Need-to-Know Information for the SCHOOL NURSE

Raising awareness of Marfan syndrome and related disorders

THE MARFAN FOUNDATION

CONTENTS

1 INTRODUCTION

"The practice of school nursing began in the United States on October 1, 1902 when the initial role of the school nurse was to reduce absenteeism by intervening with students and families regarding healthcare needs related to communicable diseases. While the nurse's role has expanded greatly from its original focus, the essence of the practice remains the same. The school nurse supports student success by providing healthcare assessment, intervention, and follow-up for all children within the school setting." (National Association of School Nurses, 2002)

As the health needs of schools have changed since 1902, the role of school nursing has evolved to meet those changing needs.

The school nurse plays an important leadership role in prevention of illness or injury, identification of medical problems through screening programs, and overall health maintenance. This is particularly important when a student has a chronic condition that requires more intense monitoring. One such condition is Marfan syndrome. It is estimated that 200,000 people in the United States have Marfan syndrome or a related connective tissue disorder, although many are not diagnosed until it becomes life threatening.

The school nurse is in a unique position to identify students in need of an evaluation for Marfan syndrome. Sensitive and professional communication of this concern can help identify, diagnose, and manage students who might otherwise have slipped through the cracks. Early diagnosis and management is crucial in order to assure that those affected have hope for a normal life-span. If not diagnosed and managed appropriately, it is anticipated that many, if not most, affected people will have a potentially fatal aortic dissection and rupture in their thirties or forties (The Marfan Foundation, 2000).

What is Marfan syndrome?

Marfan syndrome is a life-threatening genetic disorder 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 disorders related to Marfan syndrome that cause people to struggle with the same or similar physical problems, and anyone affected by these disorders 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 disorders, including:

- A detailed medical and family history, including information about any family member who may have the disorder 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 disorders 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 active 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 disorders are related to Marfan syndrome?

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

Related disorders 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 disorder 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 disorder have dislocated eye lenses and Marfan syndrome skeletal features. Most people with this disorder 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, widely-spaced 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.
- **Ehlers-Danlos syndrome.** A group of genetic connective tissue disorders characterized by unstable, hyper-mobile joints; loose, "stretchy" skin; and tissue fragility.

Many of the related disorders 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 disorders—and vice versa.

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

Further, since the diagnosis of the disorder 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 sometimes 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 IDENTIFICATION OF STUDENTS IN NEED OF EVALUATION

The school nurse's role in identification of Marfan syndrome

A school nurse will never be called upon to make the diagnosis of Marfan syndrome, but plays a critical role in recognizing the features and possibly encouraging the student's parent to speak to their doctor about further evaluation.

Screenings already conducted in most schools provide opportunities to raise suspicion of Marfan syndrome including:

- general appearance
- postural screenings
- vision screenings
- Body Mass Index (BMI) screenings
- sports pre-participation physical evaluations

General appearance

A number of skeletal features may indicate Marfan syndrome, as outlined above. The presence or absence of any one feature is not enough to diagnose or rule out Marfan syndrome. Children with Marfan syndrome frequently grow at accelerated rates. For instance, by age two, many children with Marfan syndrome will already have height greater than the 95th percentile. As a result, children who stand out above the rest of their classmates should be observed for other features.

Postural screenings

One of the possible skeletal features in identifying Marfan syndrome is scoliosis. Since many school districts have a routine scoliosis screening program, this may be one of the best opportunities to identify children who have additional features of Marfan syndrome and may need an evaluation for the disorder.

Approximately 50 percent of people with Marfan syndrome develop scoliosis, although only one third require treatment for it. Therefore, although scoliosis may not be severe enough to warrant aggressive treatment, it may be enough to initiate suspicion of Marfan syndrome. Look at a child with scoliosis for other features of the disorder.

Vision screenings

People with the Marfan syndrome often experience dislocated lenses (extopic lentis), retinal detachment, or nearsightedness. School based screenings may suggest that a student has poor vision, perhaps of unknown cause. A student may also report problems with depth perception. Since nearsightedness, or myopia, is relatively common in the general community, it cannot be assumed that a student who is near-sighted has Marfan syndrome. If the student has poor vision and has other features of the disorder, further follow-up is warranted.

Body Mass Index (BMI) screenings

As with other school based screenings, it is important to put BMI into the context of the whole person and recognize the features of Marfan syndrome. A thin stature, either particularly tall or tall for one's family, in combination with other features of Marfan syndrome as described earlier, indicates the need for further evaluation.

