

AGS and Nutrition

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Outline

- Nutrition needs in AGS
- Macronutrients
- Caloric supplementation
- Fat soluble vitamin deficiencies
- Mineral deficiencies
- Questions

Introduction

- Malnutrition in children with AGS results from
 - Fat malabsorption
 - Liver disease
 - Pancreatic insufficiency
 - Inadequate intake
- This results in poor growth and nutritional deficiencies.

Calories

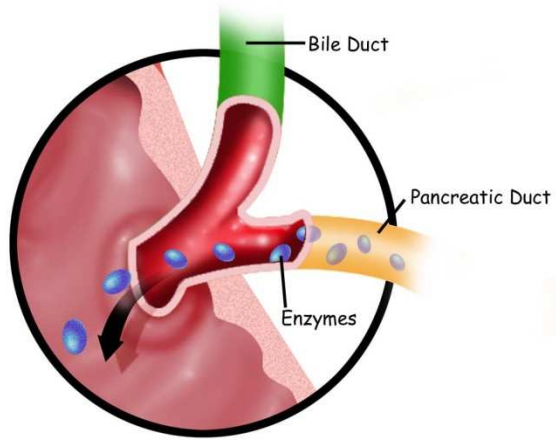
- Calorie requirements should be adequate to support growth and development and to reach or maintain desirable body weight and height.
- Factors that influence caloric requirements include body composition and level of activity.
- Children with AGS need to consume more than healthy children of the same age and sex.

Macronutrients

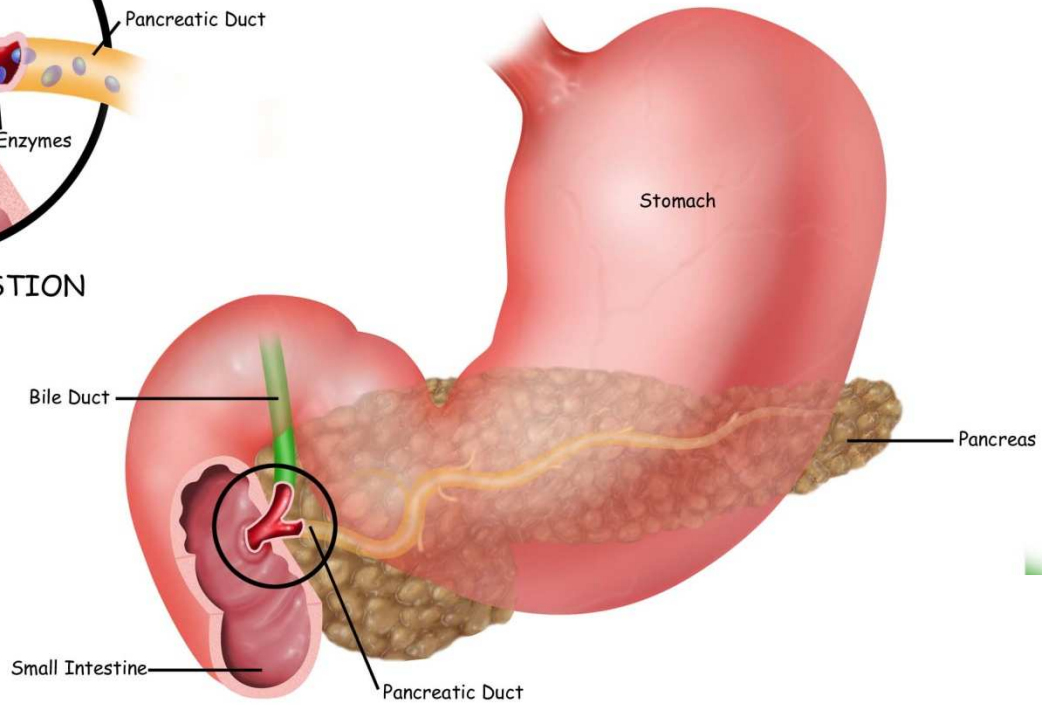
- Fat, protein, and carbohydrates.
- General recommendations: 50-60% of calories from carbohydrates, less than 30% from fat, and 20-30% from protein.
- Infants and younger children need a slightly higher proportion of fat (25-40%).
- Children with AGS need greater amounts of fat and similar amounts of carbohydrates and protein as compared to the general population.

