortably. Avoid activities involving heavy weights or intense isometric exertion. The student may be doing physical therapy outside of school; ask parents about coordinating with the student's physical therapist so that in-school physical education can complement it.

Modifications for required equipment

- If the student needs a brace during sports activities, his or her maneuverability, flexibility, speed, and endurance may be affected. The physical education teacher should be informed by an orthopedist what restrictions the brace creates. When a back or body brace is worn, the head and neck should be protected during physical activity by suitable padding.
- If the student has had chest wall surgery, the surgeon will need to inform the physical education teacher about any further restrictions that are necessary.
- If mouth guards are required for an activity, the student may need custom mouth guards to accommodate the narrow mouth.
- If hernias are present, the student may need to wear a supportive truss and the student will need instruction in proper lifting techniques. The physical education program may need to minimize activities that involve lifting or climbing.

Addressing fatigue issues

- Decreasing duration of an activity
- Decreasing size of playing area
- Allowing frequent “time out” periods
- Permitting participation at the student’s own rate, with freedom to rest as necessary
- Eliminating competitive and emotional stress factors

Collision and contact concerns

- Assign zones of play
- Use individual activities
- Change nature of implement (e.g., foam balls instead of hard balls)
- Group students according to size, abilities, and needs
- Provide clear and concise directions, rules, and regulations
- Provide play area free of obstacles, barriers, or hazards
- Ensure proper padding of facilities and equipment according to activity

Addressing visual and perceptual motor limitations

- Use brightly colored objects
- Use soft objects (e.g., foam)
- Use velcro to assist in catching
- Decrease distances
- Change implement or increase the size to decrease speed of flight and movement (e.g., a whiffle ball instead of a softball)
- Provide playing areas that are free of hazards
- Make sure the play area is well lit
- Familiarize the student with the play area prior to the start of an activity

Curriculum suggestions

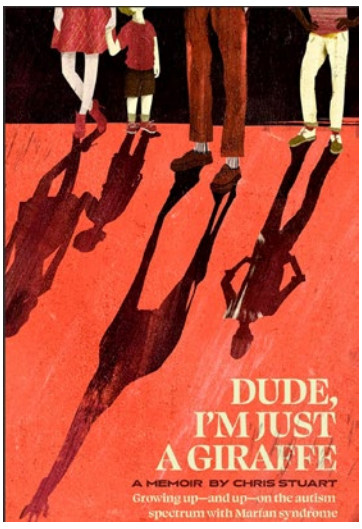
Grades K-3

- Movement exploration activities, games of low organization (with limitations as described above)

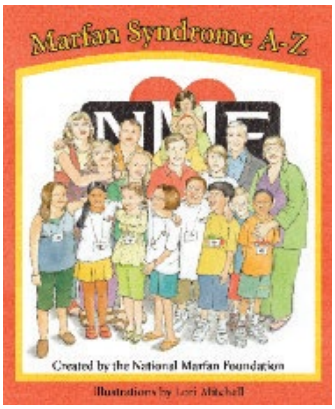
Grades 4-12

- Archery, billiards/pool, board games, bowling, bicycling (stationary and/or leisurely) croquet, dance/rhythms (rhythmic elements, singing games, folk, square, social), darts, golf, gymnastics (balance activities), horseshoes, relaxation exercises, shuffleboard, walking, aquatics/water activities (safety skills, swimming strokes, pool exercises)

4 ADDITIONAL RESOURCES



The Marfan Foundation Publications



Marfan Syndrome A to Z: This book for students ages 4–8 features vivid illustrations of diverse students and families in real-life situations. *Marfan A to Z* normalizes the student's experience with Marfan syndrome. A specialized illustration style lets students complete coloring in the pictures (more coloring pages are available online) and have fun finding the hidden pictures on each page. A glossary of helpful terms is included for parents. Written by a committee of parents and professionals and illustrated by Lori Mitchell.

Marfan Does Not Mean Martian: This book for students ages 8–12 is an inspirational story about Marvin, a boy diagnosed with Marfan syndrome, and the friendship he builds with his new neighbor Joe. The story educates students about Marfan syndrome while teaching them about diversity and acceptance. Written by Elias Clark Turner, a teenager who has Marfan syndrome, and illustrated by Alexandra Dubow.

