

Dear Parents and Students,

This school packet is meant to foster communication between you and the educational and social institutions serving your family. It provides information intended to present a general understanding of Loeys-Dietz syndrome and its maintenance.

The packet contains an area for you to enter important information and emergency contacts. It can be used in conjunction with a doctor's note or a personal note from you for more detailed information that is specific to your child. It is our hope that this will alleviate some of the discomforts of explaining a syndrome that has recently been defined. Portions of this packet should be updated as the medical/social needs of your child change.

Families often ask "how much information is necessary and who needs to know about the diagnosis?" This is an individual choice, but we do recommend that school settings, day care, and others who spend significant time caring for your child know about the diagnosis. They might not need to know all of the details, but it is necessary that they have basic information in case of an emergency. Thus, portions of this packet (including emergency symptoms and instructions) should be copied and given to institutions/people who care for your child.

It is our desire that every child or adolescent have full access to a rich educational environment and at the same time, is able to easily access the special accommodations that may be needed. Some children need additional assistance and/or accommodations and others do not. It is important to have good communication with your child, their teachers, principal and/or school nurses so that everyone feels comfortable and informed.

We hope you find this to be a helpful tool.

Sincerely,
The Loeys-Dietz Syndrome Foundation

sample letter

Date:

RE:

Dear Educator:

A student in your school has been diagnosed with Loeys-Dietz syndrome. The following packet of information is intended to assist communication between this student, our family and the school. Included you will find emergency care information for our Loeys-Dietz student along with some general information about Loeys-Dietz syndrome.

Thank you for taking the time to familiarize yourself with Loeys-Dietz syndrome on the behalf of this student. As a student at your school, his/her health should be kept in mind. However, while he/she may have some limitations in PE class or may need some special assistance in school, he/she should be encouraged and supported to fully engage in your school system.

Some children will not need ever need special accommodations in the classroom setting. But this student may and we encourage him/her, and you, to be honest and open about needs and school capabilities of providing any requested support.

Our hope is that this student will be provided the best learning environment in order to empower him/her to lead a full and rewarding life during the school years.

If you have any questions about this student or Loeys-Dietz syndrome, please don't hesitate to contact us. We look forward to working with you this school year.

Sincerely,

Parent's name
contact information

Emergency Vascular Information

for Students with Loeys-Dietz syndrome

Name: _____

Date of Birth: _____

Emergency Contact(s): _____

Relationship: _____

Phone: _____ Alternate Phone: _____

Allergies: _____

General Information

LDS characterized by aneurysms of the aortic root (the base portion of the aorta, the largest artery of the body that sends blood from the heart to the rest of the body) and throughout the arterial tree, tortuous (twisting) vessels, hypertelorism (widely spaced eyes) and uvula (piece of flesh that hangs in the back of the mouth) abnormalities. A variety of other craniofacial (head and face), skeletal, skin and cardiac findings have also been described.

Individuals diagnosed with Loeys Dietz syndrome have a predisposition to develop aortic tears and ruptures at sizes not thought to be dangerous to the general population or those with other connective tissue disorders.

Surgical repair of the aorta is performed long before the individual is deemed to be at high risk for aortic tear or rupture. In most cases the student should be considered to be at a low risk for a serious event and should be encouraged to participate in school activities to the extent of their ability.

In the event that this student should present with any of the following characteristics his/her emergency contact(s) should be notified immediately and contacting the local emergency room should be considered.

- weak or unusual blood pressure
- severe pain in the head, neck or abdomen
- weakness of the limbs
- symptoms of a stroke
- dizziness
- loss of consciousness
- shortness of breath
- difficulty breathing or speaking
- vision disturbances

Emergency services should be notified that the student has Loeys-Dietz syndrome and is at increased risk for tears of the arteries and that imaging should be done promptly.

Loeys-Dietz syndrome

General Facts For Schools

What is Loeys-Dietz syndrome (LDS)?

