



# *Shwachman-Diamond Syndrome Foundation*

*Support... Research... Cure*

Founded in 1994, our primary objectives are to provide support and education to pediatric and adult patients and their families affected by SDS (Shwachman-Diamond Syndrome). We also educate the general public, medical community and fund research toward greater understanding of this illness, improved treatment and hopefully a cure.

Our **Board of Directors** is comprised of patients and parents of patients with SDS. We meet every month to discuss ways to help families, organize family conferences, develop educational materials, and plan fundraising events.

The SDSF **Medical Advisory Board** consists of physicians from medical centers throughout the U.S. and other countries. They provide families with counseling, present at our family conferences and help to spread awareness of SDS in the medical community. They also review research grant proposals submitted to SDSF for funding.

We provide **Family Support** to all families through internet and phone communication. We supply educational materials to families with newly diagnosed individuals. We also provide a bi-annual week long family conference at Camp Sunshine in Casco, Maine. Families attending are able to get medical information with updates from our medical advisors as well as meet and share their experiences with other families. Additionally, we support the SDS Patient Registry and the bi-annual SDS International Scientific Congress.

**For more information or  
to contact us please go to:  
[www.shwachman-diamond.org](http://www.shwachman-diamond.org)**

# Shwachman-Diamond Syndrome

## First Reported in 1964

- Autosomal recessive disorder characterized by exocrine pancreatic insufficiency and bone marrow failure. The major causal genes are on chromosome 7 and chromosome 15.

## General features of SDS

- Bone marrow failure manifesting as neutropenia, intermittent or constant, anemia and/or thrombocytopenia. Some patients may present with myelodysplastic syndrome (MDS) or even acute myeloid leukemia.
- Failure to thrive, growth retardation - many patients are below 3rd percentile for height.
- Pancreatic exocrine deficiency. This improves in about 50% of patients with age.
- Skeletal abnormalities.

## Other features which may present

- Neuro developmental issues, kidney, liver disease and issues with the immune system, dental dysplasia, osteoporosis and abnormal blood sugars.

## Diagnostic criteria and testing

- Genetic testing is available, about 10% of SDS patients present clinically but have no identified genetic mutation.
- Most patients present in infancy with growth failure, feeding difficulties and/or recurrent infections.
- Documented evidence of exocrine pancreatic dysfunction and hematological abnormalities. CBCs and often bone marrow and biopsies are needed to check on cytopenias, particularly neutropenia.
- Pancreatic testing may include serum trypsinogen, isoamylase, 72 hour fecal fat balance test, elastase. Rule out of other possible causes of exocrine pancreatic dysfunction.

## Treatment and Management

- Pancreatic insufficiency is treated with enzyme replacement therapy. Fat soluble vitamin levels should be monitored regularly and vitamin replacement provided when indicated.
- Anemia and thrombocytopenia may require transfusion therapy.
- G-CSF can be considered for recurrent bacteria and/or fungal infections in the presence of severe neutropenia. The aim in long-term use of G-CSF is not to obtain normal granulocytic counts but to prevent infections.
  - Serious hematological complications which may include MDS, Aplastic Anemia and acute Leukemia. These require detailed surveillance with CBCs and bone marrow biopsies for early detection and consideration of bone marrow transplantation.

ICD9 Diagnosis 255.2 Congenital Adrenal Hyperplasia  
255.4 Adrenal Insufficiency

Letter of Medical Necessity

Emergency management of CAH briefly reviewed:

1. For fever  $> 101$ , administer 3x the usual maintenance dose of hydrocortisone, supply ample electrolyte containing fluids, call physician. This higher dose of medication should only be given for 2-3 days, then go back to the usual dose.  
-If infant/ young child cannot tolerate oral medications, try rectal hydrocortisone suppository (for infant,  $\frac{1}{2}$  of 25mg suppository every 8 hours).  
-If infant/ young child has diarrhea, administer instead 25mg IM injection of Solu-Cortef (hydrocortisone). Children aged 5-10 years should get a 50mg injection, and adolescents and adults should get 100mg.
2. If infant/child requires an injection of Solu-Cortef, it is prudent to bring the child for medical attention, since at this point IV fluids and medication are often required. Blood pressure, pulse, weight, serum electrolytes, and glucose should be measured.
3. Salt-wasting adrenal crisis can occur in infants/children with CAH who are either seriously ill, and/or have not received medication. This is a dangerous and potentially life-threatening situation requiring immediate medical attention. IF YOUR CHILD APPEARS LETHARGIC OR UNARROUSABLE GIVE THE INJECTION OF SOLU-CORTEF AND GET EMERGENCY HELP.

Prescriptions were given for injectable and rectal hydrocortisone with instructions for use; injection technique was demonstrated.

A copy of this note has been placed in the patient's chart.  
A copy of this note has been sent to the patient's parents.



in addition. Higher requirements of pancreatic enzymes should alert the clinician to the possibility of a concomitant unrelated enteropathy.

Enteric-coated enzyme preparations prevent gastric acid-peptic degradation and therefore deliver a higher concentration of enzymes to the intestine than uncoated preparations. The capsules should be swallowed whole, without chewing. If the patient cannot swallow capsules, they can be opened and the enteric-coated granules mixed with milk, juice or pureed fruit. The resulting mixture should be swallowed immediately without chewing. Pancreatin is inactivated at high temperatures, and excessive heat should be avoided when the granules are mixed with liquids or food.

