

Physical Education Goals for Children with Marfan Syndrome

Helen D. Iams, MD, MS

This document is a detailed discussion of physical activity needs and restrictions for children who have Marfan syndrome. It also includes practical suggestions for curriculum modifications for the PE teacher.

What is Marfan syndrome?

Marfan syndrome (MFS) is a genetic disorder of connective tissue. Because connective tissue is found throughout the body, Marfan features can occur in many different parts of the body. Marfan features are most often found in the heart, blood vessels, bones, joints, and eyes. Sometimes, the lungs and skin are also affected. Marfan syndrome does not affect intelligence. It occurs in both boys and girls and in all racial and ethnic groups.

General approach to physical education for students with Marfan syndrome

Children with Marfan syndrome require modification to their physical activity for a variety of reasons that we will address below. Even though they have physical limitations, it is important for each affected child to remain as active as possible to maintain both physical and mental health.

Marfan syndrome causes a wide variety of physical problems in both type and severity. Each child has a different combination of these problems. Because of this great variability of physical problems, most children with Marfan syndrome will need a custom-designed exercise program in lieu of standard physical education. The information provided below is designed to give physical educators the information they need to create gym programs that help children with Marfan syndrome be as active as possible. The information can also serve as a guide for persons responsible for a child's physical activity outside of the school setting, at, for example, summer camp or an after school program.

Parameters for exercise should be set by the child's physicians. The physical educator can then select activities for the child based on that child's needs, abilities, and limitations. These children are often seen by a number of different specialists, such as cardiologists, orthopedists, and ophthalmologists. Each specialist should be consulted individually to find out what limitations would be prudent from their perspective.

How Marfan syndrome affects students' ability to exercise

Marfan syndrome affects connective tissue throughout the body. Because connective tissue is a component of many parts of the body, Marfan syndrome affects the body in many ways. We will identify in greater detail the features of Marfan syndrome and discuss how they can affect the child's ability to be physically active.

Musculoskeletal System

The most obvious features of Marfan syndrome are in the musculoskeletal system.

- The child may be tall and have elongated extremities.
- The child may be very thin and may lack muscle bulk and strength.
- Joints may be hyperextensible and prone to dislocating. Joint contractures may also be present.
- The combination of underdeveloped muscles and joint hypermobility can contribute to poor coordination and delay in acquiring gross and fine motor skills.
- The spine may be curved, resulting in scoliosis or kyphosis which can greatly limit the range of motion of the back. The child may need to wear a brace to support the spine and this brace may limit range of motion even further.
- The chest wall might be deformed, creating a sunken or protruding chest. Most often, chest deformities are merely a cosmetic problem. Occasionally these deformities will impair normal functioning of the lungs. To address the problems with lung function, the child may have had surgery or may need to wear a brace. Both of the solutions may cause problems by limiting exercise capacity and range of motion.
- The child may have a very narrow and highly arched palate and may not be able to use a standard mouth guard.

How Marfan syndrome affects students' ability to exercise, continued

Ocular System

The eyes are commonly affected by Marfan syndrome.

- The lens may dislocate.
- The retina can tear.
- The cornea is often flatter than usual, making it difficult to fit contact lenses. Contact lenses, if worn, tend to fall out easily.
- The child may be near-sighted and may require corrective lenses.
- In some cases, bifocals may have to be worn over contact lenses.
- The child may have difficulty with visual perception and with eye-hand coordination.
- The child may have difficulty in following the flight of an object.
- The child may have difficulty tolerating bright light.

Cardiovascular System

The cardiovascular system is affected in approximately 90% of people with Marfan syndrome. Cardiovascular features include:

- Enlargement (dilation) of the aorta
- Leaking of the aortic valve (aortic regurgitation)
- “Floppy” mitral valve (mitral valve prolapse)
- Leaking of the mitral valve (mitral regurgitation)
- Dissection of the aorta that occurs if the aorta becomes too large

Aortic dilation is of major concern because a severely dilated aorta can tear (dissect) or rupture. Children (and adults) with Marfan syndrome have regular echocardiograms to monitor the size of their aortas. When the aorta reaches a certain size, prophylactic repair of a dilated aorta can prevent life-threatening aortic dissection.

Spontaneous aortic dissection (in the absence of trauma) is rare in school-aged children, but a gradual increase in aortic size is common. Exercise modification and beta-adrenergic blocker medications are important components of cardiac management that seek to help slow the rate of aortic growth. Medication and exercise modification help reduce the force with which blood is pumped from the heart, thus reducing

How Marfan syndrome affects students' ability to exercise, continued

stress placed on the aorta. Beta-blocker medication may have side effects including fatigue, feeling sleepy, and reduced ability to concentrate.

Repair or replacement of a leaking mitral valve may be needed in a school-aged child.

If the correction required valve replacement, the child will be on a blood thinner medication to keep any clots from forming on the replacement valve. This will leave the child prone to spontaneous bleeding and easy bruising.

