Nutrition and the Patient with Progressive Central Nervous System Disease

A manual for patients and their families

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ERRATA

The Recommended Dietary Allowances (RDA) have been changed for a few vitamins and minerals after this publication was in print. Please note the following changes from the RDA of 1989.

	Males		Females	
	1980	1989	1980	1989
Vitamin A (RE)	1000	1000	5000	800
Thiamin (mg)	1.4	1.5	1.0	1.1
Riboflavin (mg)	1.6	1.7	1.2	1.3
Niacin (mg)	18	19	13	15
Vitamin B6 (mg)	2.2	2.0	2.0	1.6
Folacin (mcg)	400	200	400	180
Magnesium (mg)	350	350	300	280
Iron (mg)	10	10	18	15
Zinc (mg)	15	15	15	12
*Calcium (mg)	800	800	800	800

*Calcium has been increased to 1200 mg for females between ages of 14 to 24 years of age.

This book is dedicated to
our patients and their families
with whom we have worked
for so many years.

About the Authors

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We plan in future editions of this book to include more information on other neurologic disorders and on the growing body of research on nutrition and the brain.

Comment from Dr. Perlman (2009)—the careful reader will see that there have been changes over the past several years, with respect to the food pyramid, recommended daily servings from each of the food groups, and recommended daily allowances of vitamins and minerals. These will be reflected in the next edition of this book. The need to be an educated consumer has not changed.

Acknowledgments

The authors of this book would like to thank the many people whose help and encouragement have brought our efforts to fruition:

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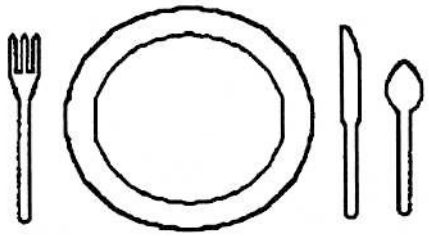
1. Introduction

It is important that one ensures an adequate intake of macronutrients (carbohydrates, fats and proteins) as well as micronutrients (*vitamins* and *minerals*). This can often be difficult to achieve when associated problems include trouble in chewing and in swallowing, lack of transportation for food shopping, often limited budgets, and misinformation about nutritional requirements.

In spite of this, it is possible to obtain well balanced meals affording an adequate nutritional status, when simple dietary guidelines are followed.

It is the purpose of this publication to present a broad overview of nutrition and its importance in the overall care of the human body, with a focus on patients with neurologic disorders of movements (e.g. incoordination and tremor in Friedreich's *Ataxia* (FA), Olivopontocerebellar Atrophy (OPCA) and related diseases (Spinocerebellar Ataxia, Multiple System Atrophy); extraneous movements in Huntington's Disease (HD) and Tardive Dyskinisia (TD).

This book is designed to serve as a guide to good nutrition in an age where food myths and faddism have penetrated the market.



2. The Four Food Groups

Some fifty *nutrients*, including water, are required daily by the body for optimum health. Nature has facilitated this for man by assuring that by obtaining the ten "leader" nutrients (Protein, Fat, Carbohydrates, Vitamin A, Vitamin C, *Thiamin*, *Riboflavin, Niacin,* Calcium and Iron) through normal food intake, the other forty simply fall into place (as they are obtainable from these same foods, when they're taken in the recommended amounts).

It is almost impossible for one particular food to provide all these essential macro and micronutrients, so the key to good nutrition is variety.

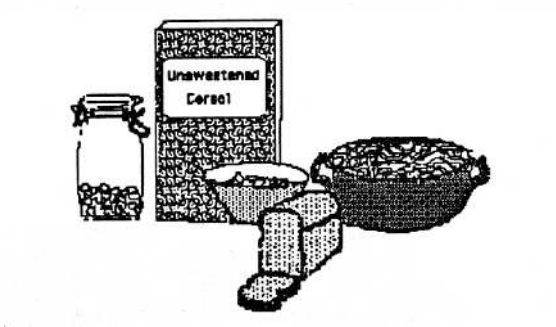
It should be noted however that sometimes the food industry adds one or more vitamins or minerals to their food item. When this happens, or when a nutrition claim is made, the nutrition labeling regulation requires the manufacturer to list these ten "leader" nutrients and their amounts per serving on the package, for example on cereal boxes.

The four food group concept was created to facilitate planning well balanced meals through variety. By dividing all nutritious foods into four distinct categories, the individual is less likely to miss out on important nutrients that may be found in one particular food item and not the other.

These four food groups are:

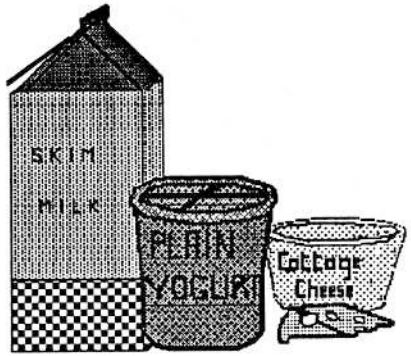
I. The grain group

It is in this group that you find nearly all the starchy food items of the average diet including rice, pasta, all the cereals and all the breads and bread products. This group is the major source of carbohydrates and provides a significant amount of thiamin, niacin and iron (usually added to the particular bread or cereal through a process called *enrichment* and/or *fortification*).



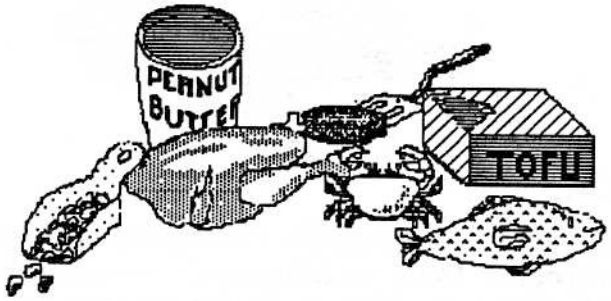
II. The milk group

As the name suggests, this includes dairy products such as milk, cheese, cottage cheese and yogurt. Dairy puddings and ice cream are also found in this group. Protein, riboflavin and calcium are the major nutrients found here. It should be noted that a portion of ice cream may have but half the nutrition of an equivalent portion of milk or cheese due to the sugar and cream that are among its main ingredients.



III. The meat group

Red meat such as beef, lamb, pork and veal as well as white meat such as chicken and fish, are the obvious members of this group. Eggs, peanut butter, nuts, and dried legumes are also part of this group. Protein, thiamin, niacin, and iron are the primary nutrients. Again, consider the caloric density relative to the amount of nutrition. The best example is that of peanut butter, where 4 Tablespoons (400 *calories*) are equivalent to 2 oz (100 calories) of lean meat or chicken or fish with respect to protein content (the caloric difference being related to the amount of fat and sometimes carbohydrates if sugar is added).

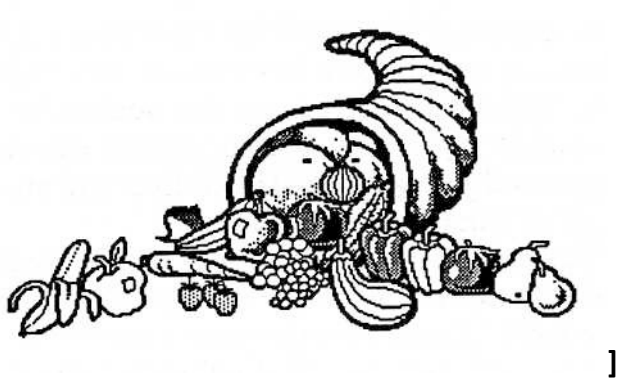


IV. The fruit and vegetable group

This group includes everything from potatoes to citrus fruits to spinach, squash, corn etc., all of which offer a wide variety of vitamins and minerals and above all have a high content of fiber. Carbohydrates in their complex form are the main macronutrient in this group. The thing to remember is that dark green, leafy or orange vegetables are recommended 3 or 4 times per week for Beta *Carotene* which is the precursor for Vitamin A, and citrus fruit is recommended daily for Vitamin C.

In general fruits have a higher content of the simple sugar fructose which gives them a sweeter taste than vegetables.

Vegetables, on the other hand, can further be divided into green vegetables (such as broccoli, green beans, lettuce, celery) and starchy vegetables (such as corn, potatoes, peas, squash or yams). These starch vegetables are at least twice as dense in caloric content as the green ones.



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Others

Oils, margarine, butter, creams and dressing are items that complement these groups but do not replace food from any of them. They should be used wisely, depending on the individual's caloric needs.

1. <u>Macronutrients</u>

The human body, most basically, consists of elements, the 3 major ones being hydrogen, oxygen and carbon. These 3 elements are combined in several million ways to form the biochemical compounds that make up the different parts of the body, primarily water, fat, and carbohydrates. When nitrogen is added to the configuration, protein results. The position of these building blocks in relation to one another, along with the addition of other elements determines the final structure of the human tissues, (for example, the collection of proteins from an individuals' skeletal muscle is different from that in the same individuals' heart).

The major constituent of the human body is water. Fat, mostly stored in the *adipose* tissues, but also found in the nerve tissues and in the brain, is the next major component. This is followed by proteins, such as in the skeletal muscles, *somatic* proteins and blood proteins. Carbohydrates, which are primarily in the form of blood sugars and *glycogen* stored in the liver and muscles, contributes the smallest fraction of all 3 in terms of weight.