#### *Vitamin supplements*

Blood levels of fat-soluble vitamins should be measured every 6 to 12 months in young children, and supplementary therapy started if values are low. It is important to ensure compliance with pancreatic enzyme supplementation, as deficiencies of these vitamins are an indirect marker of fat malabsorption.

#### *Dietary advice and surveillance*

Height and weight should be documented at every clinic visit. All patients should receive an evaluation by a dietitian. Poor appetite and behavioral feeding difficulties are common. Such children should have a careful psychology assessment and support offered to the family by a clinical psychologist.

If oral intake is suboptimal nutritional supplements should be considered. If there are ongoing concerns about poor weight gain despite adequate pancreatic enzyme replacement therapy, it may be necessary to assess the child for other causes or conditions such as gastro-esophageal reflux, food allergy and enteropathy.<sup>67</sup>

In severe cases of persistent failure to thrive or feeding difficulties, as a last resort a gastrostomy insertion can be considered to allow overnight feeding, but weaning should be attempted once the patient is stable.

#### **Treatment of dental complications**

Oral and dental problems are common in children with SDS<sup>68</sup>. Ulceration of the oral mucosa can be associated with neutropenia. The frequency and severity of the ulceration is variable. Enamel defects have been noted, in both the deciduous and permanent dentitions. Areas of faulty mineralization of the den-

tal surface can lead to decay and can be severe in some cases. Gastric acid reflux can lead to tooth surface loss or erosion. Regular dental care and appropriate advice from an early age are crucial to minimize these oral and dental problems.

#### **Treatment of bone abnormalities**

##### *Treatment and follow-up*

Bone deformities due to metaphyseal chondrodysplasia, usually located at the hips or the knees, may require orthopedic consultation and surgical interventions. Low-turnover osteoporosis may result from a primary defect in bone metabolism that is related to the bone marrow dysfunction and neutropenia. Efforts should be made to optimize general preventive measures such as nutrition and intake of fat-soluble vitamins, as well as to promote weight-bearing exercise. Supplementation with vitamin D (in addition to other fat-soluble vitamins) and calcium should be commenced if dietary intakes are not sufficient. It is presently unknown whether bisphosphonates, anti-resorptive agents used to treat postmenopausal high-turnover osteoporosis, are safe and efficacious in SDS osteoporosis. Optimal treatment for SDS osteoporosis remains to be established.

**Radiography and bone densitometry.** Assessment of bone dysplasia (Tables 2 and 3): at diagnosis, radiographic skeletal survey; follow-up based on individual clinical and radiographic findings, X-rays for detection of deformities or stress fractures (hips, knees). Assessment of osteoporosis: bone densitometry by DXA, at prepuberty (baseline study), during pubertal years, postpubertal follow-up studies based on individual findings (low BMD, vertebral compressions, multiple peripheral fractures). Caution should be exercised when interpreting DXA results in patients with SDS; small body size and delayed pubertal development affect BMD results.

**Biochemistry.** Serum 25-OH-vitamin D and plasma parathyroid hormone (PTH) should be monitored as part of routine follow-up and maintained within normal limits after the diagnosis.

#### **Neurodevelopmental consequences and support**

Deficits in cognitive abilities across numerous domains of functioning are evident in the majority of individuals with SDS at varying levels of severity



**Table 3.** Longitudinal changes in skeletal phenotype in SDS

When?	What?	Where?
Infancy and early childhood	Delayed appearance of secondary ossification centers Wide, irregular metaphyses Osteopenia, Wormian bones	Wrist, hand, femur Ribs, wrist Tubular bones, skull
Mid-childhood	Slow development of secondary ossification centers Irregularity and sclerosis of metaphyses Osteopenia	Wrist, hand, femur Femur Tubular bones, spine
Late childhood/ puberty	Irregularity, sclerosis and asymmetrical growth of metaphyses Stress fractures, deformity Compression fractures	Femur Femur Spine
Adulthood	Compression fractures	Spine

indicating heterogeneity. Parental report indicates that over 50% of children experience delayed language development.<sup>6,40</sup> Below average intellectual reasoning abilities are also evident<sup>6,40,69,70</sup> with approximately 1 in 5 meeting the diagnostic criteria for an intellectual disability (i.e., IQ < 2<sup>nd</sup> percentile).<sup>40</sup> Difficulties in visual reasoning and visual-motor integration,<sup>40,70</sup> higher order language functioning (e.g. understanding figurative expressions, knowledge of synonyms), executive problem solving and attention have also been documented.<sup>40</sup>

Significant behavioral issues are commonly reported. In a study of 32 children / adolescents (ages 6 through 17),<sup>40</sup> 19 percent had prior diagnosis of attention deficit hyperactivity disorder, pervasive developmental disorder or oppositional defiant disorder while an additional 31 percent were reported to have some combination of inattention, restless, impulsivity, and oppositional behavior. In addition, on behavioral rating scales, parents indicated a heightened frequency of attention problems (50%) and social problems (34%). The neurocognitive deficits have been found to be independent of pancreatic involvement, otitis media, having a chronic illness, family environment, and age.<sup>40</sup> Given the structural abnormalities that are evident on neuro-imaging of the brain,<sup>71-73</sup> neurocognitive and neurobehavioral issues are likely the consequences of SBDS gene dysfunction on the brain.