Pulmonary System

The lungs can have defects called apical blebs. These are weakened areas of tissue that over inflate, much like a weak spot in a balloon. These blebs can break, causing the lung to suddenly collapse. Improper breathing techniques during activities put further stress on lungs and may lead to a lung collapse.

The lungs may be underdeveloped with reduced capacity for gaseous exchange. Children with this lung problem may have reduced exercise tolerance.

Emergencies and how to recognize them

People with the Marfan syndrome can suddenly develop symptoms that signal the need for immediate medical attention. There is need for rapid transport to a hospital if the child suddenly develops such symptoms. Both the school nurse and the physical educator should be alert for signs of distress in the child that indicate a medical emergency.

Aortic rupture or dissection is the leading cause of death in people with Marfan syndrome, and is therefore a medical emergency of grave concern. **Although spontaneous aortic rupture or dissection is rare in school-aged children, it can occur.** Therefore it is important to know how to recognize the symptoms. Aortic rupture or dissection is typically painful, described by people as “tearing chest pain that bores through” to their back. Much less commonly, acute dissection may present as sudden collapse or syncope. However, aortic dilation can be pain-free and the only symptom may be a shortness of breath.

Sudden shortness of breath and chest pain can also be a signs of spontaneous collapse of the lung. Spontaneous lung collapse is not life-threatening but does require immediate medical attention. Other possible signs of cardiac or pulmonary problems are fatigue and an irregular heart rate. Any of these symptoms are serious and if the child complains of these symptoms, he should be promptly transported to a local emergency room.

Children with Marfan Syndrome are prone to retinal detachments. Affected children who complain of any vision problems, including flashing lights, spots in their vision, or sudden loss of vision, should be removed from physical activity and referred immediately to the school nurse.

Children who have had a valve replacement will probably be on a blood-thinner. Any child on a blood thinner can develop a spontaneous bleed or excessive bruising and should be set to the school nurse for evaluation if these occur. The replacement valve may make a clicking sound that can be heard when standing near the child. This is normal and is the sound of the device working.

Activities that should be avoided

The physical demands of some sports can lead to devastating problems for students with Marfan syndrome. Although there are a number of safe activities for a child with Marfan syndrome, some activities and situations should be avoided. These include:

- Most competitive sports—as exercise is often done at maximal capacity during competition. Golf is generally an exception.
- Activities that place excessive stress across the joints could cause dislocations or damage to the joint surface.
- Wrestling and many gymnastic activities.
- Isometric sports, such as weight-lifting, that create high blood pressure.
- SCUBA diving—because the lungs may not withstand stress from pressure gradients.
- Contact sports and sports requiring sudden exertion, because a blow to the chest or the strain of jumping or stretching can result in serious injury and even aortic dissection.
- Sports with a risk of falling or impact, such as gymnastics, diving or skating.
- Demanding endurance activities, such as competitive running and bicycling.

Almost all physical activity involves some aspect of body collision if not presented safely. Therefore care must be taken with most activities. If the child is on a blood thinner, a collision could result in heavy bleeding. Exercise to the point of exhaustion should be prevented to avoid the increases in heart rate, blood pressure and force of muscular contractions that are present in an exhausted state.

Suggested physical activities and activity modifications

General Modifications

It is not possible to create a single exercise program that would be safe for all children who have Marfan syndrome. Each child's physicians should provide the physical educator with information about safe activity levels for that child. The physical educator can then design activities within these levels. It may be helpful for the physical educator to provide the physician with a list of planned activities in the PE curriculum. If possible, have the child help in selecting activities. This will increase their enjoyment of the program.

A child may need assistance in developing a realistic self-concept of abilities and limitations. It is important to recognize that the child's level of comprehension can be misjudged. Because of their increased height, children with Marfan syndrome may be treated as older than their actual age.

Modifications to address cardiovascular issues

- The child with Marfan syndrome should be encouraged to take part in noncompetitive activities performed at a sub-maximal level of effort.
- Select activities promoting the concept of self-competition to minimize the effects of peer pressure for the child to exceed physical exertion limits.
- Instruct the child in self-monitoring techniques and provide an exercise environment in which an affected child can practice self-monitoring.
- Instruct the child in safe levels of intensity and duration.
- Include instruction in relaxation techniques, safety, and correct breathing mechanics.
- A child with Marfan syndrome must be taught to never "push through" symptoms.
- Provide adequate time for gradual warm-up and cool-down.
- Monitor the child's level of exertion more closely under extreme weather conditions because heat and cold may add additional stress and may affect the child's endurance and exertion level.

Most children with Marfan syndrome receive beta-blocker medications. These medications depress the heart rate. Therefore heart rate will not be a true indicator of exertion level. Children on a beta blocker under age

Suggested physical activities and activity modifications, continued

seven should keep prolonged heart rate under 120 beats a minute. Older children (and adults) on a beta blocker should keep heart rate under 100 beats a minute.