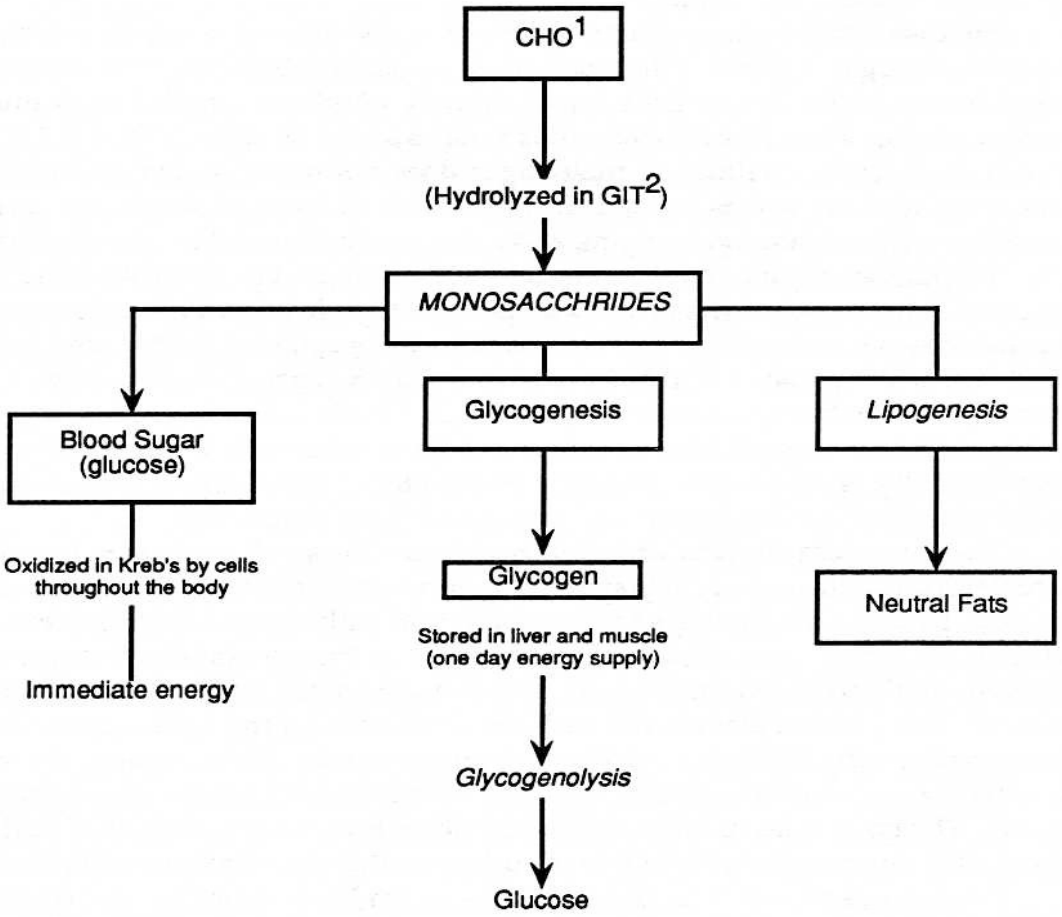
The 3 major macronutrients (fat, protein and carbohydrates) not only form the basic chemical structure of the human body, but they are also the compounds a person ingests as food. What makes the protein coming from chicken, different from the protein coming from fish or beef or that found in the human body is its configuration, i.e. the basic elements and their position in relation to one another, along with the presence or absence of other elements.

It should be kept in mind and referred to later on, that the energy yield of fat is more than twice greater than that of either carbohydrates or protein. While 1 gm of either of the latter yields four calories upon digestion and *metabolism*, 1 gm of fat yields the nine calories.

I. Carbohydrates

These are ingested in the form of sugars and starches (simple sugars specially linked together) coming mostly from the legumes, bread and cereal groups. Carbohydrate food items may also contain di-(two) or oligo-(3-10) sacchrides (simple sugars less rigidly linked together). Common *dissacchrides* are *sucrose, lactose* and *maltose*.

Metabolism of Carbohydrates



[1]CHO: Carbohydrated
[2]GIT: Gastro Intestinal Tract

Sucrose, (table sugar) is found in cane or beet sugar, molasses, brown sugar and maple sugar. Lactose is the only carbohydrate of animal origin that's of any significance in the diet and it's found in milk. A glass of milk has as much carbohydrates as a piece of fruit, but it's not as sweet because lactose is 1/6 as sweet as sucrose. Maltose or malt sugar does not occur in any appreciable amounts in foods and is formed during the breakdown of starch, eg. in the malting and fermentation of grains, and is present in beer and breakfast cereals.

Through *enzymatic* activities starting in the mouth, but occurring mostly in the small intestines, starches, *oligosacchrides* and their intermediate products are broken down into simpler sugars (*monosacchrides*, eg. *glucose, fructose* and *galactose*). Galactose results from the breakdown of milk sugar, but some foods may already contain free glucose and fructose. Glucose is found in grapes, berries, oranges and some vegetables, such as sweet corn and carrots. It is also prepared commercially as corn syrup. Fructose, on the other hand, is much sweeter than glucose and is found in honey, ripe fruits, and some vegetables.

These monosacchrides are absorbed from the small intestines into the circulatory system and taken first to the liver by the portal vein. The liver is the principal organ that regulates the metabolism of carbohydrates, storing excess glucose as glycogen and releasing glucose again for transport and delivery to the millions of different cells in the body, where it is oxidized for immediate energy release. This glucose that's taken from the circulation by the body Is constantly being replaced by the liver, so that the blood glucose level is maintained within relatively narrow limits. Muscle cells also store glycogen, for their own energy needs. The body relies on these glycogen stores for energy production during short term deprivation of food intake such as in the case of an overnight fast.

Following a meal rich in carbohydrates, as the immediate energy demands of the body are met and the glycogen stores filled, the excess flow of these circulating sugars is stored as fat in the adipose tissues, also under the control of the liver, for later use when the body needs it.

Insulin, a *hormone* secrete by the pancreas, plays a major role in the use and storage of carbohydrates. Among its functions:

1. It stimulates the uptake of glucose for oxidation by many tissues.

2. It speeds up the formation of glycogen from circulating blood glucose.

3. It halts the breakdown of glycogen back into glucose for energy needs.

4. It promotes the conversion of digested carbohydrates into fat.

Of possible concern to older patients or patients with low blood pressure is that increases in insulin stimulated by a high carbohydrate diet may weaken the internal blood pressure maintenance circuits.

Long term deprivation of carbohydrates in the diet uses up glycogen and causes stored fats to be "burned" for energy. Their breakdown products include *ketone* bodies which may build up and cause *ketosis*. *Ketogenic* diets, which restrict carbohydrates and protein but include supplements of fats and B-vitamins, have been designed for very specific disorders (childhood epilepsy and pyruvate dehydrogenase complex deficiency) where the body may not use sugars efficiently for energy but could use fats. One should not follow such a diet without medical supervision as it could be very dangerous. As little as 50 gm of carbohydrates per day should prevent development of ketosis.

II. Fats

This varied group has the *fatty* acid as its basic building block. Fatty acids with as few as four carbons and with as many as twenty six carbons in their backbone have been identified, differing also by how *saturated* with hydrogen they are. Saturated fats are solid at room temperature, eg. butter, margarine or lard. Some oils, though liquid at room temperature, have a large amount of saturated fatty acids, eg. coconut oil. Solid shortening and margarine, though actually of vegetable origin, have had hydrogen incorporated to make them behave like butter or lard.

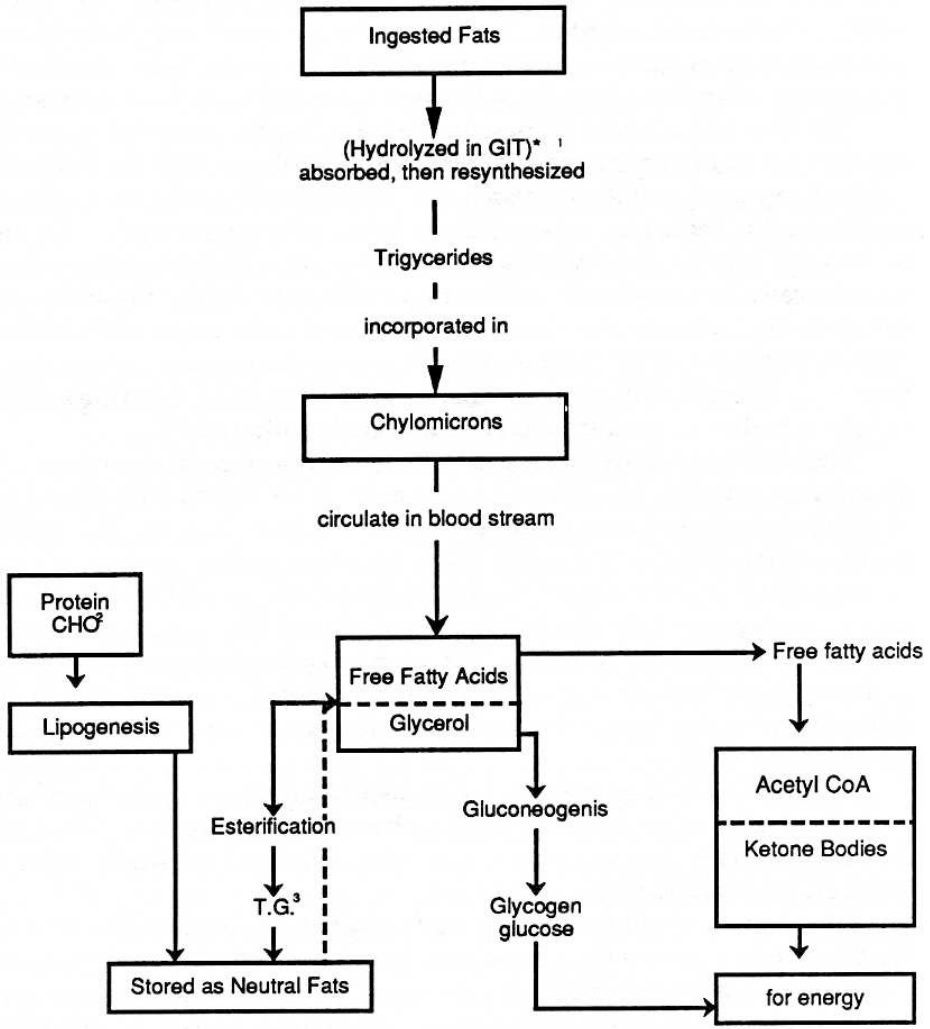
The most basic and important function of fats, besides providing *satiety,* caloric density (nine calories per gram of fat as compared to four calories per gram of protein or carbohydrates), and palatability, is their provision of the essential (cannot be made by the body) fatty acids *linoleic acid* and *linolenic acid*, which have been found to be altered in some neurologic disorders. *Arachidonic acid* can be synthesized in the body when linoleic acid is supplied in the diet.

The most obvious source of fat in the human diet is, of course, oils, margarine, butter, cream, and lard. These are often referred to as the visible fats. Most people have become accustomed to indiscriminately add such items to their meals, giving an unnecessary caloric load without contributing significantly to the meals' nutritional value. The invisible fats on the other hand are found in most dairy products (that do not carry the descriptive label "nonfat"), in the marbling of meats, and in most pastries and peanut butter. Nuts are also very high in fats as is the avocado. Safflower, corn, cottonseed and soybean oils are very rich in linoleic acid whereas peanut and olive oils are rich in oleic acid (which can be manufactured in the body) and correspondingly lower in linoleic acid.