Fat Intake

- Children with AGS need more fat than recommended for healthy counterparts.
- This is due to increased malabsorption of fat from lack of bile acids and in some patients, lack of pancreatic enzymes, BOTH of which are needed in the digestion of fat.



NORMAL DIGESTION



Fat Intake

- Children with AGS should not be put on a low-fat diet due to need for calories and essential fatty acids that are derived from fat.
- If a child has to be on a low-fat diet because he can't tolerate fat, it is necessary to ensure the child is consuming enough calories to grow in the form of carbohydrates and protein.
- A registered dietician (RD) can evaluate a child's diet and make suggestions on how many calories a child needs and ways to increase caloric intake.

Pancreatic Insufficiency

- Occurs in some children with AGS.
- Pancreatic enzymes are not properly released into the small intestine.
- These enzymes are necessary in the breakdown and absorption of mainly fat, but also carbohydrates and protein.

Pancreatic Insufficiency

- Methods to diagnose pancreatic insufficiency:
 - 72 hours fecal fat test = GOLD STANDARD
 - Fecal elastase
 - Pancreatic simulation test
- If pancreatic insufficiency is diagnosed, your doctor may add pancreatic enzymes to your medical regiment to better absorb nutrients, especially fat.

Essential Fatty Acids (EFA)

- EFA are fats that are necessary for maintaining the integrity of the skin and the structure of cell membranes and synthesizing important biologically active compounds.
- EFA are derived from nutritionally ingested fats.
- If fat is restricted too much in the diet than a child may develop essential fatty acid deficiency.

EFA Deficiency



FIGURE 85.—Acne vulgaris. A. Cystic acne of face. B. Subsiding tropical acne of trunk. C. Extensive acne of chest and shoulders.

Medium Chain Triglycerides (MCT)

- While reduced flow of bile into the intestine leads to poor digestion of most dietary fat, a specific type of fat - medium chain triglycerides or MCT – can still be well digested in children with AGS.
- Formulas that contain MCT oil or it can be used as a nutritional supplement to add calories and fat to food.



Special Formulas

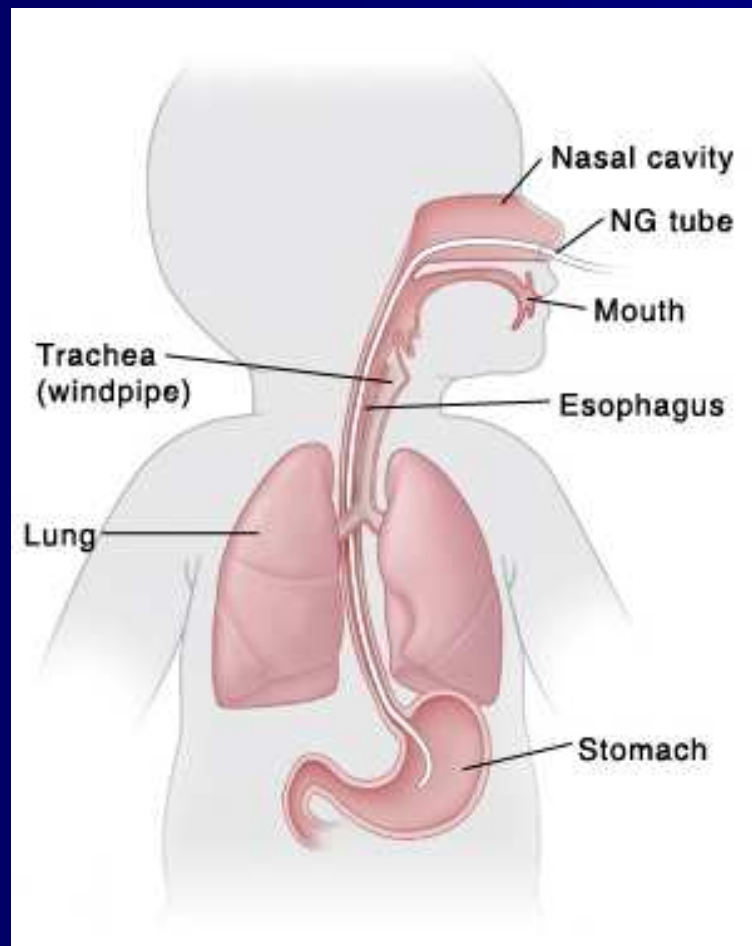
- Pregestimil
 - Infant formula with highest percentage of MCT oil
- Peptamen
 - Toddler formula with highest MCT oil
- Vital Junior
 - Structured lipid



Caloric supplementation

- A nasogastric tube (NG) is one that is passed through the nose down into the stomach. A nasogastric tube can be used to place nutrients directly into the stomach when a child cannot get sufficient calories by mouth.
- A gastrostomy tube (g-tube) is a tube that is placed into your child's stomach.
 - PEG (Percutaneous endoscopically placed gastrostomy)
 - Surgically placed gastrostomy
 - Combination
- If a child has an NG or g-tube he can still eat food but these are ways to supplement his nutrition.