Because they are often thin, children with Marfan syndrome are sometimes suspected by classmates, or by a teacher or school nurse, of having an eating disorder. Because eating disorders get so much attention, it may be difficult to convince people that being thin is a part of Marfan syndrome and not a sign of an emotional problem.

Doctors do not understand why people with Marfan syndrome have so much diffi-culty gaining weight. There is no special diet, dietary supplement or weight lifting program that will add desired pounds or muscle.

Sports Pre-participation Physical Evaluations (PPE)

Because many young people with Marfan syndrome are very tall and have flexible joints, features which are perceived as desirable in competitive athletes, they are often encouraged to participate in sports. It is generally expected that all school athletes will have a thorough pre-participation evaluation before being allowed to play. If conducted properly, the PPE provides a great opportunity to identify individuals who have not been diagnosed previously. Unfortunately, the quality and comprehensiveness of PPE forms vary greatly from state to state.

The following are recommendations from the American Heart Association regarding pre-participation screening in athletes:

- A complete and careful personal and family history and physical examination designed to identify (or raise suspicion of) those cardiovascular lesions known to cause sudden death or disease progression in young athletes, is the best available and most practical approach to screening populations of competitive sports athletes, regardless of age. Such cardiovascular screenings are an obtainable objective and should be mandatory for all athletes. They recommend that both a history and a physical examination be performed before participation in organized sports, and repeated every two years.
- Athletic screenings should be performed by a healthcare worker with the requisite training, medical skills, and background to reliably obtain a detailed cardiovascular history, perform a physical examination, and recognize heart disease.
- Athletic screening evaluations should include key questions to determine:

  (1) prior occurance of exertional chest pain/discomfort or syncope/near-syncope as well as excessive, unexpected, and unexplained shortness of breath or fatigue associated with exercise;

  (2) past detection of a heart murmur or increased systemic blood pressure; and

  (3) family history of premature death (sudden or otherwise), or significant disability from cardiovascular disease in close relative(s) younger than 50 years old or specific knowledge of occurrence of conditions such as hypertrophic cardiomyopathy, dilated cardio-myopathy, long QT syndrome, clinically important arrhythmias, or Marfan syndrome.
- Because of the heterogeneity in the design and content of pre-participation examinations, it is recommended that a national standard for pre-participation medical evaluations be implemented.

3 WHAT TO DO IF YOU THINK A STUDENT NEEDS FURTHER EVALUATION

Marfan syndrome is relatively uncommon and because the severity of the outward physical characteristics can vary greatly, 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 aortic dissection and sudden death.

When routine screenings indicate the need for further evaluation, the school nurse plays a critical role and should encourage the student's parent to speak to their doctor about further evaluation.

When referring a student for evaluation for Marfan syndrome, take care to provide a complete explanation of why you are making the referral.

A sample letter to the parent and a checklist of Marfan features are attached. Both may assist you in communicating your concerns to the parent without creating undue alarm.

4 SPECIAL CONSIDERATIONS 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. A template for developing an Individualized Health Plan (IHP) for children with Marfan syndrome is attached.

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 disorder. 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.

5 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 disorder 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 disorder 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)

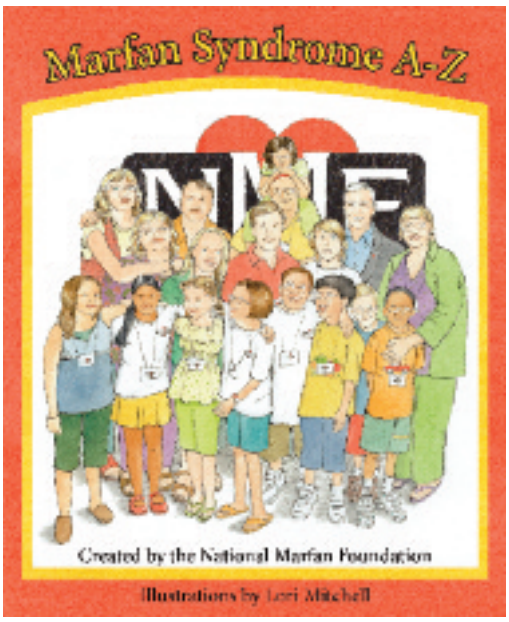
Grades 4-12

- 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)