Almost all the fats presented to the digestive tract for breakdown occur as *triglycerides*, where three fatty acids are attached to a *glycerol* molecule (of carbohydrate origin). Their digestion takes place primarily in the upper small intestines, by the action of highly specialized groups of *enzymes* called the pancreatic lipases. As the food passes from the stomach into the small intestines, it stimulates cholecystokinin, a hormone that causes contractions of the gallbladder and emptying of bile into the small intestines where it aids in the complete digestion of fats. Since one fatty acid is removed at a time, the breakdown of triglycerides yields di(2)glycerides and then mono(1)glycerides. Only about ¼ to ½ of the triglycerides are completely *hydrolyzed* to glycerol and fatty acids. *Cholesterol* esters are hydrolyzed by the enzyme cholesterol esterase to form cholesterol and fatty acids.

Therefore the end products of *lipid* digestion that are presented for absorption include fatty acids, glycerol, monoglycerides, some diglycerides and cholesterol. At the end of the small intestines, these are complexed with bile salts to form water-soluble compounds called *micelles*. At the point of contact of the micelle with the cells lining the small intestines or *villis*, the lipids are released and cross the cell border by mechanisms not yet fully understood. Fatty acids that contain twelve carbon atoms or less are absorbed into the portal circulation without further modifications. They are attached to the blood protein *albumin*, to make them water soluble for their transportation, and may be used within the liver or the portal circulation. On the other hand, fatty acids that contain fourteen carbon atoms or more are *resynthesized* with glycerol into new triglycerides within the epithelial cells of the small intestines and then taken into the circulation via the lymph ducts. The blood is therefore the means of transportation of these lipids from one side to another. Cholesterol and triglycerides, however, cannot exist in the free state in the circulation because fats are insoluble in water. Proteins, then, provide the mechanism for their transport in the aqueous medium of the blood, enveloping the cholesterol and trigylcerides and forming *chylomicrons*. These are high in the blood after a fatty meal and are cleared from the plasma about twelve hours later. The very low density lipoproteins (VLDL) and the low density lipoproteins (LDL) transport fats in the blood from the liver and the intestines to the body cells. Their increased levels in the blood is associated with a high risk of a heart attack. While the high density lipoproteins (HDL) are also fat transport mechanisms, their increased levels in the blood has a protective effect against heart attacks. This is due to the belief that the HDLs transport the fats from the cells to the liver for recycling and/or disposal.

Metabolism of Fats



[1] GIT: Gastro Intestinal Tract
[2] CHO: Carbohydrates
[3] T.G.: Triglycerides

The liver and adipose tissues are the specialized organs that control lipid metabolism, as discussed under "Carbohydrates" above. The synthesis of new lipids (*lipogenesis*) and the breakdown of old lipids (*lipolysis*) are taking place continuously. These reactions are helped by specific enzymes under the control of nervous system or hormonal mechanisms. It is important to stress that muscles can effectively use fatty acids to generate their energy requirements. As a matter of fact, all cells of the body, except those of the brain, spinal cord and the red blood cells can use fatty acids to obtain energy. In the young nervous system however, the brain cells can, after a period of starvation, adapt to the utilization of ketone bodies derived from fat and ketogenic amino acids.

The balance of fats has in the past decade, become important, as studies have shown that a diet high in saturated fatty acids is associated with higher blood cholesterol levels an with the onset of coronary heart disease. This saturated portion of the diet (i.e. fats in red meat, and other animal sources, as well as *hydrogenated* vegetable fats and coconut and palm oils) should not exceed 10% of the caloric density of the diet, while the total fat calories should remain under 30% (Robinson et al. pp. 529-337). This seemingly simple dietary manipulation is what presumably can help lower blood cholesterol. Nevertheless it is rather difficult to plan that type of diet without ending up with a diet naturally low in cholesterol as well.

A number of other measures will also bring about lowering of serum cholesterol, primarily weight loss by the obese, increased activity, and reduced intake of total fats. But, authorities agree that if dietary modifications are to be effective, they should begin early in life.

Recent studies indicate that a high intake of fish (especially fatty fish) lowers the risk of heart disease (Anderson and Sprecher, 1987). This comes from the observation that the inhabitants of Greenland consume large amounts of fatty fish and yet have one of the lowest incidents of heart disease, compared with other populations. No one is certain what exactly in fish lowers the blood cholesterol but it has been postulated that it is the fatty acid linoleic or omega 3 fatty acids. There has however been no data to suggest that fish oil capsules are of any benefit. On the contrary, their intake can result in a decrease of platelet counts which results in a lower clotting ability in the blood. (Comment from Dr. Perlman (2009)—recent evidence of elevated levels of mercury in certain types of fish warrants restriction of intake, especially in children and pregnant women, eg. Shark, Swordfish, King Mackerel, Tilefish, Albacore Tuna).

One cannot talk about fats and coronary heart disease without discussing cancer, as there has been a strong association between the incidence of colon cancer and breast cancer and the per capita consumption of total fat, saturated fat, and cholesterol (Robinson et al., pp. 496-497).

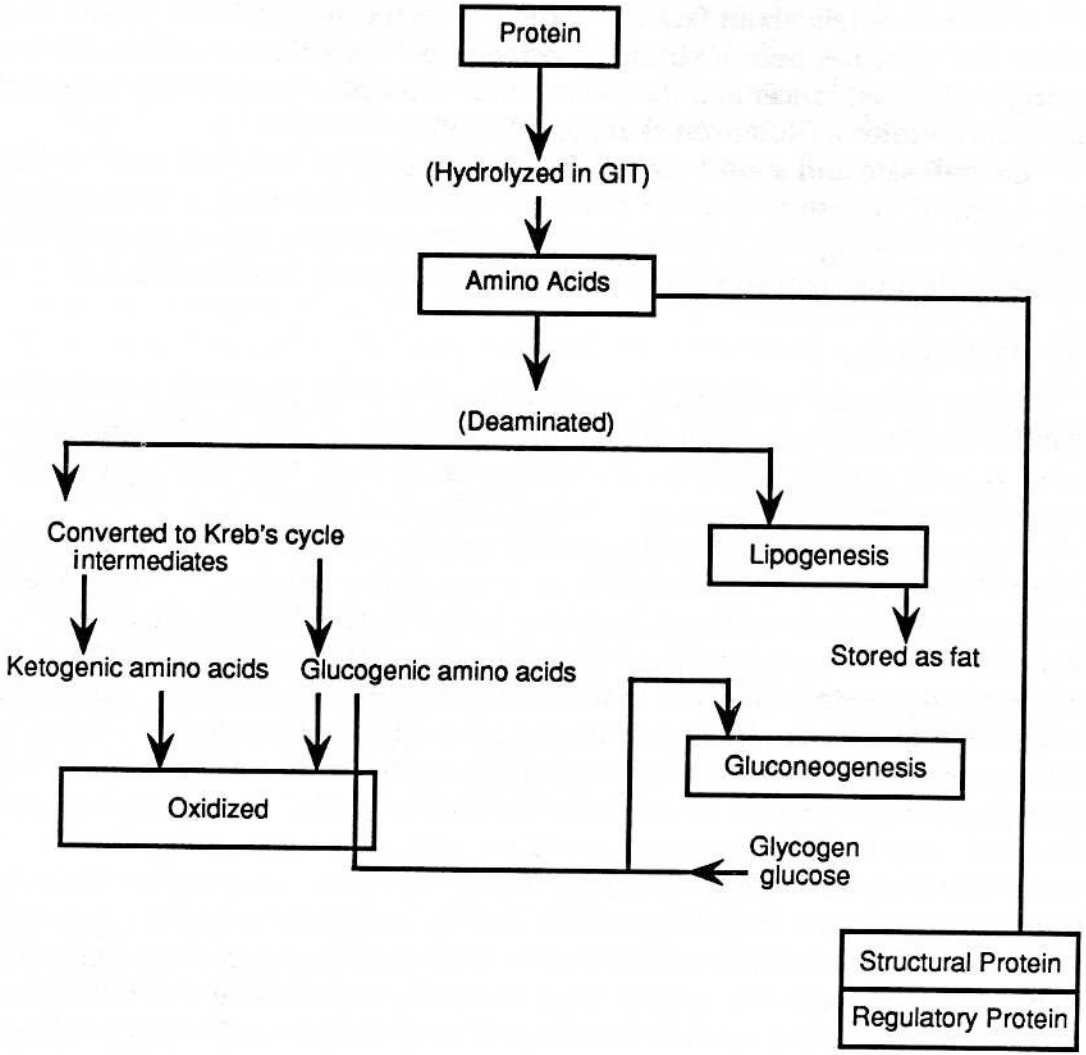
Overall safe and sound dietary practices should include selection of lean cuts of meat, the use of fish and poultry more often than the use of red meat, cooking by broiling, baking, or roasting, use of lowfat or nonfat dairy products and the substitution of soft margarines for regular margarines and butter.

III. Protein

Proteins constitute the chief solid matter of muscles, organs, and endocrine glands. They're also major constituents of teeth, bones, nails, hair, skin and blood cells. It is no wonder that they are often referred to as "the building blocks". Furthermore, they have innumerable regulatory functions.

Enzymes, hormones, blood proteins, nucleoproteins, contractile proteins are all highly specialized "reusable tools" that are made of proteins. Proteins are also potential sources of energy. If the diet does not provide enough calories, the body uses the dietary (*exogenous*) protein, along with the breaking down of its own (*endogenous*) protein (initially from worn out cells in the mucous membranes and excess digestive enzymes) to meet its energy needs. Protein digestion starts in the stomach and continues in the small intestines (*duodenum and jejunum*). The final products of protein metabolism are amino acids. These, as they are absorbed from the intestinal tract, are taken up by the veins going to the liver (portal circulation). The liver redirects the amino acids to each and every cell in the body, which absorb the amino acids needed to synthesize the numerous proteins that they need for their functions. Likewise amino acids and products of amino acids are constantly being added back into the circulation. This milieu of amino acid has been appropriately named "the amino acid pool". It must be stressed that the cell cannot distinguish between endogenous or exogenous amino acids. In a typical American diet the total protein requiring digestion could be as much as 160 gm: 90 gm from food and up to 70 gm from the endogenous sources. The "amino acid pool" that is available to any given tissue at any given time thus includes the exogenous sources (dietary) as well as endogenous ones (tissue breakdown).

