Pediatric Interventions
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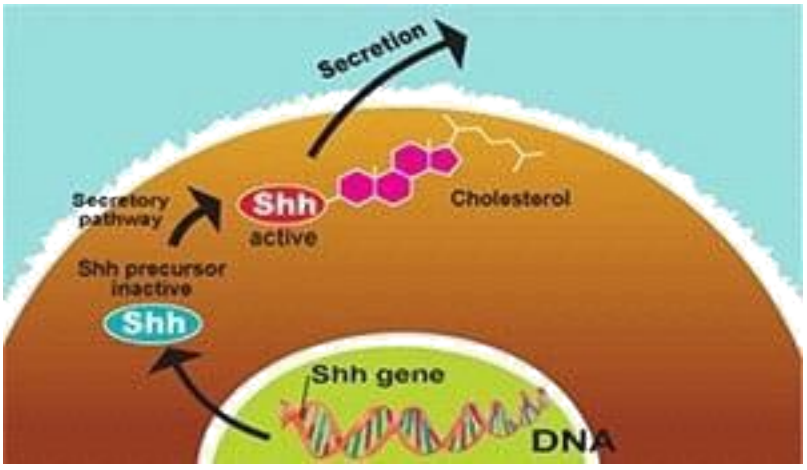
Cholesterol: The Good, Bad and The Ugly

The concept of good and bad for dietary substances depends on the circumstance of the individual person. Much of the information that the public receives is oversimplified. To a person dying of thirst in the desert, any water is very good. To a person who just drank two gallons of water on a dare, another glass of water might be fatal. The concept of good and bad cholesterol is similar to the water analogy.

The type of cholesterol that is associated with high density lipoproteins and helps to remove cholesterol from certain tissues was termed “good” cholesterol or HDL cholesterol (High Density Lipoprotein-associated cholesterol). The type of cholesterol associated with low density lipoproteins and which transports cholesterol to tissues that require it was designated as “bad” cholesterol or LDL cholesterol (Low Density Lipoprotein-associated cholesterol). ***If, however, the tissues of a certain person have a significant overall deficiency of needed cholesterol, then both LDL and HDL cholesterol are good for that person.***

Therefore, a purified cholesterol supplement cannot be inherently “good” or “bad” and the body will distribute it to the locations where it is needed the most. If the person has adequate amounts of cholesterol, however, no additional supplementation would be needed.



Abnormalities in cholesterol metabolism present in SLOS (Smith-Lemli-Opitz syndrome disorder) and autism also impair the function of a developmental signaling protein with the bizarre name "Sonic Hedgehog." Sonic hedgehog (SHH) is named after the character from the popular Sega Genesis video game. The original hedgehog gene was found in the fruit
fly Drosophila and was named for the appearance of the mutant fly offspring in which the embryos are covered with pointy spines resembling a hedgehog. The first two types of hedgehog proteins were named after certain species of hedgehogs and the third was named after the video game character.
Cholesterol must covalently bond to SHH before SHH can function properly. In addition, some forms of SHH have both cholesterol and the fatty acid palmitic acid covalently attached to the protein. (Palmitic acid is required for the production of a soluble Hedgehog protein complex and long-range signaling in humans). The attachment of cholesterol activates the sonic hedgehog protein, and without adequate cholesterol, SHH protein function is impaired.

Everyday supplementation with high cholesterol foods, such as egg yolks, might prove to be a useful therapy to try for a few months for children with ASD who have cholesterol values that are low (<160 mg/dL). Unfortunately, egg allergy is common in autism and may increase with a steady egg diet, and compliance may be difficult for children who dislike eggs. Although very high blood serum cholesterol values are associated with heart disease, ***values that are low (below 160 mg/dl) are associated with increased violent behavior, suicide, depression, anxiety, bipolar disorder, Parkinson's disease, and increased mortality from cancer.***

Surprisingly, high cholesterol protects against some infectious diseases like tuberculosis, which has been uncommon in the USA since The Great Depression, during which there was a substantial lack of high cholesterol foods because of financial hardship. Vegetarians have a much higher incidence of tuberculosis than meat eaters.

It is possible that the overemphasis on a low cholesterol diet may also be associated with the recent marked increase in cases of tuberculosis. Low cholesterol values are also associated with manganese deficiency, celiac disease, hyperthyroidism, liver disease, malabsorption, and malnutrition. Pregnant women with low cholesterol are twice as likely to have premature babies or babies with small heads.

LDL cholesterol (so-called bad cholesterol) protects humans against infection. Deadly Staphylococcus bacteria produce endotoxins that have the ability to kill human cells, including red blood cells. LDL was found to protect human red blood cells from this toxic effect of endotoxins while HDL was not protective. A

study at the University of Pittsburgh found that in young and middle aged men, those that had LDL-cholesterol below 160 mg/dl had a significantly lower number (of total and various types) of white blood cells than men with LDL-cholesterol above 160 mg/l.

Functions of Sonic Hedgehog (SHH):

- Plays a central role in developmental patterning, especially of the nervous system and system.
- Important in the growth and differentiation of a variety of cell types, including the development of T-cells in the thymus.
- Purkinje neurons secrete SHH to sustain the division of granule neuron precursors in granule layer in cerebral development. Abnormal cerebellar development.
- As a transcription regulating protein, SHH alters which genes function at a given time.