6 ADDITIONAL RESOURCES

The Marfan Foundation Publications

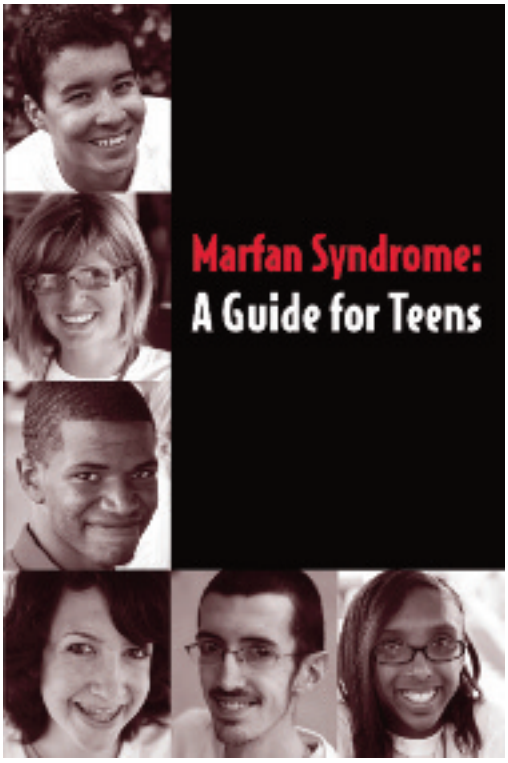
Marfan Syndrome A to Z: This book for students ages 4–8 features vivid illustrations of diverse students and families in real-life situations. *Marfan A to Z* normalizes the student's experience with Marfan syndrome. A specialized illustration style lets students complete coloring in the pictures (more 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.

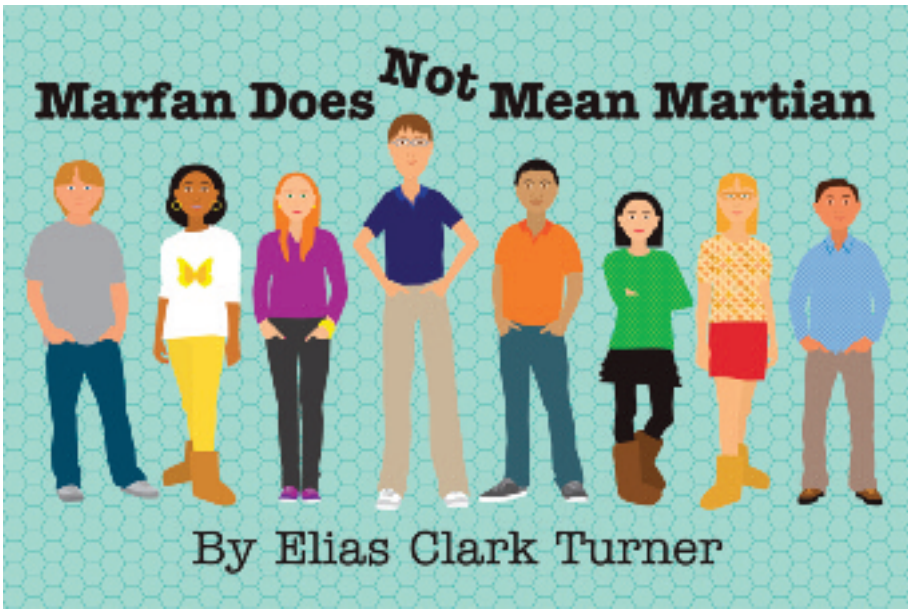


Marfan Does Not Mean Martian: This book for students ages 8–12 is an inspirational story about Marvin, a boy diagnosed with Marfan syndrome, and the friendship he builds with his new neighbor Joe. The story educates students about Marfan syndrome while teaching them about diversity and acceptance. Written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

Marfan Syndrome A Guide for Teens: A comprehensive resource for teens, this booklet addressing all aspects of living with Marfan syndrome from diagnosis and treatment to psychosocial issues such as dealing with family, friends and school. It includes a glossary of Marfan-related terms and listing of additional resources, and features photographs of teens living successfully with Marfan syndrome.

Marfan Syndrome Need-to-Know Information for the School Nurse: A companion resource to this teacher guide, this CD-ROM contains a wealth of information and resources tailored to the school nurse.





These and other resources are available at marfan.org

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 28.