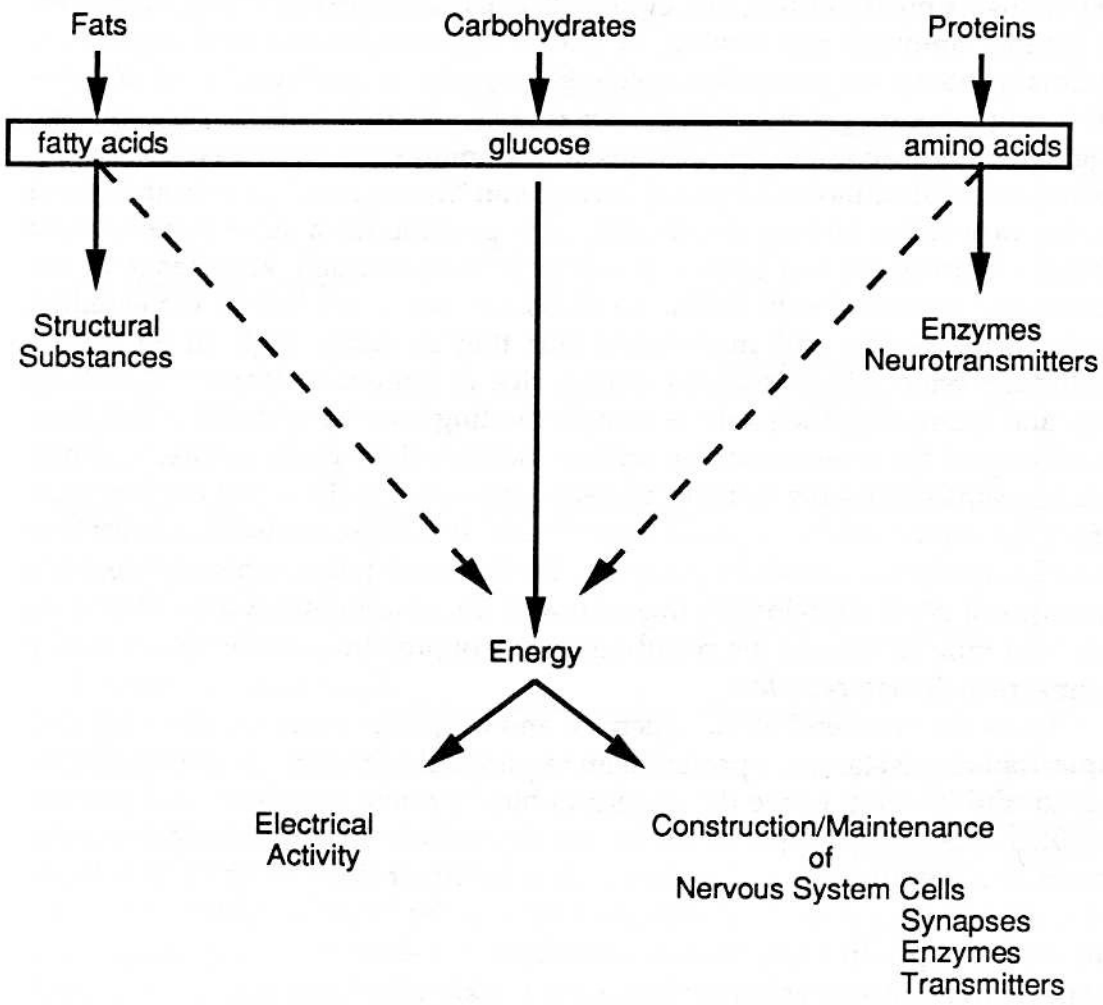
Metabolism of Protein



Twenty amino acids have been identified: nine are essential (i.e. must be obtained form the diet as the body cannot make them) and eleven are non-essential (the body can make them). What determines the "quality" of a protein is the availability of the *essential amino acids*. Although the major dietary sources of proteins are meat, poultry, fish, eggs, milk and cheese, protein is also found, but in smaller amounts per serving, in grains, legumes, cereals and vegetables. Animal proteins are generally considered superior in quality to those of vegetable origin because of this distribution of essential amino acids. For instance, eggs and milk are complete in all the essential amino acids. Whereas rice, lacking in one essential amino acid (lysine), becomes an "incomplete" protein and lysine in this case is the *limiting amino acid*. It is possible for a strict vegetarian to obtain adequate protein intake, but only with a thorough knowledge of the amino acid composition of foods. To elaborate, beans are low in the essential amino acids cystine and methanione but they're fairly high in all others including lysine. As mentioned above, rice is limited in lysine. By eating rice and beans together, one is complementing the rice with the beans, thus obtaining all the essential amino acids at the same time. Furthermore, legumes, whole grains, nuts and vegetable proteins provide a satisfactory combination of the amino acids for strict vegetarians. It must be stressed however that these have to be eaten at the same meal. By the same token, when appreciable amounts of plant proteins are ingested with small amounts of animal protein (such as milk or cheese), the resulting quality of protein is as effective as if only animal protein had been fed.

From the other end of the spectrum and in spite of evidence showing that Americans ingest far more protein than require, the average consumer continues to find his way to the drug store to buy "protein powders" and protein supplements. Some people are unfortunately led to believe that protein supplements of a specific type will enhance their health or the quality of their lives. Athletes or people with conditions affecting the nervous system are often the targeted groups. In truth, there is no evidence to show that increased protein intake will enhance physical performance. Luckily, the body is able to rid itself of the excess of nitrogenous waste generated by too much protein intake, unless a kidney or a liver disease is present. In either of these cases, the individual can do himself substantial harm by consuming too much protein.

Neurologic Goals of Metabolism



4. MICRONUTRIENTS

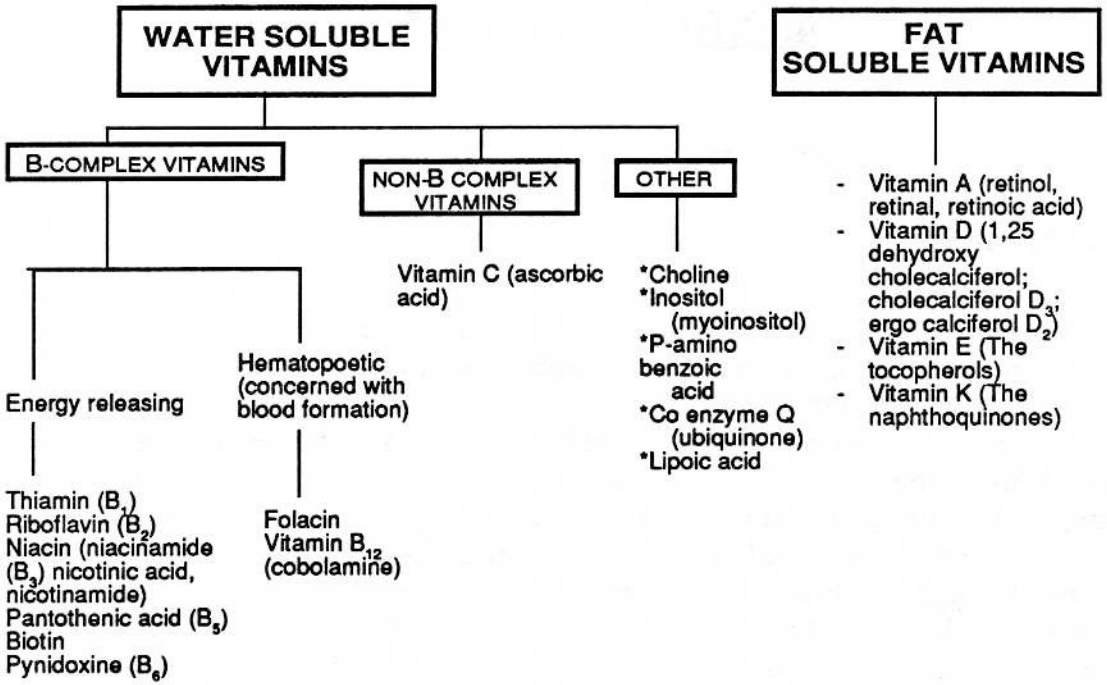
I. Vitamins

Vitamin is the name given to a group of potent organic compounds that along with certain minerals are essential in minute qualities for specific body functions of growth, maintenance and reproduction. None of the vitamins (with the exception of niacin, vitamin D, and Vitamin K) can be synthesized in the body hence they must be supplied in the diet. Vitamin D is synthesized in the skin with the help of the sun. Niacin and Vitamin K are made from the amino acid tryptophan and by intestinal bacteria, respectively.

Vitamins have gained a lot of attention, because of their association with the cure of specific diseases (eg. *beriberi* (thiamin), *pellagra* (niacin), and *scurvy* (vitamin C). The importance of vitamins in overall health should not be underestimated, but one must not be misled into believing that vitamins are cure-all substances. With the exception of certain disease states, the need and value of vitamin supplements are often exaggerated and the popularity of mega dose supplements is worrisome. While, it's been known for some time that mega doses of the fat soluble vitamins can result in toxicity, only recently have toxic effects and dependency states been associated with excess intake of the water soluble vitamins.

A daily multivitamin with mineral tablet is recommended for individuals whose diet lacks variety from all the food groups. It can also be recommended for those who follow a weight control regimen which restricts the total amount of food to be consumed. Mega doses of any vitamin or mineral, especially if it is to be taken as an individual nutrient supplement, are highly discouraged unless indicated by your physician.

The following tables summarize the vitamins in their major functions and dietary sources in order of content, the recommended allowances, the results of deficiency, and a special section that describes the results of overdosage or toxicity.