Dude, I'm Just a Giraffe, is Chris Stuart's memoir about his childhood in Florida and New York to growing into adulthood in Arizona. Free copies are available through the Help and Resource Center. The book is also available for a fee on Amazon.

My Marfan Magic: Being different is magical. Meet Marvin the bunny... he has Marfan syndrome. That's what makes him different, but being different is magical. A children's book (ages 2-10) developed by Kathleen Bolton from our Help and Resource Center.

For the School Nurse: A companion resource to this teacher guide.

Emergency Care Plan

The Emergency Care Plan (ECP) attached is customizable and will help record information about the student's condition and provides contact information for family, physicians, hospitals, and an ambulance service in the event of a medical emergency. Teachers, as well as the school nurse, principal, and other adults in the student's life should have a copy of it readily available.

See page 23.

Classroom Activities

The attached classroom activities include a word search, crossword puzzle, and word cloud. These tools were developed to help teachers educate their students about Marfan syndrome.

See pages 24–28

These and other resources are available at Marfan.org

EMERGENCY CARE PLAN

Student:		Date:
DOB:	Grade:	Homeroom:
Parent(s)/Gaurdian(s):		
Contact Information	Home Telephone Number:	
Parent	Work Telephone Number	Cell Telephone Number
Mother		
Father		
Healthcare Contact Information	Name	Telephone Number
Primary Care Provider/Pediatrician		
Hospital Preference		
Ambulance Preference		
Health Insurance		
Cardiologist		
Orthopedist		
Geneticist		
Pulmonologist		
Ophthalmologist		
Health Conditions		
Cardiac Concerns:		
Vision Concerns:		
Orthopedic Concerns:		
Signatures		
Reviewed by:		Date:
Student:		Date:
School Nurse:		Date:
Parent/Guardian:		Date:

CLASSROOM ACTIVITY:

WORD SEARCH

Marfan syndrome is a genetic condition that affects many different parts of the body. Find the Marfan-related words and terms below in the puzzle and discuss how the different words are related to Marfan syndrome.