#### *Assessment, monitoring, and treatment*

In order to maximize ongoing development, comprehensive assessments using standardized tests and clinical observation to monitor cognitive, behav-

ioral, social, and adaptive functioning are warranted from time of diagnosis through to adulthood. Specifically, during the infancy/pre-school period (diagnosis to 4 years of age), it is advised that comprehensive developmental checklists be used so that referrals to specialists (i.e., speech and language therapist, occupational therapist, developmental pediatrician, developmental psychologist); assessment and intervention can occur at the earliest sign of possible issues. In addition, it is recommended that serial neuropsychological assessments be completed to coincide with key stages of brain maturation, namely 6-8, 11-13, and 15-17 years of age. These age groups also parallel changes in expectations in learning at school. Assessments should include evaluation of intellectual abilities, attention (working memory, sustained attention, and divided/dual attention), higher order language, visual perception, visual-motor functioning, executive skills, academic readiness/achievement, behavior, and functional independence. The identification of an individual's strengths and weaknesses, consequently leads to individualize recommendations for intervention, which are reviewed and adapted at the follow-up assessment at the next critical stage of development. Counselling for parents should parallel the neuropsychological assessments of their child to support them in enhancing interactions with, and in developing realistic expectations for, their child.

#### **Conflicts of interest**

The authors declare no conflicts of interest.

# Supporting Students with Bone Marrow Failure Diseases

A Guide for Parents & School Personnel



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Aplastic Anemia & MDS  
International Foundation

FIGHTING BONE MARROW FAILURE DISEASES  
THROUGH PATIENT SUPPORT AND  
RESEARCH SINCE 1983

## We are here to help!

The Aplastic Anemia & MDS International Foundation is an independent nonprofit organization. Our mission is to support patients, families, and caregivers coping with:

- Aplastic anemia
- MDS (myelodysplastic syndromes)
- PNH (paroxysmal nocturnal hemoglobinuria)
- Related bone marrow failure diseases



This publication provides basic information about bone marrow failure diseases and their symptoms, along with tips about how to best support the student with a bone marrow failure disease in the school environment. Although the Aplastic Anemia & MDS International Foundation strives to provide the most accurate and up-to-date information, and this information has been thoroughly reviewed by experts, it does not warrant or guarantee this information. Patients should always seek medical advice from a qualified physician and discuss these materials, individual questions, and concerns with their physician.

**For more information, call us at (800) 747-2820, or visit us online at [www.AAMDS.org](http://www.AAMDS.org).**



Aplastic Anemia & MDS  
International Foundation

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# Introduction

School personnel, including school nurses, teachers, and school administrators, can have a significant impact on the well-being of students with chronic and serious illnesses, including those with bone marrow failure diseases. By working together with parents and healthcare providers, they can create a positive, safe and supportive atmosphere for the student. This type of coordinated care facilitates understanding and enhances awareness and sensitivity by everyone in the school setting, including other students. By reinforcing the need for compassion, patience, and understanding for chronic and serious illnesses, school personnel will create a positive learning environment.

The first step for creating a safe and supportive school environment is the creation of a coordinated care plan. This typically involves bringing everyone involved with the student into a meeting to discuss the student's medical,

emotional, and learning needs. At this meeting it is also critical to clarify the roles and responsibilities of each person. Appropriate expectations and possible limitations the student may face should also be discussed. The benefits of developing this type of coordinated care plan for the student include:

- Better school attendance
- Fewer symptoms, including better alertness
- Fewer restrictions on participation in physical activities
- Fewer emergencies

This guide provides you with the information you need to know to create a positive school environment for the student with a bone marrow failure disease. The first section includes basic disease and treatment information about the three main types of acquired bone marrow failures diseases: aplastic anemia, myelodysplastic syndromes (MDS), and paroxysmal nocturnal hemoglobinuria (PNH). This is followed by an overview of the physical, emotional, and learning challenges a student may face. The third section provides tips and guidance for supporting positive re-entry into school. A resource section is provided at the end, with recommended reading and trustworthy Web sites where you can find additional information about bone marrow failure diseases, as well as tips and advice for creating a positive school environment.



answers  
support &  
hope

# About Bone Marrow Failure Diseases

## What is a bone marrow failure disease?

In simple terms, a bone marrow failure disease is when something stops a person's bone marrow from creating the right amount of healthy blood cells. There can be many reasons for this. Sometimes one of the blood forming stem cells in the bone marrow becomes defective and creates defective blood cells. Sometimes a person's body attacks and kills blood forming stem cells so they can't make blood cells. For the most part, doctors don't know what causes bone marrow failure diseases.

In order to understand bone marrow failure diseases better, it's important to first know certain facts about blood.

## What is blood made of?

Blood is made of blood cells floating in plasma. The plasma is mostly made of water with chemicals in it. These chemicals include proteins, hormones, minerals, and vitamins.