*Dietary substances not established as vitamins but often given vitamin status

I. FAT SOLUBLE VITAMINS

VITAMIN	MAJOR FUNCTIONS	MAJOR DIETARY SOURCE	RESULTS OF DEFICIENCY	OVERDOSAGE AND/OR TOXICITY	R.D.A. Male	R.D.A. Female
A	Essential for bone growth and development. Maintenance of skin and mucous membranes. Maintenance of visual purple and vision in dim light. Essential for normal reproduction.	Liver; cream; butter; egg yolk; carrots; sweet potatoes; squash; apricots; spinach; collards; broccoli; cabbages; dark leafy greens; cantaloupe.	Night blindness (early symptoms); retarded growth; cracked and decayed teeth	Painful bones and joints; dry scaly skin; headache; drowsiness; nausea; loss of appetite; loss of vision	1000 R.E.	800 R.E.
D	Necessary for the formation of normal bones in promoting the absorption of calcium and phosphorus from the intestinal lumen.	Fatty fish; eggs; liver; butter; fortified milk; exposure to sunlight.	Rickets (in children); *osteomalacia* (in adults); *muscular spasm and twitching*.	Loss of appetite; nausea; vomiting; thirst; calcification of lungs, kidneys and other soft tissue; resorption of bones	Children: Adults: Adults:	0-18 yrs: 10 mg 19-22 yrs: 7.5 mg 22- over: 5 mg
E	Potent *antioxidant*. Protects cell membranes, vitamin A, vitamin C, and *unsaturated fatty acids* from oxidation.	Salad oils; liver; fortified whole grain cereals; milk; eggs; butter; leafy vegetables.	In animals reproductive failure, muscular dystrophy and liver degeneration. In man *hemolysis* of red blood cells (mild anemia). *Impaired neuromuscular function, ataxia*.	Possible increase in blood pressure, inclusions in muscle	10 mg (14.9 IU)	8 mg (11.9 IU)

II. WATER SOLUBLE VITAMINS

VITAMIN	MAJOR FUNCTIONS	MAJOR DIETARY SOURCE	RESULTS OF DEFICIENCY	OVERDOSAGE AND/OR TOXICITY	R.D.A.	
					Male	Female
Thiamin (B_1)	Essential in the release of energy to cells from carbohydrates. Essential in digestion, fertility and normal function of nerve tissues.	Port; wheat germ; whole grains; enriched cereals; nuts; eggs; liver; oyster; grains; beans; peas; poultry.	Mental depression, *polyneuritis.* Peripheral disorder of cardio vascular systems. Neurologic disorders as in Wemieke-Korsakoff syndrome seen in alcoholics. Increased concentration of *pyruvic* and *Lactic acid* in the blood together with a low urine concentration of thiamin are also suggestive of deficiency.	Not reported yet	1.4 mg	1.0 mg
Riboflavin (B_2)	Promotes resistance to disease. Involved in maintenance of nervous system (*myelin* sheath). Releases energy to cells from water, protein and fats.	Dark green vegetables; liver and other meats; fish; eggs; milk; nuts.	*Somatitis* and other skin disorders especially around nose and lips and genitalia. Anemia and *neuropathy* are late developments. Burning and itching of eyes.	Not reported yet	1.6 mg	1.2 mg

VITAMIN	MAJOR FUNCTIONS	MAJOR DIETARY SOURCE	RESULTS OF DEFICIENCY	OVERDOSAGE AND/OR TOXICITY	R.D.A. Male	R.D.A. Female
Niacin (B_3)	Required in maintenance of NAD and NADP. Acts as *coenzyme* in lipid metabolism. Promotes healthy skin, nerves, appetite and digestion. Prevents pellagra.	Poultry; liver; fish; shellfish; nuts; whole grains; legumes; brown rice.	Pellagra inflammation of mucosa an intestinal tract, with vomiting and diarrhea, red beefy tongue, loss of appetite, weight loss. Sore tongue, mouth and throat with *glossitis*. Neurologic symptoms which include confusion, dizziness, poor memory, hallucination and irritability.	Flushing, stomach pain. Releases histamine which could aggravate peptic ulcer disease as well as asthma in patients who have either. *Hepatic* toxicity with release of *hepatic* enzymes in the plasma, homeostatic *jaundice*. Increased incidence of gouty arthritis since in high doses it competes with uric acid for excretion. Possible cardiac arrhythmia, pruritis and rash, increased blood glucose tolerance tests.	18 mg	13 mg
Pyrodoxine (B_6)	Required for absorption and metabolism of proteins and fat; involved in formation of red blood cells.	Liver; fish; meat; whole grains; cereals and breads; avocados; spinach; green beans; bananas; yeast.	Glossitis and other skin disorders. *Convulsions* in infants and anemia in adults. Nervous Irritability, *weakness, ataxia.*	*Sensory neuropathy* (with 2 gm/day for 4 mos. has been reported). Antagonizes L-Dopa in patients with Parkinson's disease. May cause dependency state.	2.2 mg	2.0 mg
Cyanoco-balamin	Essential in building of nucleic acids, formation of	Kidney; chicken; liver; lean meat; salt water	Pernicious anemia; *Neurologic degeneration*	Not reported yet.	3.0 mcg	3.0 mcg

VITAMIN	MAJOR FUNCTIONS	MAJOR DIETARY SOURCE	RESULTS OF DEFICIENCY	OVERDOSAGE AND/OR TOXICITY	R.D.A. Male	R.D.A. Female
(B_{12})	red blood cells and functioning of the nervous system.	fish; oysters; milk.	, *ataxia*			
Pantothenic Acid (B_5)	Essential coenzyme in metabolism of carbohydrates, proteins and fats, important in formation of hormones and acetylcholine.	Liver; kidney and other organ meats; eggs; whole grains; nuts; soybeans; dark green vegetables; yeast.	Vomiting; abdominal pain; sleep disorders; fatigue; *Neuritis; burning sensation of feet.*	Minimal occasional diarrhea and water retention.	4-7 mg	4-7 mg
Folic Acid (Folacin)	Aids in synthesis of nucleic acid and in the maturation of red blood cells.	Liver; kidney; food yeast; dark green leafy vegetables; nuts; grains; milk.	Megaloblastic anemia; anorexia; diarrhea and other symptoms of sprue and celiac disease which may produce folate deficiency.	May mask the hematological symptoms of pernicious anemia secondary to vitamin B_{12} deficiency which may be irreversible.	400 mcg	400 mcg
Biotin (H)	Involved (along wit other B vitamins) in formation of fatty acids and release of energy from carbohydrates, red blood cell maturation.	Egg yolk; milk; liver; kidney; green vegetables; grains; nuts; fish; milk.	Experimentally produced in man: Fatigue Depression Nausea Anorexia	Not reported yet.	No Standard	No Standard
Ascorbic Acid (C)	Essential for health of bones, teeth, gums, and blood vessels, formation of collagen. Improves iron absorption. Helps resist infection and aids in healing wounds.	Citrus fruits; berries; green vegetables; sweet peppers; new potatoes; melon; tomatoes.	Scurvy; bleeding gums; loose teeth; skin is rough, brown and dry; muscle degeneration; poor wound healing.	Kidney stones. Acidification of urine which may influence the excretion of other drugs. Increased lytic sensitivity of red blood cells (hemolysis). May impair bactericidal activity of white blood cells. Rebound scurvy causes	60 mg	60 mg

VITAMIN	MAJOR FUNCTIONS	MAJOR DIETARY SOURCE	RESULTS OF DEFICIENCY	OVERDOSAGE AND/OR TOXICITY	R.D.A. Male	R.D.A. Female
				Clinisticks to read false negative and the clinitest to read false positive for glucose. May decrease the absorption of vitamin B_{12}. Increase absorption of iron, and impair utilization of copper.		

II. **Minerals**

Minerals, like vitamins, are substances that are needed in very small amounts by the body for survival. Minerals were once considered to be contaminants in food, but are now known to be essential nutrients. As new analytical instruments that can detect trace concentrations are developed, the number of such minerals is expected to increase.

Calcium, phosphorus, sulfur, magnesium, iron, iodine, and zinc are the major minerals for which specific dietary allowances have been set. On the other hand, trace minerals such as copper, manganese, fluoride, chromium, selenium and molybdenum have had an "estimated safe and adequate daily dietary intake" suggested.

Mineral elements are present in organic compounds, inorganic compounds and free ions. They enter into the structure of every cell of the body. Minerals are also constituents of enzymes (such as iron in the cytochromes), of hormones (such as iodine in thyroxine), and of vitamins (such as cobalt in vitamin B12). Minerals in general play vital and varied roles in the body and are indispensable to survival.

For the normal individual, there is a balance that usually exists between the intake of a mineral and its excretion. Precise mechanisms conserve the amounts of minerals that are needed. At the same time a protective mechanism guards against an overload that might be toxic. There are limits to this, and toxic effects from minerals have occurred.

In selecting foods for their mineral content, several factors must be considered:

1. the concentration of the mineral in the food.

2. whether the food has lost some of its mineral through refinement or in the cooking process.

3. whether the food contains the mineral in an available form.

Selecting a variety of foods as discussed in the Four Food Groups is the best assurance to supply sufficient amounts of mineral elements. Fats, sugars, highly refined cereals, and flours are practically devoid of most mineral elements. Fabricated foods may lack important trace minerals for which no established recommended dietary allowances have been set.

Clinical deficiencies of mineral elements are not widespread because methods to evaluate the adequacy of mineral intake are limited. Often blood levels of the mineral is kept constant due to the body's ability to mobilize its own stores if the intake of the mineral is low (eg. the body mobilizes calcium from the bones when the intake is low in order to keep the blood calcium within a normal range at the expense of bone, an example of *homeostasis*). Because of these stores, it generally takes a long time for a clinical deficiency to occur. Since many minerals are integral constituents of certain enzymes, in some instances measuring the enzyme levels can give a fairly reliable index of the adequacy of the mineral in the body.