Sample letter and checklist of Marfan syndrome features

A sample letter to the parent and a checklist of Marfan features attached will assist you in communicating your concerns to the parent without creating undue alarm.

See pages 29–30

Individualized Health Plan (IHP)

A template for developing an Individualized Health Plan (IHP) for children with Marfan syndrome adapted with permission from Sunrise River Press is attached.

See pages 31–35

National Association of School Nurse Statements

Official statements from the National Association of School Nurses on both Individual Health Plans (IHP) and Emergency Care Plans (ICP) are attached.

See pages 36–40

EMERGENCY CARE PLAN		
Student:		Date:
DOB:	Grade:	Homeroom:
Parent(s)/Guardian(s):		
Contact Information	Home Telephone Number:	
Parent	Work Telephone Number	Cell Phone 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 Condition:		
Cardiac Concerns:		
Vision Concerns:		
Orthopedic Concerns:		
Signatures		
Reviewed by:		Date:
Student:		Date:
School Nurse:		Date:
Parent/Guardian:		Date:

Adapted with permission from Sunrise River Press

SAMPLE LETTER TO PARENTS

Dear _____ :

I have observed that _____ has the features checked on the reverse side. Because people with these features sometimes have a medical condition called Marfan syndrome, I strongly suggest s/he be checked for this condition.

Having these features does not always mean a person has Marfan syndrome, but it does mean a person should have an evaluation by a doctor familiar with the syndrome. Please take this letter to your child's doctor and ask for an evaluation.

For more information about Marfan syndrome, including what is needed for an evaluation, you can contact The Marfan Foundation at 800-862-7326 ext. 126 or on the web at marfan.org

Please feel free to call me if you would like to discuss these recommendations further.

Sincerely,

_____ _____

Name Telephone

CHECKLIST OF MARFAN FEATURES

SKELETAL SYSTEM

- ☐ Tall, thin stature (often but not always)
- ☐ indented or protruding chest bone
- ☐ long arms and/or legs
- ☐ wrist and thumb signs (a measurement of loose joints and long fingers)
- ☐ scoliosis or kyphosis
- ☐ reduced extension at the elbows
- ☐ flat feet and/or hammer toes
- ☐ joint hypermobility
- ☐ highly arched palate and/or crowding of the teeth
- ☐ long, narrow face (dolichocephaly)
- ☐ underdeveloped cheekbones (malar hypoplasia)
- ☐ deep set (enophthalmos) or down slanting eyes
- ☐ receeding jaw (retrognathia)

CARDIOVASCULAR SYSTEM

- ☐ dilated aorta
- ☐ dissected aorta
- ☐ mitral valve prolapse
- ☐ dilation of main pulmonary artery
- ☐ calcification of the mitral annulus
- ☐ descending aortic dilation or dissection
- ☐ heart murmur

PULMONARY SYSTEM

- ☐ spontaneous pneumothorax
- ☐ apical blebs

OCULAR SYSTEM

- ☐ dislocated lens (ectopia lentis)
- ☐ abnormally flat cornea
- ☐ underdeveloped (hypoplastic) iris or dilator muscle causing increased miosis
- ☐ severe nearsightedness (myopia)

SKIN

- ☐ unexplained stretch marks
- ☐ recurrent incisional hernias

CENTRAL NERVOUS SYSTEM

- ☐ lumbosacral dural ectasia (swelling or bulging of the dura)

FAMILY HISTORY

- ☐ family member with early cardiac death, unexplained or identified as Marfan syndrome
- ☐ family history of aortic aneurysm
- ☐ height unusual for family
- ☐ height unusual for age

OTHER OBSERVATIONS

- ☐ _____
- ☐ _____
- ☐ _____

Please keep in mind that any of the above features taken individually does NOT mean a person has Marfan syndrome. However, when observed in combination, further evaluation by a doctor familiar with connective tissue disorders may be necessary.

INDIVIDUALIZED HEALTH PLAN (IHP)

Adapted with permission from Sunrise River Press. "Individualized Healthcare Plans for the School Nurse." Book and software package available at: sunriseriverpress.com

Use this outline as a guide for development of a comprehensive IHP for Marfan syndrome.