## What are the three basic types of blood cells?

1. Red blood cells are also called erythrocytes (i-RITH-ruh-sites). They make up almost half of blood. Red blood cells are filled with hemoglobin (HEE-muh-gloe-bun). That's a protein that picks up oxygen in the lungs and brings it to cells all around the body.
2. White blood cells are also called leukocytes (LEW-kuh-sites). They fight disease and infection by attacking and killing germs that get into the body. There are several kinds of white blood cells, each of which fights a different kind of germ.
3. Platelets are also called thrombocytes (THROM-buh-sites). They are small pieces of cells that help blood clot and stop bleeding.

## How are blood cells formed?

The process of making blood cells is called hematopoiesis (hi-mat-uh-poy-EE-suss). Blood cells are made in the bone marrow. That's a spongy tissue located inside some bones. It contains young parent cells called stem cells.

These blood-forming stem cells can grow into all three types of blood cells. They make copies of (clone) themselves, and they also produce mature blood cells.

When blood cells are fully mature and functional, they leave the bone marrow and enter the blood. Healthy people have enough stem cells to keep making all the blood cells they need every day.

## What are the symptoms of low blood counts?

Most of the symptoms experienced by people with bone marrow failure diseases are caused by low blood counts. Specific symptoms depend on the type of blood cell affected.

A person with a low red blood cell count may:

- Feel a little tired or very tired
- Feel less alert or have trouble concentrating
- Have a loss of appetite or lose weight
- Have paler-than-normal skin
- Have trouble breathing
- Have rapid heartbeat
- Have reduced ability to exercise or climb stairs



A person with a low white blood cell count may:

- Have repeated fevers and infections
- Get bladder infections that may make it painful to pass urine, or make you urinate more often
- Get lung infections that cause coughing and difficulty breathing
- Get mouth sores
- Get sinus infections and a stuffy nose
- Get skin infections

A person with a low platelet count may:

- Bruise or bleed more easily – even from minor scrapes and bumps
- Get heavy menstrual periods
- Get nose bleeds
- Get tiny, flat red spots under your skin, which are caused by bleeding; These spots are called petechiae (puh-TEE-kee-ie).
- Have bleeding gums, especially after dental work or from brushing your teeth.

### **How are bone marrow failure diseases diagnosed?**

Diagnosis is made through blood tests and a bone marrow biopsy. Once diagnosed, treatment and evaluation is given under the care of a hematologist.

### **What are common symptoms of bone marrow failure diseases?**

School personnel or fellow students may be among the first to notice a student's symptoms. These can include complaining of fatigue, experiencing nosebleeds or having excessive bruising on various body parts. The most common symptoms are:

- Bleeding from a cut or other wound that does not stop

- Small red or purplish spots like a skin rash (these are called “petechiae” and result from very small hemorrhages)
- Shortness of breath
- Chronic fatigue
- Decreased alertness
- Decreased attention span
- Lethargy
- Pale skin
- Ringing or buzzing in the ears
- Frequent colds, sore throats, or other infections

It is important to note that these symptoms can also signal many other illnesses. Only a doctor can make diagnosis of a bone marrow failure disease. A diagnosis can only be made after examining the student's blood and bone marrow.

### **What is aplastic anemia?**

Aplastic anemia is a non-contagious, rare disease that causes the bone marrow to stop making all the blood cells the body needs. It is newly diagnosed in 600 to 900 people every year in the United States. It can strike people of any age, race and gender, but it is more common among children, teenagers, and young adults.

There are two types of aplastic anemia: acquired and hereditary. Acquired aplastic anemia is much more common than hereditary aplastic anemia. In patients with acquired aplastic anemia, researchers believe that a patient's own immune system attacks their bone marrow stem cells. Hereditary aplastic anemia is usually diagnosed in childhood and is less common than acquired aplastic anemia. People who develop hereditary aplastic anemia usually have other genetic or developmental abnormalities.



In about 75 out of 100 of cases of acquired aplastic anemia, there is no known cause. For the remainder of patients, it can be linked to one of several causes, including:

- Medications, such as anti-inflammatory drugs, anti-seizure medications and treatments for lupus and rheumatoid arthritis
- Exposure to certain toxins such as pesticides, arsenic, or benzene
- Radiation and chemotherapy used to treat cancer

The symptoms of aplastic anemia are caused by low blood counts. They can include fatigue, increased bleeding, bruising and susceptibility to infections, among others. (See “What are the symptoms of low blood counts?” on page 5 for more information)

There are a number of treatments for aplastic anemia:

- Blood transfusions may be given when blood counts are low. This temporarily replaces the cells the bone marrow has stopped producing.
- Immunosuppressive drug therapy can suppress the immune system’s attack against the bone marrow, permitting it to recover and begin producing cells again. About 70 out of 100 patients can be successfully treated with this therapy, but it can take several months to respond. The most common immunosuppressive drugs used to treat aplastic anemia are ATG (antithymocyte globulin) and cyclosporine.
- Growth factors are naturally occurring chemicals (proteins) in the body that cause bone marrow to make more blood cells. Man-made growth factors can be used to increase blood cell production and may be prescribed at any point during treatment.
- Bone marrow/stem cell transplant is considered the only cure for aplastic anemia. Transplants are most successful in younger patients with a related donor whose tissue type matches their own.