G A O R T A L L O G T X L E H N

L O N G A R M S Y N D R O M E N

A D O C T O R M A R F A N N A L

S S E Y E S T R T G L Y G G R U

S K E L E T O N U R S E L E T N

E F E E T M E D I C I N E N R G

S S R E H G S E M F I N G E R S

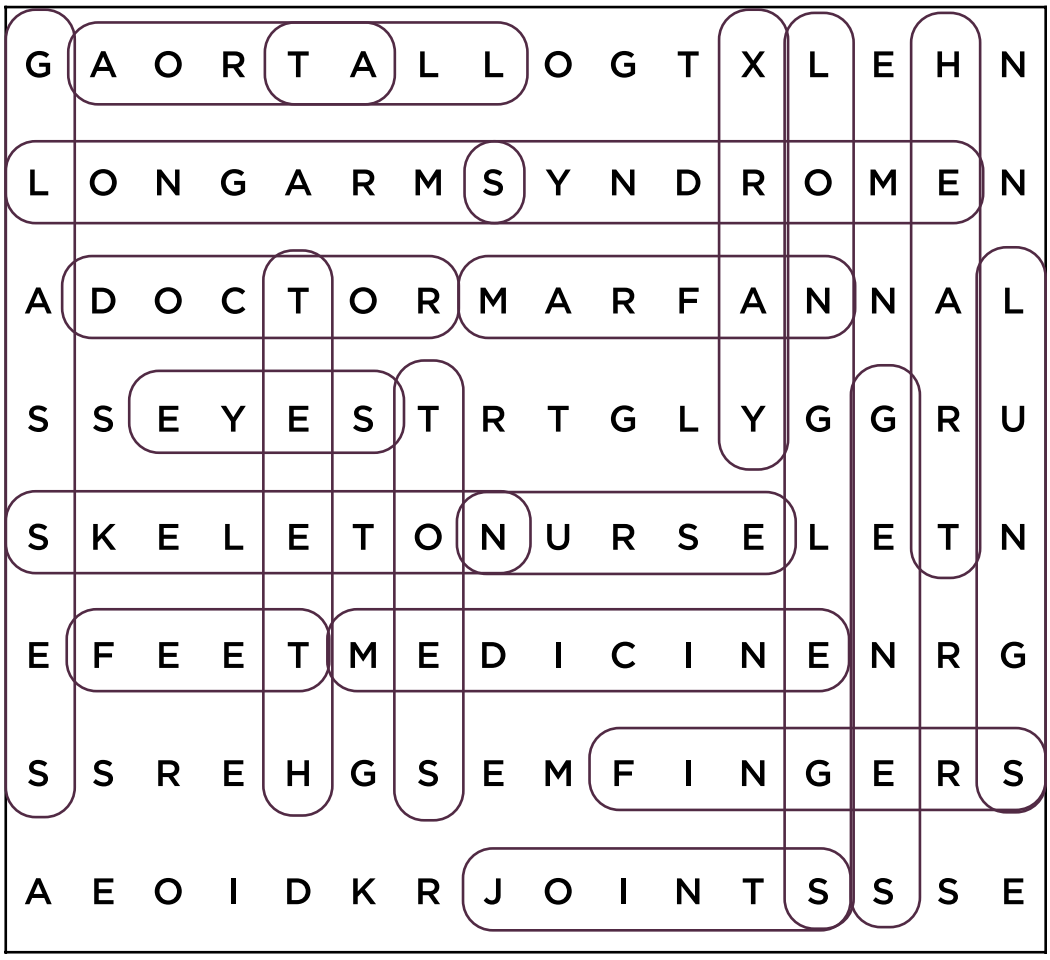
A E O I D K R J O I N T S S S E

aorta
doctor
eyes
feet
fingers
genes
glasses
heart
joints
long arms
long legs
lungs
marfan
medicine
nurse
skeleton
syndrome
tall
teeth
toes
xray

CLASSROOM ACTIVITY:

