

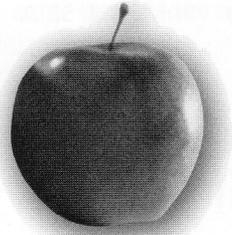
Alagille Syndrome In The Classroom



- Symptoms
- Care
- Solutions



Alagille
Syndrome
Alliance



*A Guide to Understanding
the Needs of AGS Students*

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Since the body does not digest food properly, many important nutrients are not being absorbed. The child with AGS, therefore, should take vitamins to aid in proper nutrition.

- Vitamin K (Phytonadione) A fat-soluble vitamin absorbed by the intestine and stored in the liver that is necessary for the function of clotting factors in blood coagulation.
- Vitamin E (Alpha Tocopherol Polyethylene Glycol Succinate, TPGS, Liqui-E) Antioxidant that protects red blood cells from hemolysis.
- Vitamin A (Aquasol A) Needed for bone development, growth, visual adaptation to darkness, testicular and ovarian function, and as a cofactor in many biochemical processes.
- Vitamin D (ergocalciferol = Calciferol, Drisdol) Stimulates the absorption of calcium and phosphate from the small intestine and promotes release of calcium from bone into blood.

Hospitalizations

Need for hospitalization will vary greatly between children with AGS because the condition has such a wide spectrum of organ system involvement. The most common reasons for admission to the hospital would be for testing, such as liver biopsies and cardiac catheterizations which may require a one to two day absence. The child with AGS may also break major bones (i.e. femur) which may require immobilization casts and may mandate a prolonged absence from school for that reason. Rarely a child may have such severe liver disease that he requires prolonged hospitalization for a liver transplant. In all of these cases efforts should be made to continue to supply the child with her classwork so that she may have the option of staying current with her classmates. In some cases, tutors may be needed.

Communication

The most important thing to remember with an AGS child is to communicate with the family. Parents and teachers can help each other

- Phenobarbitol (Barbita, Luminal) Mainly used as an anticonvulsant, but is used in patients with liver disease to induce liver enzymes in order to help lower bilirubin and improve itching slightly. Main side effect: sedation, sleepiness.
- Antihistamine (Benadryl = diphenhydramine or Atarax = hydroxyzine) Medications which block the action of histamine in the body. Histamine is one of the mediators of "itchiness". Used to improve itchiness and for mild sedation. Main side effects: drowsiness or unusual irritability and moodiness.

Pancreatic Enzymes in AGS

Many AGS patients have pancreatic insufficiency and require enzyme supplementation therapy. Pancreatic insufficiency results from the pancreas not producing certain enzymes which aid in digestion. Lack of these enzymes results in malabsorption or failure of the intestines to absorb the nutrients from food. In children with malabsorption the nutrients from their food pass through their body unabsorbed and exit in their stool. The child, therefore, has difficulty gaining weight and maintaining adequate nutrition despite a normal or large intake of food. Teachers should be aware that these children may require extra or urgent bathroom privileges.

The treatment for pancreatic insufficiency is to supplement each meal with pancreatic enzymes taken in capsules. In order for the food to properly mix with the enzymes, the capsules must be taken with the meal. Depending on the degree of pancreatic insufficiency, the child may take 1 to 4 capsules with meals and 1 to 2 capsules with snacks.. There are many different types of pancreatic enzymes with names such as PANCRECARB[®], Pancrease[®], Creon[®], Ultrase[®] or Viokase[®]. The child will require dosing of this medication with all meals and snacks including those eaten in school.

Vitamins

Since many AGS children experience impaired digestion related to improper liver function, there is a considerable issue with growth and weight gain.

stools, but the benefits from the calories and vitamins in the fat that is absorbed usually leads to the recommendation that children not be put on a low-fat diet.

Medications

Most school systems have procedures in place for children taking medications during school hours. Discussions between the health care coordinator for the school, the parents, and possibly the child's physician, should help to make the procedure as easy as possible for the student and as unintrusive as possible for their class.

Most medications that children with AGS are taking are intended to improve bile flow and lower bile acids in order to improve their jaundice and itching, and to supplement their vitamin levels:

- Ursodeoxycholic acid (Actigall, Urso, Ursodiol) A bile acid that improves bile flow. May improve itching by improving bile flow. Main side effect: diarrhea.
- Cholestyramine (Questran or Colestipol) A resin which binds mainly bile acids, cholesterol, and some drugs and vitamins. The drug is used to remove bile acids from the system and lower cholesterol by stimulating increased production of bile acids from a cholesterol backbone. Powder which is mixed with fluid or applesauce. Main side effect: rash, constipation, abdominal distention.
- Rifampin (Rifampicin) An antibiotic originally developed for therapy of tuberculosis. It has been found to improve itching in patients with liver disease possibly by altering uptake of bile salts by liver cells. Main side effect: colors body fluids such as tears, sweat and urine to a red-orange color; may cause drowsiness and mild nausea.

vitamins, and maintaining normal growth and development, often through supplemental feeding or in some cases formula feeding through a nasogastric tube or direct gastric tube on the abdominal wall. It is most important to monitor a child's growth, development and nutritional status.