NG tube



Types of buttons

- Balloon buttons
 - MicKey
 - Mini-One

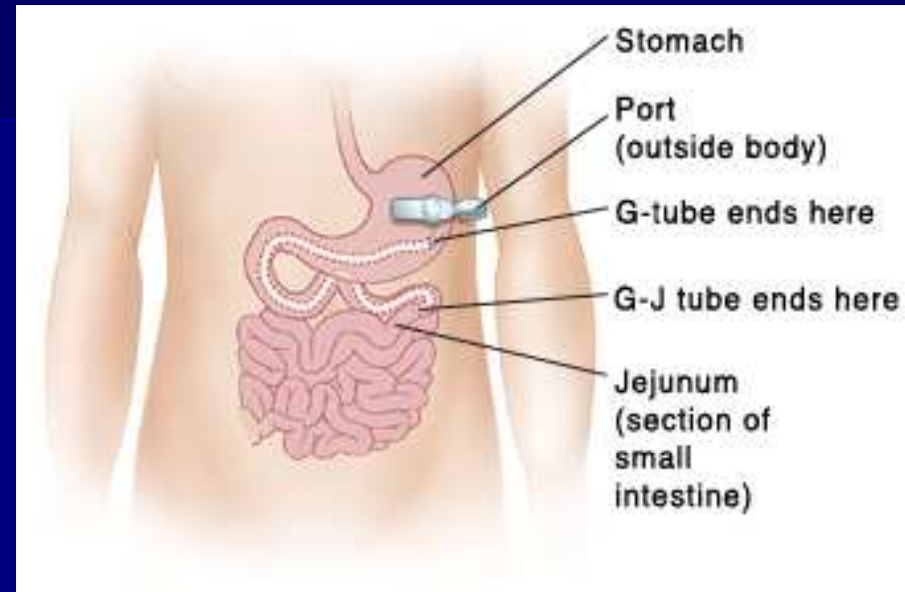


- Non-balloon buttons
 - BARD
 - Mini-One



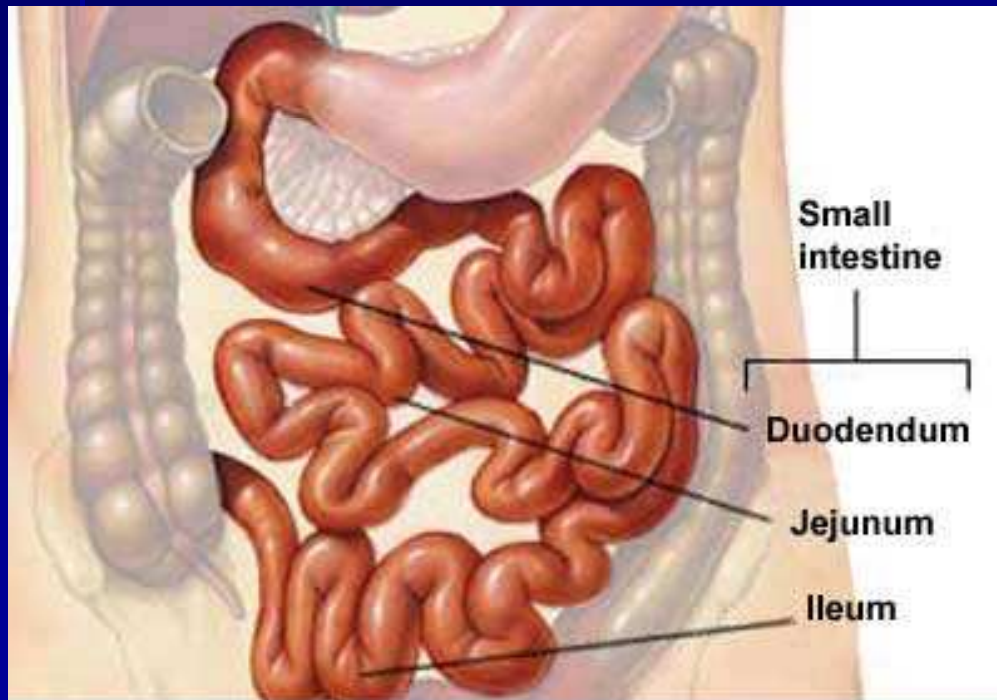
Gastrojejunal tubes

- Two ports
 - A gastric port: Usual used for medication delivery, venting, and drainage if stomach not moving normally
 - Jejunal port: Formula delivery
- Most common complication is dislodgement of jejunal portion into stomach requiring fluoroscopic or endoscopic replacement



Jejunal tubes

- Not commonly used



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High Calorie, High Fat Diet

- Individuals with AGS have increased energy needs and increased fat needs due to malabsorption
- Foods/Additives to add calories:
 - Whole milk, cheese, butter, margarine, or oil
 - Peanut butter or high calorie spreads for sandwiches and crackers
 - Ice cream or whole milk yogurt
 - Salad dressings, mayonnaise
 - Cream, half and half
 - Cream cheese, sour cream
 - Powdered milk (can add to casseroles, meatloaf, puddings, custards, etc)

Oral Supplements



- For Infants:
 - Standard infant formula and breastmilk has 20 calories per ounce. We can concentrate or fortify both formula and breastmilk up to 30 calories per ounce.
 - Polycose
- For Children and Adults:
 - Carnation Instant Breakfast
 - Pediasure, Boost Kid Essentials (both 1.0 and 1.5 versions)
 - Boost, Ensure (regular and Plus versions)
 - Resource Breeze, Glucerna
 - Duocal, Benecal, Polycose



Enteral Supplements

- For Infants:
 - Use infant formula (Pregestimil) and / or breastmilk fortified with MCT oil
- For Children / Adults:
 - Pediasure Enteral, Boost Kid Essentials 1.5, Peptamen Junior, Vital Junior, Elecare, Neocate Junior