III. MINERALS

MINERAL	MAJOR FUNCTIONS	MAJOR DIETARY SOURCE	RESULTS OF DEFICIENCY	OVERDOSAGE AND/OR TOXICITY	R.D.A. Male	R.D.A. Female
Calcium	Plays vital role in controlling the excitability of peripheral nerves for heart function and muscle contractibility. Essential component of bones & teeth. Involved in blood clotting.	Milk, yogurt, sardines, salmon with bones, hard cheese, cottage cheese, turnips, collards, kale, mustard greens, broccoli, oysters, shrimp.	Associated with osteoporosis, *muscle twitching a cramping.*	Hypercalcemia; calcifications of soft tissue; renal stones.	800 mg	800 mg
Phosphorus	Component of DNA and RNA, the substances that control heredity. Involved in energy metabolism. Needed for formation of bone mineralization.	Meat; fish; milk; eggs; cheese; legumes; nuts; whole grain cereals	Poor bone mineralization, *rickets* in children. Occurs with prolonged use of absorbable antacids and is characterizes by weakness anorexia, malaise and pain in the bone, neurological and psychiatric disorder in adults. Hypercalcemia.	Not seen in humans with normal parathyroid and kidney function.	800 mg	800 mg
Manganese	Essential part of several enzyme systems involved	Nuts; unrefined grains; vegetables and fruits;	Poor reproductive performance;	Adverse effect on central nervous	2.5-5 mg	2.5-5 mg

MINERAL	MAJOR FUNCTIONS	MAJOR DIETARY SOURCE	RESULTS OF DEFICIENCY	OVERDOSAGE AND/OR TOXICITY	R.D.A. Male	R.D.A. Female
	in protein and energy metabolism.	legumes.	growth retardation; congenital malformation in the offspring. Abnormal formation of bone and cartilage.	system when element is injected or inhaled; Parkinson's disease-like syndrome		
Fluoride	Vital component in bones and teeth enamel.	*Fluorated* water; small ocean fish; tea	Tooth decay, *periodontal* disease; osteoporosis	Mottling in the teeth in children; fluorosis; crippling skeleton deformities; osteosclerosis	1.4-4 mg	1.4-4 mg
Magnesium	Essential component of many enzyme systems. Essential in maintaining electrical potential in nerve and muscle membranes. Constituent of bones and teeth.	Whole grain cereals; nuts; legumes; meat; milk; green leafy vegetables.	*Neuromuscular dysfunction* and psychological abnormalities. Hypomagnesemia seen mostly in malabsorption, renal disease and alcoholism.	Diarrhea	350 mg	300 mg
Iron	Major constituents of hemoglobin, *myoglobin* and a number of enzymes.	Liver; navy beans; meat; egg yolk; whole grain bread; vegetables; peaches; apricots; prunes.	Iron deficiency anemia.	Iron poisoning.	10 mg	18 mg
Zinc	Essential constituent of enzyme involved in most	Animal meats; seafoods (particularly oysters);	Loss of appetite; failure to grow; skin	Acute gastrointestinal	15 mg	15 mg

MINERAL	MAJOR FUNCTIONS	MAJOR DIETARY SOURCE	RESULTS OF DEFICIENCY	OVERDOSAGE AND/OR TOXICITY	R.D.A. Male	Female
	major metabolic pathways.	eggs; liver; wheat germ.	changes; impaired regeneration of wounds; decreased taste activity and *dysgeusia*; Hypogonadism and dwarfism.	irritation and vomiting. May aggravate marginal copper deficiency.		
Iodine	Integral part of thyroid hormone (thyroxin) which has important metabolic roles	Seafood; iodized salt.	Goiter	May cause hyperthyroidism	150 ug	150 ug
Copper	Constituent of a number of enzymes. Essential in iron metabolism. Integral part of *ceruloplasmin*.	Oysters; nuts; liver; kidney; corn oil; margarine.	*Hypocupremia* observed in protein-calorie malnutrition; anemia, *spasticity.*	Abnormal storage in Wilson's disease	2-5 mg	2-5 mg
Chromium	Helps maintain normal glucose metabolism.	Brewer's yeast; meat products; cheeses; whole grains and condiments i.e. thyme; black pepper.	Impaired glucose tolerance	Not reported yet.	0.05-0.2 mg	0.05-0.2 mg
Selenium	Essential component of enzymes that protect cells against oxidative damage. Maintenance of muscle and red blood cells integrity. Ameliorates the toxicity of other metals i.e. mercury, arsenic, silver,	Seafood; kidney; liver; meat.	Keshan disease characterized by abnormalities in the heart muscle, reported in China where plant foods are low in selenium.	Alkali disease in cattle grazing in soils high in selenium content, characterized by stiffness, blindness, deformity of the hooves, and loss of	0.05-0.2 mg	0.05-0.2 mg

MINERAL	MAJOR FUNCTIONS	MAJOR DIETARY SOURCE	RESULTS OF DEFICIENCY	OVERDOSAGE AND/OR TOXICITY	R.D.A. Male	Female
	and copper.			hair.		
Molybdenum	Cofactor for flavoprotein enzymes. Interrelated metabolism between molybdenum, copper, and sulfur.	Organ meats; grains; legumes.	Not reported yet.	Antagonizes copper. Gout-like syndrome.	0.15-0.5 mg	0.15-0.5 mg

5. TARDIVE DYSKINESIA—an example of nutritional therapy

People often note benefits from over-the-counter dietary supplements (Vitamin C in cold prevention, vitamin E for scar healing, calcium for leg cramps), long before advantages can be shown scientifically.

But up until recently, there has been no evidence that modifications of dietary intake could improve the symptoms of a neurologic disorder independent of cause. In the past decade or so, many hypotheses concerning nutrition in brain function have emerged and been tested. These include:

1. High linoleic acid intake in reducing the frequency and duration of *relapses* in patients with Multiple Sclerosis. (Brown et al, 1980)

2. Chronic ingestion of a low *tryptophan* diet to reduce levels of brain *serotonin* and demonstrate a heightened sensitivity to painful stimuli in laboratory animals (implying a use for tryptophan supplements to block pain). (Fernstrom, 1977)

3. Use of a high *choline* diet to alleviate the symptoms of Tardive Dyskinesia. (Growdon et al, 1977)

4. Use of minhydrin oil to replace arachidonic acid and ameliorate neurologic symptoms of deficiency. (Auerbach et al, 1989)

Because of our own experience with (3) and the amount of scientific research that has been done with it, it will be used as an example and will be the scope of discussion of this chapter.

Tardive Dyskinesia is an *extrapyramidal syndrome* caused by the chronic administration of major *neuroleptic* agents (haloperidol and other drugs used for psychosis). It is characterized by abnormal involuntary persistent movements of the tongue, lips and facial muscles, as well as abnormal muscular manifestation in the fingers, hands, arms, feet and *diaphragmatic* movements (resulting in difficulty in breathing).

Choline, a nutrient found in food, is the precursor for acetylcholine, the *neurotransmitter* found to be low in the brain cells of patients with Tardive Dyskinesia. (Klawans et al, 1980)

The brain is limited in its ability to make choline and it has been demonstrated that the administration of purified *lecithin* (choline is a component of lecithin) or a high choline diet resulted in increased serum and brain choline levels, as well as brain acetylcholine, in laboratory animals and human subjects, (Wurtman et al, 1977 and Zeisel et al, 1980), including patients with Tardive Dyskinesia, (Growdon et al, 1977). It appears that when blood levels of choline are high enough, an *influx* of choline through the *blood brain barrier* in the brain reacts with acetyl CoA (a product of food metabolism) and increases acetylcholine supplies at the nerve endings (i.e. Choline + Acetyl CoA = Acetylcholine).

This suggests that food ingredients which are precoursers of important neurotransmitters, when taken in the diet in large amounts, can indeed penetrate the blood brain barrier and increase the levels of that neurotransmitter. Theoretically then, dietary supplements such as lecithin or choline may be given to patients with disease in which cholinergic neurotransmitters are low. Researchers went on to test this theory in:

1. Huntington's Disease: in this disorder choline supplements have shown no amelioration of symptoms because HD involves degeneration of *postsynaptic* reception (receptors that interact with acetylcholine are gone). (Aquilonius et al, 1977)

2. Alzheimer's Disease: treatment with lecithin has been shown to be effective in mild memory loss in a small percentage of the participants. But it has not been proven that choline (administered as choline chloride or as lecithin) improves memory, in this dementia characterized by an early and disproportionate loss of cholinergic neurons. (Boyd et al, 1977)

3. Tardive Dyskinesia: studies have shown between 50% and 70% success in reducing involuntary facial movements with choline supplements. (Growdon et al, 1977)

Note that brain acetylcholine can be increased by several mechanisms. These are:

1. Choline supplements: however, when taken in large amounts may cause the person to have a potent fishy odor. Furthermore, this type of choline is quickly cleared from the body before it is able to have any long lasting effect on the level of acetylcholine in the brain.

2. Lecithin supplements: here the only type that is able to make any difference at the brain level is the "purified" form which is only available for experimental research and not for consumers. So one is wasting his time and money when buying the "over the counter" lecithin, which, because of its large content of impurities is unable to pass the blood brain barrier in sufficient amounts to make a difference in the levels of acetylcholine.

In either form of choline supplement listed above, common side effects were nausea, diarrhea and low blood pressure.

Wurtman and his associates have shown that oral lecithin is considerably more effective in raising human serum choline levels than an equivalent quantity of choline chloride.

3. High choline diet: Growdon and his associates at MIT found that high choline diets were more efficacious than either of the above in raising serum choline, brain choline and brain acetylcholine in normal subjects as well as in patients with Tardive Dyskinesia and that these increases tended to stay high longer than with the other techniques. This carries the good news that what we eat could indeed make a difference at the neurotransmitter levels in the brain. However, much more research is needed in the domain of nutrition and brain function.

Choline and nutrition:

Because of its widespread occurrence in foods, it would be virtually impossible to eat a regular diet low in choline that is not also lacking other nutrients. Moreover, no clinical evidence of a dietary choline deficiency has been found, so it's not possible to state that choline is an essential supplement for man.

Over the counter, especially in health food stores, choline is falsely claimed to lower blood lipids and fat deposition in the body. While it is true that inducing choline deficiency in mammals by its exclusion from the diet and by the exclusion of the nutrient essential for its synthesis (an amino acid called methionine) has been found to cause:

1. fatty infiltration of the liver;

2. *hemorrhagic* kidney damage.

Both of the conditions listed above are reversible with the administration of choline only when these are caused by choline deficiency. Unfortunately, it does not work that way in, for example, alcoholic patients with fatty liver disease of other causes, as choline supplements have been given to such patients with absolutely no results. (Goodhart and Shils, 1980)

Foods that have the highest choline content are eggs, soy beans, milk, peanuts, liver and chocolate. High choline diets are however most efficacious in raising serum choline when coupled with choline supplements in the purified form and these cannot be bought over the counter. (Zeisel et al, 1980)

6. The ALD Diet Plan

Many "inborn errors of metabolism" have been identified (most in children), where an inherited alteration in the body's ability to metabolize nutrients is the cause of a degenerative neurologic process. Supplementation and restriction diet therapies are often tried in these cases. For example, addition of carnitine is used for the carnitine deficiencies. Elimination of certain amino acids or proteins with addition of certain B vitamins is tried for many amino acid and urea cycle disorders (as in the treatment of phenylketonuria). The experimental diet treatment of glutamate dehydrogenase deficiency is being tested in certain of the OPCA's.