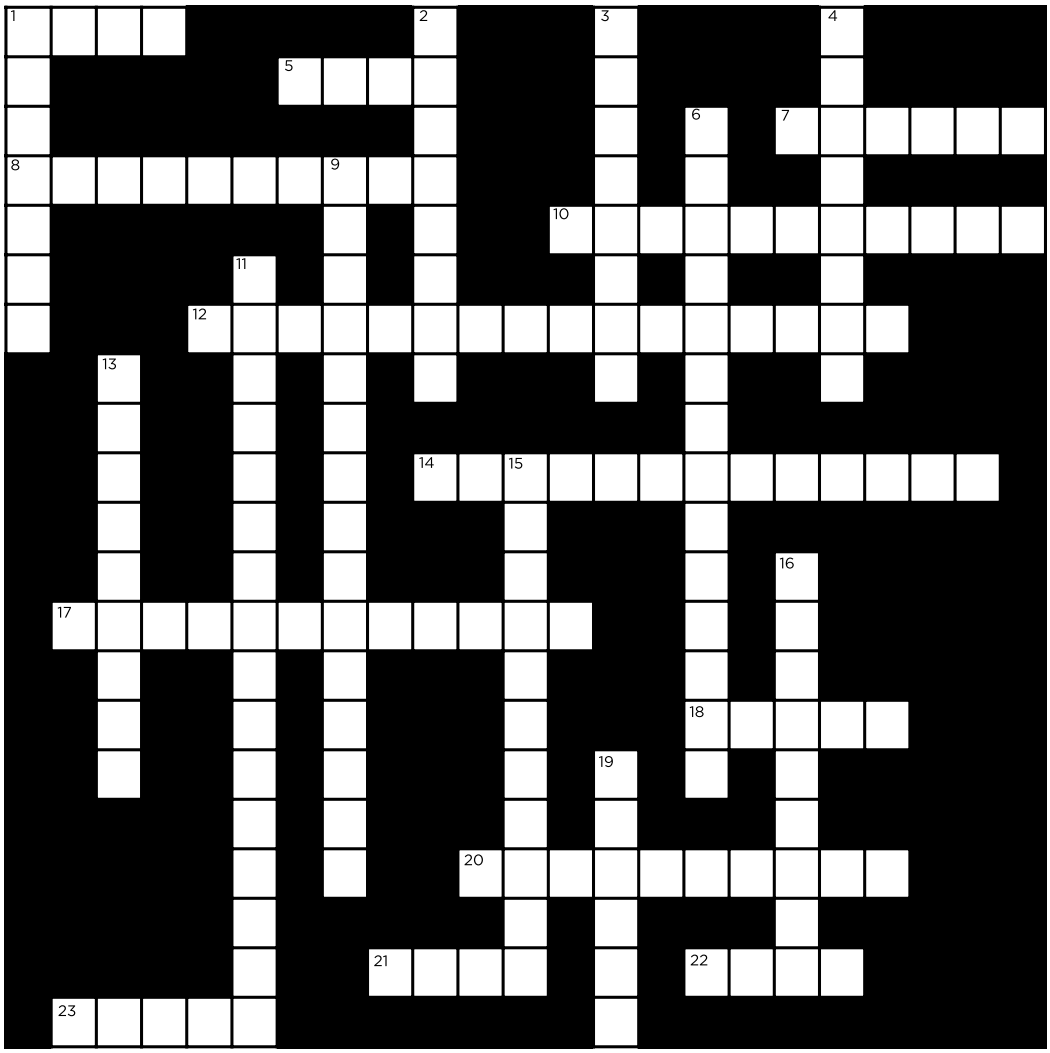
WORD SEARCH

ANSWER KEY



CLASSROOM ACTIVITY: **CROSSWORD PUZZLE**

Marfan syndrome is a genetic condition that affects many different parts of the body. Learn common terms and facts about Marfan syndrome by completing this crossword puzzle.



ACROSS

1 Pneumothorax, or collapsed _____, can be a serious complication of Marfan syndrome

5 The basic physical and funcational unit of heredity

7 _____ excavatum or carinatum, deformities of the breastbone, also called funnel chest or pigeon breast