1. Assessment

A. History

- Family health history
- Growth, development, and health prior to diagnosis
- Age at diagnosis
- Circumstances prompting evaluation and diagnosis
- Initial care and lifestyle changes
- Systems currently affected, severity
- Immediate and long-term prognosis
- Program of medical and surgical care
- Frequency of evaluations
- Results of last evaluation
- Family development of acceptance
- Student's development of acceptance
- Past supportive persons, support group involvement
- Past professional eye-examination reports

B. Current Status and Management

- Height and weight
- Deformity of the sternum and spinal curves
- Appearance of limbs, fingers, and toes
- Arm span
- Current vision, interval screening for vision complaints
- Emergency care plans (ECPs) for aortic dilatation and dissection or rupture, lens dislocation and retinal detachment, sudden shortness of breath and pain associated with breathing
- Current circulatory status: blood pressure, apical and radial pulse
- Stretch marks and abdominal, inguinal, or umbilical bulges or weaknesses
- Current respiratory status: bilateral lung sounds, respiratory rate and effort initially and if symptomatic
- Current medications
- Current therapies
- Nervous system function of lower extremities (weakness, numbness, pain)
- Current management plan
- Student and family's understanding of management plan

INDIVIDUALIZED HEALTH PLAN (IHP) continued

- Spinal bracing
- Current activity tolerance: getting to and from school, traveling around both inside and outside the campus, recreational play, and physical education classes

C. Self-care

- Student's knowledge of health-care needs and management plan
- Ability to self monitor: activity tolerance, vision, skin, breathing
- Student's ability to advocate for self with staff
- Decision making skills

D. Psychosocial Status

- Student's acceptance of family interventions
- Student's utilization of coping strategies to deal with health condition: effective, ineffective
- Family's utilization of coping strategies to deal with health condition: effective, ineffective
- Behavioral issues: risk-taking behaviors, fear of performance
- Feelings about visible deformity and activity limitations
- Participation in school activities, community activities, family activities
- Supportive persons s/he is comfortable talking to (parents, physicians, school staff, psychologist, etc.)

E. Academic Issues

- Academic achievement: past and current
- School attendance pattern: reasons for absences
- Physical education and other activity restrictions
- Modifications needed in curriculum or course requirements: alternative assignments due to physical education restrictions, extended timelines for completion of assignments, and process to obtain missed assignments due to absences for appointments or illness
- School reentry plan following surgery

2. Nursing Diagnoses (N.D.) (NANDA, 2003)

N.D. 1 Chronic self-esteem disturbance (NANDA 00119) related to diagnosis of chronic illness with multiple unknowns

N.D. 2 Risk for activity intolerance (NANDA 00094 or 00092) related to:

- joint laxity, respiratory problems
- cardiac problems
- medication side effects

INDIVIDUALIZED HEALTH PLAN (IHP) continued

N.D. 3 Risk for disproportionate growth (NANDA 00113) related to: (potential for) abnormal bone development (progressive spinal deformity, sternal protrusion or concavity, long bone development)

N.D. 4 Ineffective breathing pattern (NANDA 00032) related to:

- loss of elasticity of alveoli and potential for pneumothorax

N.D. 5 Risk for powerlessness (NANDA 00152 or 00125) related to: current (or potential for) life-threatening health complications

N.D. 6 Impaired physical mobility (NANDA 00085) related to potential for spinal deformity

N.D. 7 Disturbance in body image (NANDA 00118) related to perception of physical disability:

- scoliosis
- limb length
- joint laxity
- surgical scars
- thick corrective lenses

N.D. 8 Risk for injury (NANDA 00035) related to aortic dilatation, dissection or rupture; spontaneous pneumothorax, lense dislocation or retinal detachment

3. Goals of IHP (with related Nursing Diagnoses)

The student will participate in development and implementation of ECP. (N.D. 4, 8)

The student will participate in development of emergency evacuation plan. (N.D. 2, 8)

The student will be independent in the school setting. (N.D. 1, 2)

The student will increase acceptance and positive self esteem. (N.D. 1, 5, 7)

The student will increase his/her knowledge of the disease process. (N.D. 1, 5)

The student will assist in identifying and implementing modifications to plan of care. (N.D. 5)

The student will share personal health information appropriately with staff and peers. (N.D. 3, 5)

The student will utilize healthy adaptation and coping skills. (N.D. 1, 3, 5, 8)

The student will prevent predictable physical injury. (N.D. 3)

The student will manage his/her pain effectively. (N.D. 3,6)

The student will communicate effectively with parents/guardians and school staff about school-day health needs. (N.D. 1, 7, 8)