 - Nutren 2.0, TwoCalHN, Peptamen 1.5

Appetite Stimulants

- Some patients who struggle with poor appetite despite trying high calorie diets and oral supplements may benefit from appetite stimulants such as Periactin, Megace, Remeron, Marinol, Oxandrolone.
- These are available by doctor's prescription only and are ordered on a case-by-case basis.
- Periactin is the most commonly used in children.
 - It is a anti-histamine with the side-effect of increasing appetite.
 - Tachyphylaxis (or getting used the medication and losing its effect) is common, so usually the medication is cycled on and off for best effect.

Fat soluble vitamins

- Problems with fat digestion and absorption may lead to deficiency of fat soluble vitamins since intestinal absorption of these vitamins requires adequate hepatic secretion of bile acids and pancreatic enzymes into the intestinal lumen.
- There are 4 fat soluble vitamins: A, D, E, K.

Fat soluble vitamins

- Supplementation of fat-soluble vitamins is essential for children with AGS and chronic cholestasis to optimize growth and nutritional status and in order to prevent deficiencies.
- Each fat-soluble vitamin should be supplemented separately so that the dose of each can be adjusted according to blood levels.

Vitamin A

- Important for:
 - Eye vision
 - Bone and tooth formation
 - Helps cells in the body function normally
 - Helps fight off infections and keep the body healthy
 - Works in the lining of the lungs to help fight infection
 - Keeps intestines stay healthy
- Not getting enough vitamin A can lead to night blindness. This means your eyes do not adjust when going from a lighted area to a dark area, such as when entering a dark movie theater.