Testing for cholesterol, cholesterol transport proteins, and homocysteine can be beneficial. The Great Plains Laboratory has developed a special cholesterol- related panel that will help to determine whether cholesterol deficiency or abnormalities in cholesterol transport are present. This panel will include the following markers: Total cholesterol, apolipoprotein A-1, apolipoprotein B, Lipoprotein (a), and homocysteine. Lipoproteins are involved in cholesterol, lipid, and vitamin E transport.

Total cholesterol: Total cholesterol measures all types of cholesterol including esterified and free.
Low values (generally values less than 160 mg/dL) are associated with genetic diseases of cholesterol. Low values are more common in hyperthyroidism, liver disease, malabsorption, malnutrition, autism, violent behavior, celiac disease, anxiety, bipolar disease, alcoholism, lung cancer, suicide, depression, and obesity associated with human adenovirus-36 infection.

In China, where mean cholesterol is much lower than in the Western world, chronic hepatitis-B virus infection is ubiquitous. Chronic carriers of hepatitis-B, but not individuals with eradicated hepatitis-B, have significantly lower total cholesterol than non-carriers, suggesting a cause-effect relationship. High cholesterol values are associated with atherosclerosis.

Apolipoprotein A-I (Apo A-1): The main protein component of HDL (high density lipoprotein). It accounts for approximately 65% of the total protein content of HDL. Apo A-I activates lecithin cholesterol especially in children.

Apolipoprotein B (Apo B): The main protein component of LDL (low density lipoprotein). It accounts for approximately 95% of the total protein content of LDL. Apolipoprotein B is necessary for the reaction with LDL receptors in the liver and on cell walls and is thus involved in transporting cholesterol from the liver to the cells. Recently the Mind Institute found that low values of Apo B are associated with autism, with the lowest values being found in low-functioning autism. LDL has been found to have protective effects against endotoxins from deadly staphylococcus.

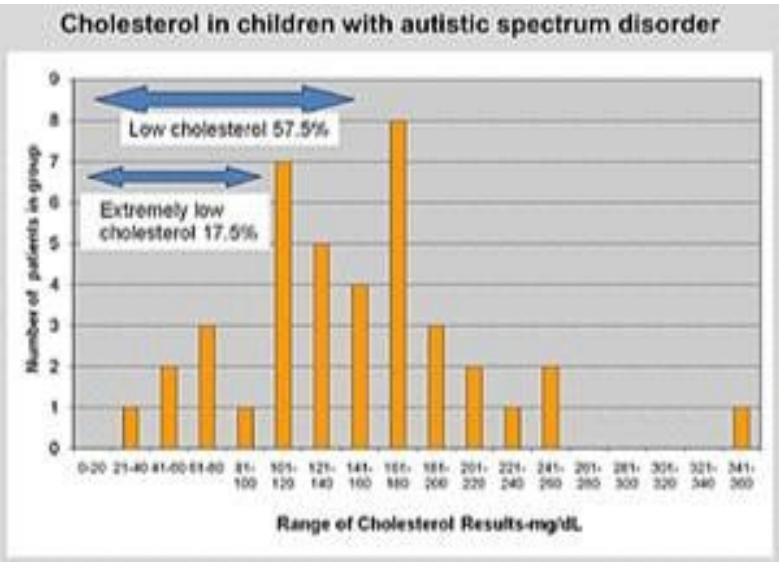
Lipoprotein (a): Consists of two components, the low-density lipoprotein (LDL) and a glycoprotein, which are linked by a disulfide bridge. High values have been implicated as a risk factor for cardiovascular disease, Alzheimer's disease, Crohn's disease, and rheumatoid arthritis.

Low values have also been found in those with autism who have higher doses of Apolipoprotein E epsilon-4 gene variants that are associated with increased risk of Alzheimer's disease. Lipoprotein (a) is unrelated to Apolipoprotein A.

Homocysteine: A sulfur-containing amino acid that can be converted to methionine by methionine synthetase or by betaine methyl transferase. The role of homocysteine in atherosclerosis gained attention after finding massive atherosclerosis in young people with the genetic disorder homocystinuria. Methionine synthetase requires the folic acid derivative 5-methyl tetrahydrofolate.

Why the brain needs cholesterol

- There is a direct correlation between the concentration of cholesterol in the brain, particularly in the myelin, and how well the brain functions.
- The brain is the most cholesterol-rich organ in the body.
- In the central nervous system (CNS), essentially all (99.5%) cholesterol is the free or unesterified form (unattached to fatty acids).
- The majority (70%) of cholesterol present in the CNS is believed to reside in the myelin (the material that insulates the nerve fibers) sheaths and the plasma membranes of astrocytes (brain support cells) and neurons.
- Half of the white matter, which contains the nerve axons that allow for transmission of brain signals, may be composed of cholesterol-rich myelin.



Benefits of cholesterol feeding in SLOS
Kelley RT. Inborn errors of cholesterol biosynthesis. Adv Pediatric 2000; 47:1-53

- Beginning to walk
- Starting to run
- Growth improvement
- Less infections
- Less UV light sensitivity
- Increased alertness
- Head-banging stops
- Decreased tactile defensiveness
- Increased sociability
- Behavior improves
- Talking has started in adults who
- were not talking before
- Verbal people say they feel better
- Many improvements in only a few
- days after supplement
- Decreased irritability
- Increased muscle tone

Adapted from http://biomedicalnutritionalintervention.com/training/