A “graded” exercise program may be recommended by the child’s physicians. In such instances, use an estimate of the child’s exercise tolerance to determine the maximal heart rate and functional capacity. The child must never be actually tested to identify the maximal heart rate. The estimation of the maximal heart rate should be based on the child’s age. It is generally recommended that Marfan patients stay at an aerobic level of work at 50-60% of their maximal heart rate. If there is no evidence of aortic dilatation, the physician may permit activity at higher levels of intensity.

Modifications that address musculoskeletal issues

The student may have muscular underdevelopment and joint laxity or, conversely, contractures. Overall lack of bulk and muscle tone may need to be addressed with strengthening exercises. Muscle strengthening is also useful when working with individuals who demonstrate joint hyperextensibility. Focus on activities that increase strength of both muscles and ligaments. However, the child should only do exercises with a weight that enables 15-20 repetitions comfortably. It is recommended to avoid activities involving heavy weights or intense isometric exertion. The child’s musculoskeletal manifestations may have required physical therapy so the child may already have prescribed exercises. Ask the family about any physical therapy the child is doing, as this may affect the physical educational activities at school.

Modifications to Improve Coordination

A child with Marfan syndrome may have difficulty with eye-hand and eye-foot coordination. Provide opportunities to practice visual tracking of objects in motion and to develop other sequential perceptual motor skills (fine motor, gross motor, balance, spatial and body awareness). The physical educator may be able to enhance the child’s body mechanics and improve posture by providing appropriate exercise.

Suggested physical activities and activity modifications, continued

Modifications for Required Equipment

If the child needs a brace during sports activities, his or her maneuverability, flexibility, speed, and endurance may be affected. The physical education instructor should be informed by an orthopedist what restrictions the brace will create. When a back or body brace is worn, the head and neck should be protected during physical activity by suitable padding. If the child has had chest wall surgery, the surgeon will need to inform the physical educator about any further restrictions that are necessary. If mouth guards are required for an activity, the child may need custom-made mouth guards to accommodate the narrow mouth. If hernias are present, the child 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.

Provide instruction in selecting appropriate clothing for physical activity to insure comfort and easy movement. Unnecessary circulatory restrictions created by clothing or bracing must be eliminated or minimized. To decrease the potential for falling, only appropriate, well fitting, and supportive footwear should be worn. The equipment the child needs to wear may cause embarrassment so provide the child with privacy for dressing and showering.

As with any other student wearing glasses during athletic activities, the child will need glasses with shatter-proof lenses and rounded edges. If the child needs to wear contact lenses, there is a greater likelihood of their falling off during physical activity due to the flatter corneas. The child may need to wear sun glasses for activities taking place outside or in brightly lit places.

Suggested physical activities and activity modifications, continued

Suggestions for Modifying Activities

To address intensity, endurance and fatigue:

- Decrease duration of an activity
- Decrease size of playing area
- Use frequent “time out” periods
- Permit participation at the child's own rate, with freedom to rest as necessary
- Eliminate competitive and emotional stress factors
- Reduce weight of implement
- Use transportation or support devices
- Utilize sitting or lying position in place of standing
- Utilize greater numbers of participants
- Utilize aquatics for support during activity
- Utilize relaxation techniques and relaxing music during activities
- Incorporate proper breathing techniques

To address collision and contact concerns:

- Assign “zones” of play
- Use individual activities
- Use “singles” rather than “doubles” in racquet games, where appropriate
- Change nature of implement (utilize “Nerf,” foam, and “rag” items)
- Group children according to size, abilities, and needs
- Provide clear and concise directions, rules, and regulations
- Provide play area free of obstacles, barriers, or hazards
- Insure proper padding of facilities and equipment according to activity

Suggested physical activities and activity modifications, continued

To address visual and perceptual motor limitations:

- Use brightly colored objects
- Use “soft” objects (foam, Nerf, rag)
- Use Velcro to assist in “catching”
- Decrease distances
- Change implement to decrease speed of flight and movement
 - o (e.g., a whiffle ball instead of a softball)
- Place child to insure a clear and close visual field
- Increase the size of the implement when necessary
- Provide playing areas that are free of hazards
- Utilize appropriate illumination
- Familiarize the child 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, shuffle- board, walking, aquatics/water activities (safety skills, swimming strokes, pool exercises)

Summary of key points

A child with Marfan syndrome should have a physical activity program that:

- provides opportunity for physical activity in order to optimize physical and mental health.
- is individualized to acknowledge the Marfan features and limitations specific to the child
- creates exercise modification that
 - o Reduces stress on the aorta
 - o Minimizes opportunity for head or chest trauma
 - o Reduces stress on joints
 - o Accommodates possible reduced lung capacity

Note: Special thanks to the late Tom Romeo who wrote the document from which this resource is adapted.