8 Marfan syndrome is caused by a mutation in the fibrillin gene on _____ 15 in the human genome

10 Bone and joint doctor

12 A tear in the inner lining of the aorta (2 words)

14 Spider fingers

17 Heart doctor

18 The large blood vessel that carries blood away from the heart

20 Part of the heart that regulates blood flow by opening and closing with each heartbeat (2 words)

21 Dislocated _____ of the eye is a common feature in Marfan syndrome and can cause severe vision problems

22 Jonathan Larson, creator of the hit musical _____, died of Marfan-related causes before his show even opened

23 A curving of the _____ is called scoliosis

DOWN

1 U.S. President thought by some to have had Marfan syndrome

2 In biology, the study of inheritance

3 An abnormal ballooning of a blood vessel

4 Bones and joints

6 A painless test that uses sound waves to take pictures of the heart and blood vessels

9 Eye doctor

11 Tissue that provides support and structure throughout the body (2 words)

13 Flat feet, also called ___ ___, that may be larger than normal make it difficult to find shoes that fit (2 words)

15 Extremities (3 words)

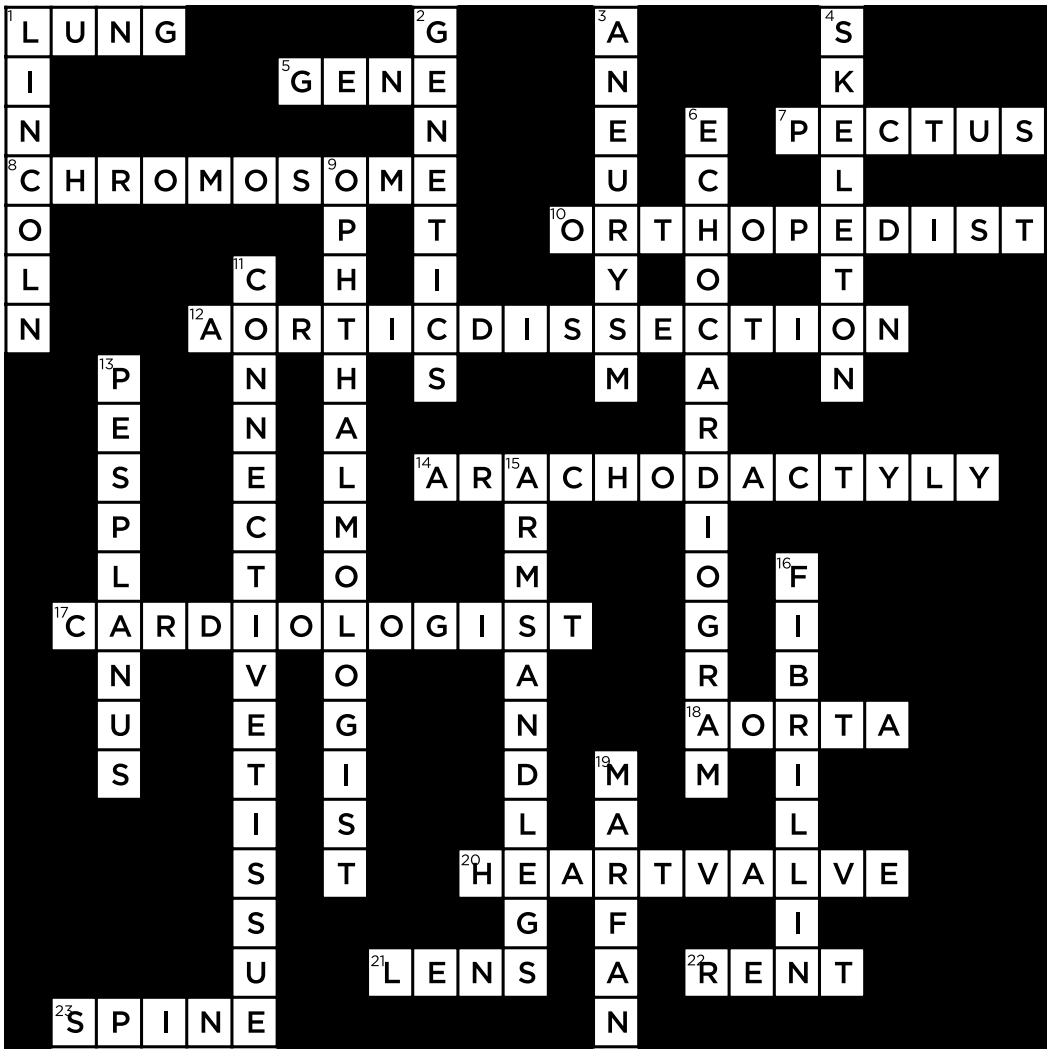
16 A protein that is essential for the formation of elastic fibers found in connective tissue

19 Antoine _____ is the French pediatrician who first identified the condition

CLASSROOM ACTIVITY:

CROSSWORD PUZZLE

ANSWER KEY



CLASSROOM ACTIVITY:

WORD CLOUDS

A “word cloud” is a visual representation of text. Creating word clouds is an easy, fun, and creative way to enforce key points of lessons, reading assignments, or research projects, and to incorporate technology into the classroom.

The 21st Century Educational Technology and Learning Blog written by Michael Gordon includes a comprehensive list of how word clouds can be incorporated into the classroom. There are many free online word cloud generators, each with varying features. Tagxedo is one that provides numerous creative options and flexibility.

Here’s a word cloud of celebrities and historical figures assocated with Marfan syndrome created by The Marfan Foundation as an example.