Aplastic anemia is a serious illness that requires immediate medical attention. Once considered a fatal illness, with standard treatments such as bonemarrowtransplantsandimmunosuppressive drug therapy, 70 to 90 out of 100 patients can now be treated successfully.


For more information on aplastic anemia, contact the Aplastic Anemia & MDS International Foundation, and order a free booklet titled Your Guide to Understanding Aplastic Anemia. Also visit [www.AAMDS.org](http://www.AAMDS.org).

### **What is paroxysmal nocturnal hemoglobinuria (PNH)?**

PNH is an ultra-rare and serious blood disease that causes red blood cells to break apart. Doctors call this breaking apart hemolysis (hi-MOL-uh-suss). It happens because blood cells are missing a protein that protects them from the body’s immune system.

Fewer than 500 people are diagnosed with PNH in the United States each year. PNH can also occur with aplastic anemia or MDS. It is rare to see PNH in very young children, but it has been seen in teenagers.

When red blood cells break apart, the hemoglobin (HEE-muh-glo-bun) inside the blood cells is released. Hemoglobin is the red part of red blood cells. Its job is to carry oxygen around your body. So patients with PNH will often have anemia and will experience fatigue and decreased alertness. The release of hemoglobin causes other symptoms of PNH. These include back pain, abdominal discomfort, trouble swallowing and blood clots.



The name “paroxysmal nocturnal hemoglobinuria” comes from:

- Paroxysmal means sudden and irregular
- Nocturnal means at night
- Hemoglobinuria means hemoglobin in urine. Hemoglobin is the red part of red blood cells. It makes urine look dark

So the name “paroxysmal nocturnal hemoglobinuria” means sudden, irregular episodes of passing dark colored urine, especially at night or in the early morning. It is important to note, however, that many people with PNH do not have this symptom.

Treatment of PNH depends on the severity of the disease. It may include blood transfusions, growth factors, blood thinners, immunosuppressive drug therapy or bone marrow/stem cell transplantation. A drug named eculizumab (Soliris®) is also approved by the FDA to treat PNH.

For more information on PNH, contact the Aplastic Anemia & MDS International Foundation and order a free booklet titled Your Guide to Understanding PNH. Also visit [www.AAMDS.org](http://www.AAMDS.org).

### **What are myelodysplastic syndromes (MDS)?**

MDS is a group of disorders in which a person’s bone marrow does not work well, and the bone marrow cells fail to make enough healthy blood cells. People with MDS can lack the right amount of red blood cells, white blood cells, and platelets. Children rarely get this disease.

The disease happens because the bone marrow cells do not develop into mature blood cells. Instead, these blood cells stay within the bone marrow in an immature state. The symptoms

and the course of MDS may vary greatly from person to person. These differences depend on which blood cells are affected.

All people with MDS have 2 things in common:

- They have a low blood cell count for at least 1 blood cell type. This is called cytopenia (sie-toe-PEE-nee-uh).
- Their bone marrow and blood contain blood cells with an abnormal shape, size, or look.

Between 10,000 and 15,000 people are newly diagnosed with MDS each year. MDS occurs mostly in people over the age of 60.

MDS is classified into separate subtypes according to how the disease manifests itself in a patient’s blood and bone marrow.

Treatment for MDS depends on the type of MDS, the severity of the disease and on any specific genetic abnormalities found in the blood cells. Treatments include blood transfusions, growth factors, immunosuppressive drug therapy and bone marrow/stem cell transplantation. In addition, there are three drugs currently approved by the FDA to treat MDS. These drugs include lenalidimide (Revlimid®), azacitidine (Vidaza®) and decitabine (Dacogen®). A bone marrow/stem cell transplant is the only cure for MDS.

For more information on MDS, contact the Aplastic Anemia & MDS International Foundation and order a free booklet titled Your Guide to Understanding MDS. Also visit [www.AAMDS.org](http://www.AAMDS.org).

# Understanding the Physical and Emotional Impact on a Student



## What are the physical problems the student may face?

There are many physical problems that can affect a student with a bone marrow failure disease. School personnel, especially school nurses, should always be kept informed of the student's condition.

Physical problems can result from the disease itself or from the treatments:

- A low red blood cell count can cause the student to feel very tired and lack energy for normal activities. They may have limited ability to participate in physical activities because of fatigue or muscle weakness. Consider reducing requirements for participation in physical activities such as gym and sports and providing a place for the student to rest during the day.
- A low white blood cell count puts the student at risk for infection and certain precautions must be taken. If the counts are very low, the student may need to be kept home to avoid colds, flu, and other infections. In some cases a special diet called a neutropenic diet may be required. This diet reduces exposure to food-borne illnesses and bacteria. It involves avoiding raw foods, including unwashed fruits and vegetables, salad bars, sushi, aged cheeses, and certain other foods.

- A low platelet count means the student may bruise or bleed easily. At school, reducing physical activity can help protect the student. Avoid contact sports, or activities that may lead to getting banged or cut. Among younger students, playground activities may need to be restricted to avoid injuries.

Students may also experience changes in their physical appearance as a result of their disease and/or treatment. These might include weight gain or weight loss, puffy face, hair loss or excessive hair growth, pale skin, or large bruises.

These physical changes can also have an emotional impact on the student. School personnel should be alert to teasing or bullying that may result from the student looking different.