INDIVIDUALIZED HEALTH PLAN (IHP) continued

4. Nursing Interventions

- Periodically update health information with parent/guardian and student. (N.D. 3, 8)
- Assist the student to manage episodes of pain during the school day (ice-packing joints, medications, rest, etc.). (N.D. 2, 4, 6, 8)
- Obtain parent and physician authorization for administration of medication. (N.D. 2, 3, 4, 6, 8)
- Develop ECPs and revise as needed. (N.D. 8)
- Decide when and how information will be shared with education staff. Review and update regularly. (N.D. 7, 10)
- Establish plan with parents/guardians and student to obtain health updates and share information. Review regularly. (N.D. 5, 8)
- Educate school staff on student's individual needs and review emergency plans. (N.D. 8)
- Assist teachers to modify physical education activities and requirements, as needed. (N.D. 6)
- Review playground safety rules with student. (N.D. 6)
- Encourage student to report changes in vision, breathing, leg pain, activity intolerance, getting through halls, stairs, on and off bus, etc. (N.D. 5)
- Encourage independence and activity that does not put the student at risk for injury. (N.D. 1, 5, 6, 8)
- Screen vision, height, weight, spine as needed or as agreed upon with parents and physicians. (N.D. 3, 7, 8)
- Encourage student to verbalize feelings about chronic illness. (N.D. 1)
- Provide positive feedback to student, parents/guardians, and staff regarding effective management of symptoms. (N.D. 1, 5)
- Provide age-appropriate information on Marfan syndrome to the student. (N.D. 1)
- If surgery is required, assist in planning for homebound instruction and reintegration back into school. (N.D. 2)
- Provide student with opportunities to take on responsibilities for his/her care and control situations when appropriate (i.e., talking to primary care provider office for appointments with parent permission). (N.D. 1, 5, 8)
- Clarify misconceptions the student has about self-care. (N.D. 1, 5, 8)
- Provide assessment of student's subjective concerns as they occur, such as respiratory difficulty, irregular heartbeat, or activity intolerance. (N.D. 2, 5, 8)
- Teach student how to report abnormal symptoms to the teacher, health office, and parents/guardians. (N.D. 4, 5, 8)

INDIVIDUALIZED HEALTH PLAN (IHP) continued

- Teach student how to advocate responsibly for self. (N.D. 7)
- Encourage student to express how s/he feels about or views him/herself. (N.D. 1, 7)
- Encourage student to ask questions at home and at school to assist in making good decisions or problem solving. (N.D. 1, 5, 7)
- Assist teachers to organize school day to minimize:
    - Pain and discomfort (N.D. 2)
    - Energy expenditure, if activity intolerance is occurring (N.D. 2, 6)
- Assist the student and parents/guardians to obtain medical emergency identification bracelet or necklace. (N.D. 4, 8)
- Encourage the student to wear medical identification bracelet or necklace. (N.D. 4,8)

5. Expected Student Outcomes

The student will:

- Effectively communicate health concerns and needs to parents/guardians and school staff. (N.D. 1, 5)
- Identify and describe potential health problems. (N.D. 4,5)
- Demonstrate avoidance of risk activities that could cause injury. (N.D. 8)
- Follow ECP(s) and evacuation plan when required. (N.D. 6, 8)
- Describe what Marfan syndrome is and how it affects his/her body (age-appropriate). (N.D. 2, 3, 4, 6, 8)
- Participate in classroom activities, with specific activity limitations as needed. (N.D. 1, 2, 4, 6, 8)
- Demonstrate compliance with activity limitations. (N.D. 2, 8)
- Demonstrate effective coping and adaptation skills. (N.D. 1, 5, 7)
- Demonstrate independent mobility in the school environments. (N.D. 6)
- Utilize measures to minimize pain and discomfort during the school day. (N.D. 2, 3, 4, 6)
- Consistently wear medical emergency information bracelet or necklace. (N.D. 4, 8)

National Association of School Nurses

POSITION STATEMENT

Individualized Health Care Plans

HISTORY:

Since the inception of school nursing, management of students with chronic health problems has been a vital role (American Nurses Association, 2001). The standard for this management: is provided by the six steps of the nursing process: Assessment, Nursing Diagnosis, Outcome Identification, Planning, Implementation, and Evaluation. Documentation of these steps for individual students who have health-related issues results in the development of Individualized Health Care Plans (IHPs), a variation of nursing care plans. IHPs fulfill both administrative and clinical purposes leading to sound planning, coordination, continuity, and evaluation of care (Arnold & Silkworth, 1999).