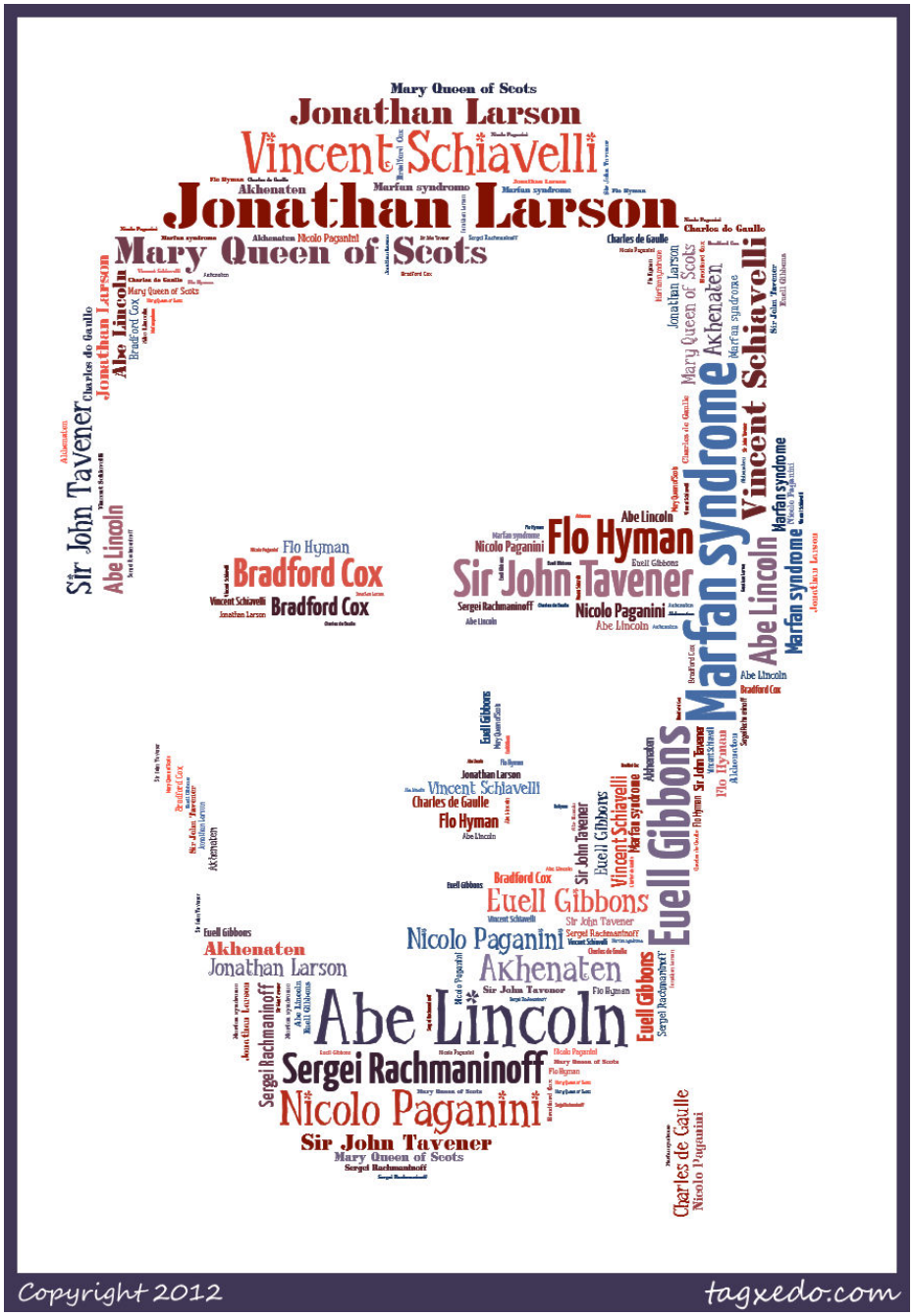
Celebrities and historical figures confirmed to have Marfan syndrome or a related condition:

Jonathan Larson, Playwright
Flo Hyman, Athlete
Vincent Schiavelli, Actor
Euell Gibbons, Naturalist
Bradford Cox*, Musician
Sir John Tavener*, Composer

Suspected of having Marfan syndrome:

Abraham Lincoln, U.S. President
Mary Queen of Scots,
Scottish Queen
Niccolò Paganini, Musician
Charles de Gaulle,
French President
Sergei Rachmaninoff, Musician
Akhenaten, Pharaoh

*Still living



THE **MARFAN** FOUNDATION

Know the signs. Fight for victory.

The Marfan Foundation, founded in 1981, is a non-profit voluntary health organization that creates a brighter future for everyone affected by Marfan syndrome and related conditions.

- We pursue the most innovative research and make sure that it receives proper funding.
- We create an informed public and educated patient community to increase early diagnosis and ensure life-saving treatment.
- We provide relentless support to families, caregivers, and healthcare providers.

We will not rest until we've achieved victory—a world in which everyone with Marfan syndrome or a related condition receives a proper diagnosis, gets the necessary treatment, and lives a long and full life.

22 Manhasset Avenue, Port Washington, NY 11050
516.883.8712 | 800.8.MARFAN | **Marfan.org**

Have additional questions? Contact our Help and Resource Center at Marfan.org/ask

Credits

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