Vitamin A

- Great Sources of Vitamin A:
 - Liver, egg yolk, whole milk, and fortified low-fat milks
 - Fortified cereals
 - Dark colored fruits and vegetables, including carrots, sweet potato, spinach, broccoli, apricots, cantaloupe, and peaches
- Vitamin A can be very toxic, so individuals with liver disease need to be careful about over supplementation.
- Careful monitoring of blood levels is essential due to the possibility of toxicity to the liver.
 - Retinol and retinol binding protein

Vitamin D

- Important for:
 - Building and maintaining strong bones and teeth
- Not getting enough Vitamin D can lead to thin, brittle bones that break easily → this can result in rickets in children.
- Great Sources of Vitamin D:
 - Sunshine
 - Fortified dairy products like milk, yogurt, cheese
 - Mackerel, canned sardines
 - Eggs

Vitamin D

- Vitamin D supplementation is often required in high doses in children with AGS.
 - D2 = ergocalciferol OR D3 = cholecalciferol (preferred form in cholestasis).
 - Co-supplementation with D- α -tocopherol polyethylene glycol succinate (TPGS) found in formulation of Vitamin E, enhances absorption of vitamin D.
- Blood levels should be checked regularly and doses adjusted to keep levels at the high end of the normal range.
 - 25-OH-Vitamin D
 - Levels should be maintained around 30 ng/dL.
- Cholestyramine (Questran) can bind to vitamin D so they should be given several hours apart.

X-ray of child with rickets



Vitamin E

- Important for:
 - Keeping red blood cells healthy
 - Keeping lining of the lungs and intestines healthy
 - Keeping muscles and nerves working properly
- Great sources of Vitamin E:
 - Vegetable oils, nuts, wheat germ, green leafy vegetables and fortified cereals
- Vitamin E comes in several forms but the best form of vitamin E for children with AGS is D- α -tocopheryl polyethylene glycol 1000 succinate (TPGS).

Vitamin E

- TPGS is better absorbed in children with cholestasis and enhances the absorption of vitamin D.
- Questran can also bind to vitamin E so they should be given at least 2 hours apart.
- Vitamin E is best measured as a ratio of serum vitamin E to total serum lipids, however, total serum lipid measurement is not easily available at all institutions.
- If vitamin status fails to normalize after several months of therapy, then injections of vitamin E will be needed.

Vitamin K

- Important for:
 - Helping blood to clot (Not getting enough Vitamin K can cause your blood to take longer to clot which can be dangerous)
 - Keeping bones healthy
- Great Sources of Vitamin K:
 - Dark green, leafy vegetables (especially spinach, broccoli, turnip greens and swiss chard)

Vitamin K

- A deficiency in Vitamin K can cause prolonged clotting time and bleeding.
- PT should be measure routinely.
- PIVKA is another measure of vitamin K status.
 - It is more accurate than PT, however, is more expensive and not readily available at all institutions.
- If oral vitamin K is not sufficient than injections can be given.

Other Common Nutrient Deficiencies

- The following nutrients can be malabsorbed in the presence of steatorrhea: calcium, magnesium, and zinc.

Calcium

- Calcium is extremely important mineral for growing children's bones.
- The heaviest bone mass that an individual achieves is called their peak bone mass.
- In order to achieve optimal peak bone mass children need to consume adequate calcium in their diets.

Calcium

- Calcium is found most widely in dairy food such as milk, ice cream, and yogurt.
- Certain foods such as orange juice and cereal bars are being fortified with calcium.
- Calcium deficiency can cause weak bones and increased risk of fractures.
- Children with AGS may need more calcium than healthy children due to their fat malabsorption.
- If albumin is low, may need to get an ionized calcium level to more accurately assess calcium status.

Magnesium

- Like calcium, magnesium is an integral part of the inorganic structure of bones and teeth.
- Green vegetables, cereals, legumes, and animal products are all good sources of magnesium.
- Magnesium depletion may lead to hypocalcemia and can contribute to the metabolic bone disease that complicates chronic cholestatic liver disease.

Zinc

- Zinc is important for proper growth and immune function.
- Zinc is not found in too many foods but good sources are lean red meats, seafood, peas, and beans.
- Zinc deficiency can cause a decreased sense of taste and smell, reduced ability to fight infections and poor growth.
- Zinc deficiency can be a result of chronic diarrhea and also can be the cause of chronic diarrhea.
- Serum zinc levels do not always reflect total body zinc status as it is an acute phase reactant and levels maybe decreased in acute illness.

Copper

- Copper has diverse functions including erythropoiesis and connective tissue synthesis.
- Copper has a wide distribution in the food supply. The richest sources of copper are oysters, other shellfish, and liver.
- A deficiency in copper in children with cholestasis is not usually a problem, rather toxicity is more of a concern since copper is excreted in bile.
- Since zinc and copper compete for absorption, zinc supplements can be given if copper levels are too high.

Thank you for your attention!!!

Questions???