Loeys-Dietz syndrome (LDS) is a genetic disorder of connective tissue that was identified and named in 2005. At that time doctors realized that even though LDS has some features of other connective tissue disorders, it is a distinct disorder. The other disorders that share features with LDS include Marfan syndrome, Ehlers-Danlos syndrome vascular type, and Shprintzen-Goldberg syndrome.

LDS characterized by aneurysms of the aortic root (part of the aorta closest to the heart) and throughout the arterial tree, tortuous (twisting) vessels, hypertelorism (widely spaced eyes) and uvula (piece of flesh that hangs in the back of the mouth) abnormalities.

A vast variety of other craniofacial (head and face), skeletal, skin and cardiac findings have also been described, but because Loeys-Dietz syndrome manifests itself in a number of ways there are never two persons with LDS that have identical medical characteristics.

All those diagnosed with Loeys-Dietz syndrome are at significant risk for dissection and rupture of arteries at sizes that are not thought to be dangerous to the general population or to those with other connective tissue disorders.

A subset of patients have orthopedic concerns, including clubfoot or flat feet, scoliosis, or cervical spine instability that are being medically monitored. The student may have had surgery, braces, and/or limitations to address these problems.

Additionally, a subset of patients have gastrointestinal complaints, including food allergies which affect the foods LDS individuals should be exposed to.

Learning problems are not a typical finding in Loeys-Dietz syndrome. However, due to joint laxity, some individuals may have some increased difficulty with balance and fine or gross motor skills.

How is Loeys-Dietz syndrome Diagnosed?

LDS is a genetic condition, and is diagnosed through medical features and a genetic test.

How is Loeys-Dietz syndrome Managed?

Students with LDS are under the care of multiple specialists for cardiovascular, vascular, ENT, orthopedics, allergic and/or gastrointestinal concerns.

It is likely this student is on medications to lower blood pressure and treat other symptoms. These may be taken during the school day.

How does Loey-Dietz syndrome Affect Education?

Because of the complexity of the disorder, the student may have absences due to sickness and medical appointments.

We thank you in advance for your support and understanding. The student should be held to the same accountability of getting assignments finished even throughout absences.

Dietary and exercise restrictions may come into play depending on the individual student.

What Should You do if Your Student's Behavior Changes?

If teachers or administration have concerns regarding a change in mood or ability, it is important to communicate with the student's family.

To Learn More about Loey-Dietz Syndrome

Find more online at the Loey-Dietz Syndrome Foundation Web site at www.loeydietz.org.



Loeys-Dietz syndrome

General School Guidelines

How You Can Help

The most important thing you can do to help this student achieve success is to be understanding and informed of these things and offer support necessary to ensure that this student remains productive and comfortable in their educational and social environments.

Emergency Situations

In the event that this student presents with certain medical symptoms (see emergency sheet), parents should be notified immediately and contacting the local emergency room should be considered. All school staff should be aware of these circumstances and information should be made available to emergency medical staff.

Exercise Restrictions

The major goal of exercise restriction is to reduce the amount of stress imposed upon the heart and other arteries. Exercise regimens that cause a sustained elevation of either heart rate or blood pressure or bring any muscle group to exhaustion should be avoided. Ensure that communication of these restrictions is clear to all school staff that have contact with this student. (See exercise restriction sheet for more specific information.)

Prescription Medications

It may be necessary for medication to be administered at school. In addition, medications may change over time. Parents are encouraged to notify school staff when changes are made and in turn, school staff are encouraged to keep the most up to date information on file.

Corrective Devices

There may be a need for wheelchair access or student assistance when corrective devices must be worn while attending school. If necessary, the use of an aid or student buddy should be considered when available.

Allergies

Allergies have been a noted problem for young people diagnosed with Loeys Dietz syndrome. Parents are encouraged to effectively communicate the student's allergies, as well as appropriate restrictions or response actions if the student is exposed to allergens.

As a result of allergies, the student may have gastrointestinal complaints that may require easy and/or more frequent access to bathrooms.