The Kennedy Institute at John Hopkins is currently running an extensive research program on Adrenoleukodystrophy (ALD) and Adrenomyeloneuropathy (AMN), which are sometimes adult-onset, genetically determined ataxic disorders that are associated with the accumulation of saturated very long chain fatty acids (VLCFA), particularly C26 (26 carbon backbone). The fatty acids that accumulate are at least partially of dietary origin. It would be important to determine in a disease caused by known build-up of harmful dietary substances or metabolic product that a partial cure (or compensation for the cause) could be affected by dietary modification (as was shown for Refsum's disease and phytanic acid). Therefore, the ALD/AMN diet is a research diet that restricts the total dietary intake of C26 fatty acids to 3mg or less. This can be accomplished by severely restricting fatty foods as well as foods that have a high content of C26. (Van Huyn et al, 1984; Suzuki et al, 1986; Rizzo et al, 1986). While it eliminates several food items within a particular food group (even in the fruit and vegetable groups), this diet is by no means unbalanced, since it contains a satisfactory number of foods within each of the Four Food Groups. Prior to commencing this diet, a detailed nutritional evaluation is required. The number of servings from the different food groups are then determined based on the patients' medical and nutritional profile which would ultimately determine his/her specific caloric and protein requirements. Use of a special oil (called glycerol trioleate, available through the mail by special order) is also required. This oil contains the long chain fatty acid C18, which has been observed to decrease the synthesis of the very long chain (C26) fatty acids while increasing the total fat, and therefore caloric content of the diet.

Three (3) fasting blood samples for VLCFA on three separate days prior to starting the diet are requested by the Kennedy Institute and then later compared with future blood samples while on the diet, as the patient's disease condition is monitored.

7. The "Ataxia Diet"

In the ataxic illnesses, amelioration of symptoms has been sought by use of lecithin and choline supplements because the neurotransmitter acetylcholine (choline is the precourser of acetylcholine) is felt to be involved in the normal coordination pathways. However, very few metabolic cause of these disease have been discovered to date, so more specific research diets have not been developed.

The Ataxia Center at NPI/UCLA has developed a carbohydrate-restricted diet for many of its neurologic patients, to enhance general conditioning, symptom-relief, and possible changes in disease progression. We restrict the carbohydrate portion of the diet, to the extent of eliminating refined flour and sugar products (table sugar) from the daily meal pattern. This includes molasses, brown sugar, corn syrup, jams, jelly and even unsweetened fruit juices, as well as white bread products, muffins, cakes, pies, donuts, etc.

Two reasons for these restrictions are:

1. Sub-groups of FA patients have a higher incidence of insulin-resistant diabetes. Insulin is the hormone that is responsible for the uptake of blood sugar by the different body tissues. When this process is slow, the sugar stays in the blood and shows up in the urine, as in other forms of diabetes, with the risk of similar medical complications. One of the integral parts

of the medical management of diabetes is controlled carbohydrate intake, with the elimination of concentrated sweets and maintenance of ideal body weight.

2. Subgroups of FA and related disorders have been thought to have difficulty turning the products of glucose metabolism into energy, possibly due to changes in the activity of two or more major enzyme systems [pyruvate dehydrogense complex (PDHC), pyruvate carboxylase (PC) and malate dehydrogenase (MDH)]. The cerebellum and its connections are some of the areas of the brain most sensitive to this kind of deprivation (an "internal energy crisis"), especially in proven cases of PDHC deficiency. (Blass, 1979.) Blood tests are available or being developed to better identify the various degrees of this metabolic problem and alert the clinician to the possible role of dietary restriction of carbohydrates in case management (allowing other energy sources to be substituted by the brain and limiting build-up of metabolic wastes). We recommend the Ataxia Diet to those who have abnormalities in glucose use or in its metabolites, following a Glucose Tolerance Test (GTT).

The Ataxia Diet is by no means a low carbohydrate diet. The difference lies in the type of carbohydrates rather than in the amounts. While it eliminates simple carbohydrates such as those found in cookies, cakes, candies, or the like, it allows complex ones such as those found in rice, pasta, unsweetened fruits, legumes and starchy vegetables. These foods, although still high in carbohydrates, have a much lower glycemic effect than the former ones (or in other words, they do not go into the blood as fast). The amount of these allowed in the diet is determined by the dietician, based on the nutrition history and whether or not the individual is obese.

Protein in the Ataxia Diet, comes from lean fresh or fresh frozen meats. The inclusion of processed meats (such as cold cuts, hot dogs, bacon, etc.) is highly discouraged, not only due to their high fat and salt content but also their high concentration of food additives and preservatives (an impact of these on brain function has been suggested although not proven).

FA and related disorders are not hypermetabolic states (i.e. do not result in increased caloric requirements, although Huntington's disease does), and maintainence of an ideal body weight is especially crucial to facilitate walking and transferring. To aid this, fat in the Ataxia Diet is limited to the hidden forms (in lean meats, fish, and poultry and low fat dairy products). Any additional fat would depend on the total caloric requirements of the individual patient and is determined by the dietitian.

Overall, 3 meals per day that are complete in all four food groups and are properly spaced, are highly recommended. This allows the body to receive a more steady flow of energy from exogenous food sources. Not everyone that is placed on this diet benefits from it to the same degree. While some people feel no difference at all, most everyone that complies with this diet reports one or more of the following:

a. Weight loss: this is largely due to the elimination of fats, fried foods and pastries, all of which are very calorically dense, while being limited in nutritional value. Complex carbohydrates, such as fruits, vegetables and legumes are also rich in *dietary fiber* (such as guar and pectin). These have been associated with prolonged *satiety* and a feeling of fullness that comes about faster during meal ingestion and lasts longer than that from simple carbohydrates, thus facilitating weight loss by suppressing appetite.

b. Improved walking and easier transferring: as one has less weight to carry, a more constant supply of energy, and no symptomatic swings in blood sugar.

c. Easier bowel movements: this again is due to the naturally high fiber content of the food items allowed and also to better activity levels.

d. Improved mood and spirit: this could in part be due to the nutritional soundness of the diet and the benefits in a, b, and c. It may not be unreasonable to suspect that this could also be the result of improved substrate utilization by the different tissues and organs including the brain and the peripheral nervous system.

(Dr. Perlman, 2009--The "Ataxia Diet" as we have recommended it, has features of common sense nutrition and includes many dietary tools that have remained in use today—glycemic index of foods; fiber; small, frequent meals; weight maintenance. No diet has yet been shown to cure ataxia.)

8. Nutrition and Huntington's Disease

In today's society where almost everyone is struggling to "watch their weight", patients with Huntington's disease have the opposite problem.

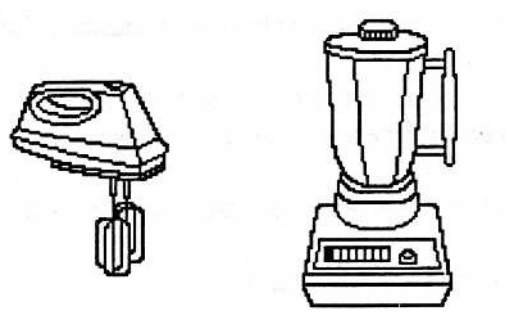
Because of their uncontrollable choreic movements and increased metabolism, they may burn almost twice as many calories as do their counterparts who do not have this disease. It is often a challenge to prevent the patient with HD from losing weight, and skipping even one meal can make a difference. Luckily, this increase in *basal metabolism* is almost always coupled with an increased appetite.

In this situation, the family or care taker should encourage the patient to consume as much food as they desire, supervising only the type of food made available. Weekly weights should be taken and any weight loss reported to the dietitian.

Aside from the choreic movements, which can make food handling quite difficult, the patient with HD often faces a more serious problem: that of choking while eating.

Let us first consider the food handling. Finger foods, ie. Those that can be handled without utensils such as sandwiches, are clearly the ones of choice, especially when assistance during meal time is limited. For items such as soups, cereal, or liquids, special utensils and flatware for the physically challenged are available through your occupational or physical therapist.

As for the swallowing problem, we have found that in more cases than not, food items that are dry and crumbly (eg. cookies, plain cake or dry hamburger) as well as those with multiple textures (eg. salads with several vegetables and dressings, soup with pieces, watermelon) present the most challenge and difficulty during swallowing. Uniformly textured food may be better handled and should be consumed with small bites that are swallowed before the next bite is taken. Distraction and talking during chewing and swallowing should be avoided, as an additional safeguard. Unfortunately, it is not uncommon for the patient with advanced HD to require a completely pureed diet.



Meal Pattern

The aim is six meals per day (three large meals and three in between snacks). These meals should be well balanced and contain at lest one item from each of the four food groups at each sitting.

Unlike the patient with FA, restricting table sugar does not seem to be a major issue, where keeping the weight on is the challenge. This is however not to say that HD patients should have their calories from sweets. Indeed, one can plan a meal pattern that has up to 4,000 calories without necessarily incorporating junk food or empty calories.

A typical meal pattern for HD would be the following:

Breakfast:
8 oz. regular milk
hot cereal (softened in milk)
3 toast slices with 3 pats of butter or margarine
8 oz. orange juice
2 eggs or 2 oz. of any leftover meat

Lunch and Dinner:
*4 oz. of poultry, fish or meat (finely chopped, ground or pureed depending on the competence of the swallowing mechanism)
2 servings of rice, potatoes or noodles (well cooked with lots of gravy)

2 slices of breads with 4 pats of butter or margarine
*2 servings of vegetable (well cooked or pureed)
8 oz. of milk

In between meal snacks:
Sandwich (2 slices of bread, 2-3 oz. of cheese or meat and mayonnaise or chicken salad, egg salad or tuna salad*)
Or: unsweetened peanut butter and jelly sandwich with a glass of milk
Or: large cottage cheese and canned fruit plate with American or Swiss cheese cubes*
It should be noted that liquids may be safest taken through a straw.

*may be pureed in severe swallowing difficulties

9. Coping with Swallowing Difficulties

Very few things in life are more frightening than choking while drinking or eating. Afraid to eat, an individual with severe swallowing problems can soon show weight loss and signs of malnutrition and dehydration.