## What are the emotional issues the student may face?

Learning to live with a chronic illness can be extremely difficult, even for the most well-adjusted child and his/her family. The lives of family members are likely to change because dealing with a bone marrow failure disease requires schedules to be adjusted for frequent doctor visits and planned or unexpected hospital stays. Parents must be vigilant about cleanliness in order to protect their child. Diets may have to change.

Any underlying mental health issues may become more apparent or worsen during the stress of dealing with these diseases. Parents and school personnel should watch for behavioral changes in the student, including increased anxiety, depression, aggression or social isolation. Different behavioral reactions may occur with different age groups.



## Elementary age

Early elementary-aged children are beginning to understand that they are part of a larger environment, but they still use their imagination for answers to overwhelming questions. They may believe their illness is “their fault.” They need to have a clear and simple understanding of their illness and what to expect. Older elementary-aged children may have a better understanding of their condition, but will be more aware that they are different from their peers. They may worry about being left out when they miss school or if their activities are restricted. Young children with chronic illnesses may also be subject to teasing or bullying.

## Teens

Adolescents are developing their own identities, gaining independence, and taking on greater responsibilities. They may be much more involved in decisions regarding their treatments. However, compliance can be an issue with some teens. Some may neglect to take their medications consistently, thus endangering their health.

Teens may also have body image issues related to the symptoms of their disease or the side effects of the medication. Things like hair loss, excessive body hair growth, weight gain and the need to wear a mask to prevent infections may make a teen even more self-conscious. Counseling with a social worker or psychologist may be important for some teens.

## How school attendance may be affected

The symptoms and treatment of bone marrow failure diseases can keep a student out of school for a long time. Frequent absences for blood transfusions and medical tests are common. In addition, the student may have frequent sick days. Low red blood cell counts can make it hard for a student to learn and concentrate. Low white blood cell counts can result in frequent infections.



# Parents and Schools Working Together:

## Tips for Successful School Re-Entry

There are many things you can do to support a successful and positive re-entry into school for a student with a bone marrow failure disease. Families and school personnel, including classroom teachers, nurses, aides and others who are in contact with the student each play an important role. This section provides tips for supporting a smooth transition.

### TIPS FOR PARENTS AND FAMILIES

- ☐ Arrange a meeting with school administrators, teachers, nurses, and guidance counselors to discuss your student's diagnosis and special needs. Provide medical information about your child, including treatment schedules and doctors appointments. Be sure the school has written authorizations to administer medications and emergency treatment plans approved by your child's doctor.
- ☐ Authorize the appropriate exchange of information between the school and your child's doctor.
- ☐ Set up regular communications with the school to inform them of any changes in your child's condition or special needs.
- ☐ Familiarize yourself with federal laws (Section 504 of the Rehabilitation Act, Americans with Disabilities Act, and Individuals with Disabilities Educational Act) that provide protections for students with disabilities. Public schools are required to provide a written plan, called an Individualized Education Plan (IEP), on how to provide education to a student with a disability. Be sure to schedule regular review meetings with school personnel regarding the educational plan for your child.
- ☐ Every public school has a coordinator for special educational plans. And some hospitals have educational consultants who can help you coordinate your child's care at school.
- ☐ Help your child develop age-appropriate skills for taking care of himself.
- ☐ With the school's and your child's permission, talk to your child's friends and classmates about the diagnosis. Give them age-appropriate information. The school nurse and counselor may be able to help you talk to classmates. Older students may prefer to have these discussions with their peers without your participation.
- ☐ Ask your child about any concerns regarding returning to school and ways that you can help. Reassure your child that these concerns are normal and that he/she has your support and the support of everyone at the school.
- ☐ Consider acquiring a medical ID bracelet for your child to wear or providing identification and key medical information on a card that stays in the student's backpack. This safeguard could be important if your child needs medical attention when no knowledgeable staff is available.
- ☐ If you rely on school buses for transportation, be sure that your child's regular bus driver has basic emergency contact information for your student.
- ☐ Consider contacting your local 911 communications facility and asking them to make a note about your child's rare disease under your address and telephone number. In the event of an emergency, this information could help medical response teams.



# Parents and Schools Working Together: Tips for Successful School Re-Entry

## TIPS FOR SCHOOL NURSES

- ☐ Become informed about bone marrow failure diseases to help facilitate discussions between school personnel and families. Order and read Your Guide to Understanding Aplastic Anemia, MDS or PNH.
- ☐ Visit the Aplastic Anemia & MDS International Foundation's Web site at [www.AAMDS.org](http://www.AAMDS.org) and the Online Learning Center at [www.AAMDS.org/Learn](http://www.AAMDS.org/Learn). There you can read the most up-to-date information on bone marrow failure diseases and view videos of experts talking about these diseases and their treatment.
- ☐ Make sure you have up-to-date information on the student's medical status, blood counts, and treatment.
- ☐ Know what medications the student is taking, when the medication is to be given, and the common side effects of each medication.
- ☐ Know what symptoms constitute a medical emergency and develop or review a plan with the student's doctor for handling such emergencies.
- ☐ Discuss with the parents any possible problems with infectious diseases that may come from other students, including chicken pox, flu, and other viruses.