Listed below are the major liver functions that doctors and nutritionists monitor. It is important to note that the severity of the effects described in this section vary from child to child and the advice of a doctor or nutritionist should be sought to determine if and what treatment may be necessary.

Absorption of fat soluble vitamins. It usually is not difficult to feed adequate numbers of calories to children with AGS. However, getting adequate numbers of calories absorbed is sometimes a great challenge. Some children may malabsorb 40% of the calories they eat. For these children, it may not be possible to eat the extra needed calories, and the insertion of a feeding tube may help. This tube allows the delivery of large quantities of nutrients overnight, including formulas that may not seem very tasty to infants and young children. Nasogastric tubes are temporary and can be removed at any time. A direct gastric tube (G-Tube) is placed for children with long-term needs. Blood tests are used to diagnose vitamin deficiencies and large oral vitamin doses usually can correct any problems that arise.

Bile flow. Because bile flow from the liver to the intestine is slow in AGS, medications designed to increase bile flow are prescribed. These medications also may relieve itching caused by buildup of bile in the blood and skin.

Blood cholesterol. The child with AGS may have elevated blood cholesterol levels, which can lead to small yellow deposits of cholesterol (xanthomas) on the skin of knees, elbows, palms, eyelids, and other body surfaces that are frequently rubbed. Elevations in blood cholesterol respond to the medications used to increase bile flow. Lowering blood cholesterol usually causes skin deposits to improve. Although these are unsightly, they are almost never associated with any dangerous symptoms.

Digestion of dietary fat. Reduced bile flow can lead to poor digestion of dietary fat. MCT (medium-chain triglyceride), a type of fat that can be well digested despite reduced bile flow, often is prescribed for infants with AGS. Later in childhood, eating foods containing fat may lead to looser, greasy

than children with some other liver disorders that may be present at the same age. Many adults with AGS are leading normal lives.

A Student with AGS may Exhibit these Symptoms...

A child with AGS can be mildly or severely affected. The most common problem seen in AGS children is severe itching that is not relieved by scratching or other traditional means. The child may display erratic behavior in the classroom as he or she tries to get comfortable. The child's attention span may be shortened as he tries to deal with his physical limitations. It is very possible for this behavior to mimic other illnesses such as ADD/ADHD.

Depending on the severity of the disease, other symptoms may be noticeable. The child may be small for her age and have brittle bones. She may be jaundiced, bruise easily, or have cholesterol bumps (xanthoma) on her hands, elbows, or knees. A child with cardiac involvement may tire easily and catch colds more quickly in winter than other children. He may miss more school days than the average student. The child may need to be protected from teasing or watched on the playground. Falls and normal childhood mishaps may need to be dealt with more aggressively.

Treatment

AGS is treated through medications and nutrition in most cases, although surgery may be needed to correct structural abnormalities in the heart and kidneys, or to reduce the amount of bile circulating in the bloodstream. Treatment focuses on increasing the flow of bile from the liver with medications, preventing or correcting nutritional deficiencies such as malabsorption of





WHAT IS ALAGILLE SYNDROME (AGS)?

Alagille Syndrome (AGS) is a rare, multi-symptomatic genetic liver disorder that is characterized by a reduced number of small bile ducts within the liver combined with abnormalities in other organs including the heart, eyes, spine, kidneys, lungs and pancreas. The incidence of AGS is less than 1 in 70,000, but this number probably underestimates the frequency of the disorder since many patients remain undiagnosed, including some with a very mild condition who appear healthy. It is usually present at birth but may not be diagnosed until later in infancy, childhood or even adulthood depending on the severity of the symptoms. It is estimated that 50-70% of cases are found to be new mutations in the child and there is no evidence of the disorder in other family members. Family members may be affected very differently; one family member may have severe heart disease, another severe kidney disease, and a third, severe liver disease.

In the majority of cases, a scarcity of bile ducts inside the liver is the most obvious symptom of AGS. This results in insufficient or no passage of bile to the small intestine and malabsorption of fat soluble vitamins and nutrients. In addition to bile duct scarcity, the numerous symptoms of AGS include prolonged jaundice (yellow-skin coloration); abnormalities in the structure of the cardiovascular system, vertebrae in the spinal column, eyes, and kidneys; narrowing of the pulmonary arteries; characteristic facial features; shortened or "clubbed" fingers; and stunted growth.