DESCRIPTION OF ISSUE:

The number of students with special health care needs in the education setting is increasing due to advances in medicine and increased access to public education as authorized by federal and state laws. Furthermore, some chronic conditions have a potential for developing into a medical emergency and require the development of an Emergency Care Plan (ECP). The ECP is a component of an IHP, not a substitute (NASN, 1998).

Standardized IHP's, both printed and computerized, are available for the most frequent chronic health problems that occur in school-age children. These standardized care plans help promote consistency of care. In addition, the use of standardized language is being encouraged in the development of IHP's to ease communication with other team members, to assist with data collection demonstrating the school nurse contribution to student health and education outcomes, and to examine linkages between interventions and outcomes (NASN, 2001). Nevertheless, individualization is essential in order to meet the unique needs of each student.

A significant task for the school nurse, especially when assigned high student ratios and/or multiple buildings, is the determination of which students require an IHP. According to Arnold and Silkworth (1999), prioritization of students and their needs is essential and begins by identifying students whose health needs affect their daily functioning, that is, students who:

- Are medically fragile with multiple needs.
- Require lengthy health care or multiple health care contacts with the nurse or unlicensed assistive personnel during the school day.
- Have health needs that are addressed on a daily basis.
- Have health needs addressed as part of their IEP or 504 plan.

Next, prioritization is accomplished by focusing on health issues that affect safety and the student's ability to learn or that the student, family, and/or teachers perceive as priorities. Ideally, the IHP is developed collaboratively with the student, family, school staff, community, and other health providers, as appropriate (American Nurses Association, 2001). Ongoing evaluation assures a commitment to achieving measurable student outcomes. IHP's are updated as appropriate and revised when significant changes occur in the student's health status.

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RATIONALE:

As a leader of the school health team, the school nurse is responsible for first assessing the student's health status; identifying health problems that may create a barrier to educational progress, safety or well being; and developing a health care plan for management of the problems in the school setting (American Academy of Pediatrics, 2001). The use of current care standards in the development of the IHP will help assure administrators, parents, and staff that the student is properly cared for. The IHP can assist in many areas:

- Professional school nurses utilize IHPs to communicate nursing care needs to administrators, staff, students, and parents.
- The IHP will create a safer process for delegation of nursing care, supporting continuity of care.
- The IHP can serve as the health plan component of a 504 plan, and for students qualifying for special education; it can be incorporated into the Individual Education Plan when the health care issues are related to the educational needs of the student.
- The IHP will serve as legal protection by showing that proper plans and safeguards, such as an Emergency Care Plan, were in place.
- Planning and delivering care based on standardized IHPs and the use of standardized nursing language will help advance professional school nurses by affording evidence-based practice.

CONCLUSION:

It is the position of the National Association of School Nurses that students whose health needs affect their daily functioning have an IHP. It is also the position of NASN that the professional school nurse should be responsible for the writing of the IHP in collaboration with the student, family, and health care providers and for seeing that the IHP is implemented, with periodic evaluation for evidence of desired student outcomes.

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Adopted: June 1998
Revised: November 2003

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National Association of School Nurses

POSITION STATEMENT

Emergency Care Plans for Students with Special Health Care Needs

HISTORY:

Children with special health care needs (CSHCN) are those who have chronic physical, developmental, behavioral, or emotional conditions or are at risk for developing these conditions. They require health and related services of a type or amount that is usually not required by typically developing children (Committee on Pediatric Emergency Medicine, 1999). In the United States, 12.6 million children under age 18 are in this category. These children are at much greater risk for requiring emergency care (Emergency Medical Services for Children, 2001). In schools, approximately 2% of children have a serious enough disability that they require special education services or supportive care (Praeger, Zickler, & Mosca, 2002). Schools, families, and communities have the responsibility to be well prepared for prompt, safe, and individualized care in the event of a medical emergency involving these children. School nurses are involved in the development of nursing care plans for students, including Emergency Care Plans, as a part of their application of the nursing process in their school nursing practice (Schwab & Gelfman, 2001).