## TIPS FOR SCHOOL ADMINISTRATORS

- ☐ Be sure that school staff who will be responsible for the student's care at school and school-related activities fully understand the student's health and learning needs and are properly trained to provide appropriate support.
- ☐ Conduct periodic review meetings with all parties to be sure the school has current and accurate information about the student's condition and needs, and to keep the lines of communication open with parents and the student's health care providers.
- ☐ Encourage teachers and other school personnel to create an environment that views a student with a bone marrow failure disease the same as other students except for the need to respond to health issues.



# Parents and Schools Working Together: Tips for Successful School Re-Entry

## TIPS FOR CLASSROOM TEACHERS

- ☐ Be aware of social, emotional, psychological, and health-related needs of the student who is dealing with bone marrow failure disease. Chronic illness does not need to stand in the way of education.
- ☐ Be aware that a student with a bone marrow failure disease often looks healthy despite their illness.
- ☐ Be aware of scheduled treatments and any resulting absences from school.
- ☐ Share information about any behavioral changes you see in the student with the family and other school personnel. It is quite common for these students to express anxiety, depression, or—in younger students—regression in your classroom. Changes in memory, attention, and perception should also be shared with all parties concerned with the care of the student.
- ☐ Expect questions from other students and be prepared to provide appropriate information. Be aware of state and federal regulations which protect the student's privacy. You will find a suggested reading list to facilitate classroom discussion at the end of this publication.
- ☐ Help the student continue studies when away from the school by providing work that can be done outside the classroom.
- ☐ Maintain regular communication with the student and the family. Working collaboratively with the family, home tutor, or hospital school program will make the transition back to school much smoother for the student.



Anyone tasked with helping a student with a bone marrow failure disease, or other life threatening illness, may experience unexpected thoughts and feelings. These may include worries about the possible death of the student and its impact on other students. If this happens, it may be helpful to speak with a physician, social worker, or nurse about your thoughts and feelings.

## What are other ways teachers can help?

**IMPORTANT NOTE:** *The suggestions below should only be implemented after discussing them thoroughly with the student's parents and ensuring that they are in alignment with school policy.*

**To help support the young student who is returning to school, teachers can initiate discussion in the classroom about health-related topics like these:**

- The need for everyone to see his or her doctor regularly
- Immunizations, proper diet, adequate exercise, and rest contribute to good overall health
- Physicians and nurses are people who help you stay well and take care of you when you are sick
- Hospitals are places where people can go when they need special care
- Most people will have some kind of illness in their life
- No one is at fault when serious illness strikes and being sick does not mean you did something bad or that you're being punished

**Teachers might also have classmates imagine they are ill and ask themselves the following questions:**

- How would I like people to treat me?
- How would I feel if I were treated differently because of my illness?
- How would I want my friends to help me when I am sick?

The student who returns to school can also explain their illness to their peers through the use of books, dramatic plays, or simple explanations with help from their parents or the school nurse.

**Prolonged absences may make the student feel lonely and out of touch with their peers. Here are ways the teacher and the student's classmates can help:**

- Keep in touch with the student and he/she family through cards, letters, and drawings.
- A video recording of the student's classmates is one of the best ways to help keep the student involved with the school. The video may include personal messages and group songs from both classmates and faculty.
- Have classmates call the student with homework assignments and information about school events.
- Let the student know their desk, cubbyhole, or locker is being saved for their return.
- Plan a special welcome for the student when he/she returns to school.

It may also help ensure a successful re-entry into school if the parents send a video message from their child back to the school. Classmates may be less surprised at any physical changes the student has undergone if they have the opportunity to see their friend in a video. A video from the student can also give the teacher an opportunity to initiate a classroom discussion about the student and his illness.

## How Can the School Help When a Student is Terminally Ill?

While the emphasis of this document is on life and living, some patients do die as a result of their bone marrow failure disease.

Unfortunately, this topic is rarely discussed. In the case of terminal illness, it is important for both the school and the teacher to be notified. Even in the final weeks of life, school can remain a rewarding experience for the student. As death approaches, classmates may want to say goodbye through cards and letters.

After a student dies, classmates may express their grief in a variety of ways. Feelings of loss should be acknowledged, but no attempt should be made to force classmates to talk about the death or to deal with grief before they are ready. Organizing a class project, such as planting a tree or collecting money to donate to charity, may help with the healing process. As is true for adults, young people will deal with grief at their own pace and in their own way.

answers  
support &  
hope





## Conclusion

We trust, now that you have read this guide, you have the basic tools and information necessary to effectively support a student with a bone marrow failure disease in the school. The information helps your understanding of what you can do to create a safe and supportive learning environment and where you can go to get additional information and guidance. The next step is to put a clear plan in place and follow that plan.

If you haven't done so yet, set up a meeting including the student's parents, school nurse, teachers, and school administrators. During this meeting, you should:

- Make sure each person has a copy of this guide and has read it before the meeting
- Review and discuss what you learned from this guide
- Identify any gaps in knowledge you may have
- Discuss and write down a plan for supporting the student on daily basis, if a crisis should occur, and during school re-entry, if appropriate

**Know that you are not alone—the Aplastic Anemia & MDS International Foundation is always there to help.**

If you would like additional guidance, have questions about bone marrow failure diseases and their treatment, or would like to talk with another parent who has gone through this process of supporting a student in the school, we can help. We wish you all the best, and we know that if you follow the tips in this guide, and your action plan, you will indeed create a positive, safe and supportive school environment for the student living with a bone marrow failure disease.