The overall life expectancy for children with AGS is unknown, but depends on several factors; the severity of cholestasis and scarring in the liver, whether heart or lung problems develop because of narrowing in the pulmonary arteries, and the presence of infections or other problems related to poor nutrition. Patients with AGS generally have a much better outcome

IDEA and Section 504

There are two laws available to help parents advocate for children in the schools. The Individuals with Disabilities Education Act (IDEA) says that any public school must provide a free and appropriate education for students with disabilities. This law has been interpreted to provide for students with learning disabilities, but IDEA is also meant to protect children with Other Health Impairments (OHI), such as AGS. A child with AGS will qualify for protection under this law so long as his health issues may affect his ability to learn. To qualify under IDEA, you must prove necessity.

Because teachers, school nurses or administrators may not understand how IDEA encompasses OHI they may, unknowingly, deny a child accommodations. If this happens, the parents may seek protection under Section 504. Section 504 of the Rehabilitation Act of 1973 offers the same modifications as IDEA but states that any agency that receives federal funds must make accommodations for people with disabilities. The advantage of Section 504 over IDEA is that it covers children in environments other than the public school.

Whether IDEA or Section 504 is used, parents, teachers, the school nurse, and administrators meet to develop a plan that sets out all of the necessary modifications a child needs or may need in the future. Be creative in setting up modifications. The bottom line is the child can get anything he or she needs in order to learn.

The Alagille Syndrome Alliance would like to thank Digestive Care, Inc., makers of PANCRECARB® (pancrelipase), delayed-release, bicarbonate-buffered enzymes, for generously sponsoring the design, typesetting, production and printing of this booklet. Their support has made possible our ability to provide quality educational support materials to the community. *A very special thanks goes out to Dr. Karan Emerick of Children's Memorial Hospital, Chicago, IL and to Richard and Beth Caldwell, Directors of the Alagille Syndrome Alliance.*



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Dear Readers

Since Spring 2001, it has been a goal of the Alagille Syndrome Alliance to develop a booklet to aid AGS children in the school system. We started off small, but with the help of Digestive Care, Inc. the booklet has been expanded and given new life.

We are immensely pleased with the product and the cooperative partnership we have shared with DCI over the past months. We hope Alagille Syndrome in the Classroom will serve as a valuable tool to increase public awareness of AGS in schools, as well as outside their walls on the playground, in students' homes, and in the numerous school districts throughout the country.

Communication is the key to understanding. We hope Alagille Syndrome in the Classroom brings understanding to teachers, staff, administrators, and parents of how bright and special, but oftentimes medically fragile, our AGS children are, and motivates them to care for the AGS child with compassion and creativity.

*Cindy L. Hahn
President, Alagille Syndrome Alliance*

About the Alagille Syndrome Alliance

The Alagille Syndrome Alliance is an international non-profit support group for people with AGS. Our purpose is to serve as the main networking resource and source of information for these individuals, their families, friends, and health care providers.

Our goals include:

- To provide a support network for people with AGS and anyone who cares for or about them, and for families who have loved ones who have died as a result of AGS.
- To enable families with one or more members with AGS to meet other families in similar circumstances by providing a forum for members to meet on a regular basis to support one another, share current information about AGS, and participate in research studies.
- To gather and distribute information about AGS and other helpful services of interest to people with AGS, families, friends, and health care providers, including information about research studies available to them.
- To support research efforts on AGS by making information about studies available to families and encouraging their participation.
- To increase public awareness of AGS.
- To involve as many families as possible in Alliance programs and services.

decide the best course of action for a particular student only if they both have all the information. Parents should be prepared to discuss the way their child's physical condition may affect the classroom environment and the teacher must ensure the parents are aware of exactly how this is happening (i.e. excessive itching or bathroom use, physical behavior that may cause injury, disturbing the class).

Also, the teacher must address any issues of concern with the family. Because a child with AGS is more prone to bruising, lower than average height and weight, and apparent attention problems, he or she may be misdiagnosed by the school. An open dialog with the family and school team can prevent any confusion.

Hints for Teachers and Staff

- Talk with the child's family. They will have information on the child's needs and how to best meet them.
- Be prepared to allow for unconventional options, such as the child removing his shoes while at his desk or needing to squirm.
- Be aware that the child may have medications to take during the day that may not be conveniently timed.
- Be alert. A child with AGS may need medical attention for minor accidents. Let her be normal but thoroughly investigate any falls, especially those where the child hits her head or may have a fractured bone.
- Be creative. Since attention span could be limited due to itching or physical limitations, work with the parents to get the most out of the child's daily routine.
- Be observant. Even the healthiest looking AGS child may need down time.
- Be an advocate. Just like any other medical conditions, AGS may need to be explained to the child's classmates (with the parents' permission). Work with the family on the best way to do this.