DESCRIPTION OF ISSUE:

In the educational setting, the Individualized Educational Plan (IEP), the 504 Plan, the Individualized Health Plan (IHP), and the Emergency Care Plan (ECP) all document preparation and planning for student success. Some students require two or more of these plans to ensure their health needs will be adequately addressed. Professional nursing standards are used in the development of IHP's and ECP's. The ECP component concerns actions to manage a specific, potential medical emergency (Arnold & Silkworth, 1999). It serves as a written, accessible, and up-to-date template for action for an individual student (Emergency Medical Services for Children, 2001). The school nurse's professional judgment is used in determining which students need an ECP and what training is needed for school personnel. Confidentiality is maintained by sharing the ECP only with school staff that have a need to know (Harrigan, 2002).

Best practice dictates that individuals who will be involved prior to or during an emergency situation with a student with special health care needs should be invited to participate in the development, implementation, and evaluation of ECP's. The child's family plays a central role in decisions of the multidisciplinary team about how the child will be managed at school and during school related activities. According to the philosophy of family-centered care, the family is the constant in its child's life (Committee on Pediatric Emergency Medicine, 1999), rather than the educational or health care system. If indicated, family can be supported to participate actively in developing its child's ECP by appropriately pairing it with someone who has greater experience with the relevant health issues needing to be addressed at school. In addition, community involvement in ECP development is critical and, when possible, should include the child's health provider, hospital emergency department, and community first responders (Emergency Medical Services for Children, 2001).

RATIONALE:

A written Emergency Care Plan, coordinated by the school nurse, ensures a plan of action is in place to maintain the student's health and safety during a life-threatening emergency (Arnold & Silkworth, 1999). In developing an ECP, the multidisciplinary team should consider the following:

- Content, format, and language that are understandable to lay people
- A system for maintaining current and ready access to information including parent and health care provider contacts (Emergency Medical Services for Children, 2001)

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- Information that is pertinent and succinct about the child's medical condition, medications, necessary supplies and equipment, and appropriate emergency intervention
- Distribution to a minimum of two appropriate school staff (AAP, 2001) in addition to the school nurse and provision for necessary training (Arnold & Silkworth, 1999)
- Prevention of an emergency as well as preparation for an emergency situation, identifying initial symptoms for concern and the response to escalating situations
- Confidentiality when determining location of ECPs
- Providing a copy of the ECP (with parent permission) to first responders and emergency department personnel (Emergency Medical Services for Children, 2001).

CONCLUSION:

It is the position of the National Association of School Nurses (NASN) that students who have special health care needs that place them at greater risk for a medical emergency should have an individualized Emergency Care Plan. NASN also believes that the registered school nurse, the student (if appropriate and he/she is developmentally able), the student's family, and health care providers, should be members of the multidisciplinary team responsible for writing and implementing the ECP. Note that the ECP should never be considered a substitute for an IHP that addresses all of a student's health needs. The Individualized Health Plan is the responsibility of the school nurse and focuses on health needs. The Emergency Care Plan flows from the IHP with special emphasis on emergency care needed for a student who may have a life-threatening episode (Harrigan, 2002) and is generally written for the purpose of directing the actions of school personnel (Schwab & Gelfman, 2001).

References/Resources:

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EVALUATION

Please take a moment to share your thoughts about Marfan Syndrome: Need-to-Know Information for the School Nurse. Your comments will help us develop new materials in the future.

On a scale of 1–5, please circle the degree to which you agree with the following statements:

Statement	1=strongly DISAGREE				5=strongly AGREE
The information is presented in clear and easy to understand language.	1	2	3	4	5
The tabs and bookmarks make the information easy to find.	1	2	3	4	5
After reading this information, I am more likely to recognize a student in need of an evaluation for Marfan syndrome.	1	2	3	4	5
This resource provides me with tools and information to make an appropriate referral.	1	2	3	4	5
The tools presented will help me to better manage the needs of an affected student in my school.	1	2	3	4	5
The "look" of the resource (photos, page layout, etc) is attractive and helps in conveying the information.	1	2	3	4	5
The CD-Rom format is convenient in allowing me to print the materials I need on a case-by-case basis.	1	2	3	4	5
The PowerPoint presentation is an effective tool to aid in educating others in my school about Marfan syndrome.	1	2	3	4	5
Have you ever had a student with Marfan syndrome in your school?	☐ Yes ☐ No ☐ Don't Know If Yes, how many?				

Please share any comments you have about the resource:

Please print and fax responses to: (516) 883-8040 Or mail to: The Marfan Foundation
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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 disorders.

- 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 disorder receives a proper diagnosis, gets the necessary treatment, and lives a long and full life.

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Credits

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