# Recommended Resources

## Books for children

*The Fall of Freddie the Leaf*, by Leo Buscaglia, 30pp, Slack Inc, 1982. Story of Freddie the leaf as he lives through all of the seasons, ending with the realization that death is a part of life. For Ages 4-8.

*Henry and the White Wolf*, by Tim & Tyler Karu, 32pp, Workman Publ Co, 2000. Henry, a very sick little hedgehog, receives treatment from the White Wolf that makes him feel even worse but eventually cures him, and through the ordeal, he holds a stone to help him stay strong and brave.

*Talking to a Child with Bone Marrow Disease*, by Marilyn Baker, 32pp, Aplastic Anemia & MDS International Foundation, 2010. This read-aloud picture book is written to encourage children to ask questions and express their feelings about their disease.

*When Molly Was in the Hospital: A Book for Brothers and Sisters of Hospitalized Children*, by Debbie Duncan, 32pp, Rayve Prod, 1994. For ages 3-12. Anna's little sister is very ill and she learns to come to terms with it.

*Why, Charlie Brown, Why?: A Story About What Happens When a Friend Is Very Ill*, by Charles Schulz, 64pp, Ballantine Books, 2002. When a friend of the Peanuts gang is diagnosed with leukemia, they all have different reactions. The book follows her treatment and recovery.

*Young People and Chronic Illness*, by Kelly Huegel, 176pp, Free Spirit Pub, 1998. For ages 9-12. This book is divided into stories of children living with chronic illness and strategies for managing these conditions.

## Books for teens

*Easy for You to Say: Q and As for Teens living with Chronic Illness or Disabilities*, by Miriam Kaufman, 285 pp, Firefly Books, 2005. Practical and frank topics geared exclusively for teens with chronic illness or disabilities.

*It's Not About the Bike: My Journey Back to Life*, by Lance Armstrong, 304 pp, Berkley Trade, 2001. Inspirational story of famed champion overcoming cancer.

## Books for parents, teachers & nurses

*Healing Images for Children: Teaching Relaxation and guided Imagery to Children Facing Cancer and Other Serious Illnesses*, by Nancy Klein, 280pp, Inner Coaching, 2001.

*Parenting Children With Health Issues: Essential Tools, Tips, and Tactics for Raising Kids With Chronic Illness, Medical Conditions, and Special Healthcare Needs*, by Foster W. Cline and Lisa Greene, Love and Logic Press, 2007.

# Recommended Resources

## Online resources

*Aplastic Anemia & MDS International Foundation (AA&MDSIF)*  
**www.AAMDS.org**

AA&MDSIF provides information and support for patients and their family members. Call (800) 747-2820 for your free information packet on myelodysplastic syndromes, aplastic anemia, or PNH, or to talk to a patient educator. You can also visit our Online Learning Center at [www.AAMDS.org/learn](http://www.AAMDS.org/learn)



*American Academy of Pediatrics*  
**www.aap.org**

This site provides general information on promoting health and well being for children. You can also search for articles on specific bone marrow failure diseases.

*National Library of Medicine (NLM)*  
**www.nlm.gov**

NLM is the world's largest medical library. Their Web site provides comprehensive information on various bone marrow failure diseases.

*National Institutes of Health (NIH)*  
**www.nih.gov**

NIH is the nation's leading medical research agency. They provide important medical discoveries that improve people's health and save lives. NIH scientists investigate ways to prevent disease as well as the causes, treatments, and even cures for common and rare diseases. On this Web site you can search for aplastic anemia, MDS, or PNH and see the research they are doing now.

*Girlshealth.gov*

**www.girlshealth.gov**

This site is sponsored by the National Women's Health Information Center (NWHIC) of the U.S. Department of Health and Human Services. It is written by and for young women in their teens. A section under "Illness and Disability" addresses many questions and concerns expressed in this age group about hospital life, school, friends, family, and dating.

*Learning Disabilities Association of America (LDA)*  
**www.ldaamerica.org**

(412) 341-1515

The LDA provides support to people with learning disabilities, their parents, teachers, and other professionals. An online course is available for parents describing how to get the services your child needs under the Individuals with Disabilities Education Act (IDEA).

## More Ways to Get Help

The Aplastic Anemia & MDS International Foundation (AA&MDSIF) is here to help. We provide the following services:

- Personalized support through patient educators
- Free educational materials on aplastic anemia, MDS and PNH
- Online Learning Center with presentations by medical experts
- Peer Support Network
- Patient and family conferences
- Print and electronic newsletters
- Clinical trials information

### Contact us today. Here's how:

**Call us:** (800) 747-2820

**Email us:** [help@aamds.org](mailto:help@aamds.org)

**Go to our Web site:** [www.AAMDS.org](http://www.AAMDS.org)

**Remember – you are not alone.  
We are standing by to support you in any way we can.**



Aplastic Anemia & MDS  
International Foundation

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FIGHTING BONE MARROW FAILURE DISEASES  
THROUGH PATIENT SUPPORT AND  
RESEARCH SINCE 1983



answers  
support &  
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