Orthopedic Concerns

Carrying heavy backpacks can negatively impact scoliosis management. It is recommended that students limit the amount of weight carried. This can be achieved by allowing the student to have an additional set of textbooks to leave at home or allowing the use of a rolling book bag.

Because of loose joints causing pain/fatigue, some individuals may require more time for test-taking, or access to a computer for note-taking (as typing may be a more efficient and less painful means of note-taking for the student).

Medical Appointments

Students may be absent from school more frequently due to doctor appointments and hospitalizations that are very important to maintaining health. Traveling may be necessary. Students and families are encouraged to ask for school work that can be completed during long absences to ensure that the student does not fall behind in class curriculum.

Participation

As children age they become more cognizant of their physical or medical "differences." The stress of diagnosis and management may take an emotional and physical toll on this young person. Young people with Loeys Dietz syndrome are encouraged to participate to their full medical ability.

Every effort should be made to ensure that this student remains comfortable in their educational environment. If this student's behavior or interactions with other students changes it is important that the parents be notified. This will allow everyone to work together to help this student understand his/her genetic condition and achieve their full potential in a healthy environment.

During the course of a student's learning experience their individual situation may change frequently due to medical circumstance. Families, students and schools should work together on an ongoing basis to provide the best educational experience possible.

To Learn More about Loeys-Dietz Syndrome

Find more online at the Loeys-Dietz Syndrome Foundation Web site at www.loeysdietz.org.



Loeys-Dietz syndrome

Exercise Restrictions

General Information

The main goal of restricting the activity of those diagnosed with Loeys Dietz syndrome (LDS) is to **reduce the amount of stress imposed upon the heart and arteries.**

An individual with LDS should be encouraged to remain cardiovascular active. These activities work to naturally lower blood pressure and heart rate and encourage/promote cardiovascular health and fun over the lifespan.

As a general rule, many of the activities that are performed in the first few years of school are generally safe. As children get older, and the requirements and intensity of PE classes increase, there needs to be frank communication regarding the level of student participation that is safe.

If the student can maintain normal conversation during activity, this is a good indicator that activity is in a safe range.

Activities that should be Avoided

It is a general rule to recommend that those diagnosed with Loeys Dietz syndrome avoid:

- a sustained heart rate of above 140
- exercises that involve straining any muscle group to the point of exhaustion.

Discouraged activities include, but are not limited to, the following;

- competitive exercise performed to the point of exhaustion
- running sprints
- strenuous contact sports where there is a high risk of a sharp blow to the body
- isometric exercise that involves muscle straining such as weight lifting, chin ups, rope climbing, sit ups and other activities in which a muscle group is brought to exhaustion
- tumbling and gymnastics due to the risk of cervical spin instability

Activities that should be Encouraged

Most routine activities in Physical Education programs constitute part of a healthy lifestyle that will naturally lower heart rate and blood pressure over time.

In most circumstances those diagnosed with Loeys Dietz syndrome are encouraged to participate to the extent of their abilities in any activity that involves movement in the absence of muscle straining or a sustained elevation of blood pressure or heart rate.

It has been found that once students reach a certain maturity level most have the ability to use self control to stay within a safe boundary while participating in most physical education activities given proper informed supervision.

Activities that should be Encouraged, Cont.

Encouraged Activities include, but are not limited to, the following;

- regular aerobic exercise performed in moderation
- isometric exercise in extreme moderation in low repetitions, being careful to avoid straining
- swimming, bike riding, hiking, running (in moderation) and tennis, etc

Participation

If there comes a time when participation in physical education is inappropriate then every effort should be made to maximize the potential of the student by allowing them to explore other endeavors. Simply sitting on the side lines may cause a disturbance in their educational and social development and is not a viable option.

To Learn More about Loey-Dietz Syndrome

Find more online at the Loey-Dietz Syndrome Foundation Web site at www.loeydietz.org.