Swallowing disorders and *dysphagia* are not uncommon in patients with incoordination problems. One of the keys is to identify the types of foods that one has a problem handling and subsequently avoid these and/or change their consistency to one better tolerated. We found the majority of our FA patients who have difficulties with swallowing have the most choking episodes with thin liquids (examples of which are juices and plain water) as well as with dry crumbly foods (examples of which are dry cake or corn bread). Once these "culprit foods" have been identified, efforts can be made to handle them.

Anyone with swallowing difficulties can actually identify the type of food that they may have difficulties with, through taking the "Bolus Specificity quiz" on the next page.

Another point to note is the head position one assumes while eating. To best identify the position that's associated with the least likelihood of choking, a cinesophogram or modified barium swallow study (MBSS) can be performed. This is a test where one ingests food items (usually 4 are used: liquid, pureed, cookie-type and paste-like consistencies), that are coated with a fluoroscopic solution. The swallowing mechanism is then followed on video. This can be interpreted by the clinician, who will help determine exactly where the defect is taking place. The clinician can also offer recommendations for different head positions to compensate for the problem (as well as the consistency of food that causes the lowest risk of choking). In general the head should be tilted slightly down (chin tuck) to protect the airways during swallowing. Clearing of the throat with sips of liquid between bites may be recommended.

One should not lie down for at least 2 hours after a meal, to avoid *regurgitation* and possible choking.

It is also important to eat slowly and in a calm quiet place with full concentration on what's being chewed and about to be swallowed.

Through practicing these few simple steps, a person with impaired swallowing can learn to adapt and minimize episodes of choking.

Bolus Specificity Quiz

BOLUS SPECIFICITY	BOLUS CHARACTER-ISTICS AND/OR EXAMPLES	NO DIFFICUL-TIES	SOME DIFFICUL-TIES	GREAT DIFFICUL-TIES
I. Temperature	Hot	. . . . .	. . . . .	. . . . .
	Warm	. . . . .	. . . . .	. . . . .
	Cold.	. . . . .	. . . . .	. . . . .
II. Taste	Strong	. . . . .	. . . . .	. . . . .
	Mild	. . . . .	. . . . .	. . . . .
	Bland	. . . . .	. . . . .	. . . . .
III. Consistency				
a. Viscosity (stickiness)	Juice	. . . . .	. . . . .	. . . . .
	Milkshake	. . . . .	. . . . .	. . . . .
	Yogurt	. . . . .	. . . . .	. . . . .
	Pudding	. . . . .	. . . . .	. . . . .
b. Particulateness (Crumbliness)	Mashed Potatoes .	. . . . .	. . . . .	. . . . .
	Plain cake	. . . . .	. . . . .	. . . . .
	Cookies	. . . . .	. . . . .	. . . . .
	Hamburger	. . . . .	. . . . .	. . . . .
c. Multiplicity of Textures	Macaroni & Cheese	. . . . .	. . . . .	. . . . .
	Beef Stroganoff . . .	. . . . .	. . . . .	. . . . .
	Salad with bleu cheese dressing . .	. . . . .	. . . . .	. . . . .

10. Irregular Bowel Movements

Constipation is an all too often encountered problem when activity levels are reduced, due to use of a wheelchair, or even in the older individual. It is easy for people who fall into these categories to become "laxative dependent", accustoming the gastrointestinal tract to the chemical stimulation of laxatives.

Products containing phenolphthalein (Correctol, Ex-Lax, Feen-A-Mint) or bisacodyl (Carter's Little Pills, Dulcolax) irritate the inner lining of the intestine, creating artificial *peristalsis* that moves the fecal mass through. This, like chronic diarrhea, can result in poor absorption of micronutrients and water contained in the stool. Use of mineral oil can wash out fat soluble vitamins. A dependent state results, when these laxatives are taken on a regular basis: anywhere from 1 pill per week to 2-3 per day.

In extreme cases, such as the person who has been taking laxatives regularly for 15 to 20 years, this state of dependence becomes irreversible even with increased fiber in the diet. Regular use of laxatives should be highly discouraged.

A high fiber diet has historically been used to treat and relieve constipation. Foods that have a high content of non absorbable material (commonly referred to as "high fiber foods", containing one or more of the following: *cellulose, hemicellulose, pectin, guar* and bran) are incorporated into this diet. These substances, which are unique to whole grain products, fruits, vegetables, and legumes, relieve constipation through absorbing and holding on to water in the bowel, increasing the net weight of the stool and softening them, making them much easier to pass.

Aside from the fact that a diet rich in fiber does alleviate constipation, other benefits have also been show to result from a high fiber diet. These included prolonged relief of hunger after a meal, decreased blood sugar level and decreased glucose in the urine of diabetics. Decreased incidence of colon cancer (due to the shortened transit time of stool through the bowel that results from high fiber diets) may also be a benefit. Most recently, these high fiber diets (especially oat bran) have been suggested to lower blood cholesterol, the mechanism of which is not well understood, but seems to be related to the fiber's ability to bind the bile in the small intestine and decrease cholesterol synthesis in the body.

Most people generally know that fruits and vegetables are high in fiber, but very few realize that items within these two groups vary widely in their actual fiber content. The following is a list of the highest sources of fiber:

Vegetables: peas, baked beans, parsley, turnip greens, yams, lentils, mustard greens, broccoli, eggplants, beets, mushrooms, parsnips.

Fruits: stewed prunes, dried peaches, blackberries, whole oranges, white grapes, raspberries, cranberries, apricots, dried prunes.

Whole grain products: wheat bran, all bran, crispbread or rye crackers, shredded wheat biscuit, whole wheat bread, soya flour. Oatmeal and oat bran also fall into this category.

In severe cases of constipation, one may not seem to be able to get enough roughage to achieve regular bowel movements and frustration may result. It is for those people that the Bran Plan has been developed.

On the supermarket shelves there is an inexpensive, unattractive, yellow box in the cereal section with the title "Miller's Bran". It has more fiber per serving than any other food item available. The reason why not too many people bother with it is because they just don't know how much or even how to eat it since it looks and tastes like saw dust. In addition, tremendous care in its care should be taken if the patient has swallowing problems. Raw bran (or miller's Bran) is hardly an appetizing "food" item to ingest, but the majority of our patients that have been placed on it have accomplished regular bowel movements and some were able to be weaned off laxatives completely. To them, this is worth the "unpleasant" experience of ingesting it on a daily basis.

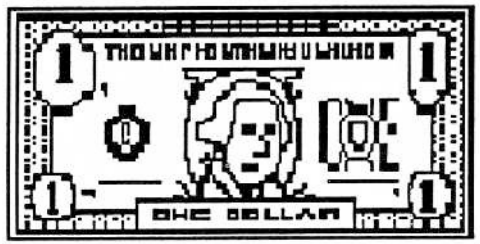
We find the easiest way to take any of these fibers is to mix them in a semi solid food item such as applesauce or oatmeal. One should then swallow the mixture without attempting to chew it. Starting out with 1-2 teaspoons per day, preferably in the morning, and increasing slowly over a period of 2 weeks not to exceed ¼ cup has proven helpful to most of our patients. If abdominal distress such as cramping, bloating and/or flatulence develop, one must decrease the dosage or discontinue it altogether and report to the physician or the dietitian. Drinking adequate amounts of water during the day cannot be overemphasized. The easiest way to remember to drink water is by taking in 4 oz of water before eating or drinking anything else.

Therefore, a high fiber diet, one that contains at least one serving of the foods listed above per meal, coupled with 1, 2 or 3 tablespoons of fiber supplement per day (depending on the individual), taken as directed, can be the cure for constipation from which many millions of Americans currently suffer.

We've also tried a few of our patients who had occasional episodes of diarrhea on this Bran Plan, and it has helped in its alleviation. Nevertheless in persistent and severe cases of diarrhea or if there is trouble with bowel control, a physician should be consulted.

11. Eating Well on a Budget

It is not uncommon for someone with a disability that limits his income to be on a strict budget which could affect the very food choices he makes. This however does not need to result in malnutrition. Milk and eggs happen to be the two protein sources that have the highest *biological value* of all and yet they're among the cheapest items on the supermarket shelves. Nonfat skim milk powder can make an excellent protein supplement, when added to regular milk for those who need it (consult with your dietitian). Candy, cookies, cakes or ice cream can cost up to twice and sometimes three times as much as chicken or fish per pound! So, to quiet the sweet tooth, consider fruit in season, when bargain prices can be found. Even noodles, rice, legumes and vegetables, all complex carbohydrates, are relatively cheap yet nutritious food items that will cut down on hunger pangs longer than the simple carbohydrates above. Cookies, donuts and commercial cakes and pies do have the distinct advantage of both a relatively long shelf life as well as being finger foods that are easy to grab at and eat! While those are difficult points to ignore, arguments for not having them around the house are not easy, unless substitutes are considered. So how about stocking the refrigerator with cottage cheese and fruit containers, unsweetened peanut butter on carrot sticks or cheese cubes on whole grain crackers! Even vitamin pills (if recommended) can be bought at a bargain. There is no evidence to suggest that name brand vitamin preparations are better than the generic ones. Ironically enough, some of the multi million dollar pharmaceutical companies that make the brand name vitamins are the same ones making the no name ones and sell both to your neighborhood drugstores under different names and different prices! So it doesn't take a millionaire to prepare well balanced and complete meals.



12. Malnutrition

Malnutrition is a state that can develop in an individual who, over a period of time, has failed to meet his or her caloric and/or protein requirements.

Contrary to most peoples' belief, malnutrition does not have to take on the classical presentation seen in pictures from India or Africa.

Malnutrition comprises three categories:

1. *Marasmus* type: the individual is noticeably underweight, but his blood proteins and his vitamin levels are within a normal range. The hazard of this state, with limited fat, and skeletal muscle reserves, becomes most significant when the person becomes ill or has to have surgery or be hospitalized for a length of time. Hospital foods have never had high ratings in palatability, and it is often a common occurrence that the person will be NPO (nothing by mouth) for at least a couple days depending on the tests or procedures he is to have. A small fat reserve and good skeletal muscle mass would definitely come in handy for situations like that and speed recovery. It must also be stressed that age makes a big difference as the older the person is, the more likely he is to have complications and delayed recovery when malnutrition is in the picture.

2. *Kwashiorkor* type malnutrition: an overweight or even obese individual may actually be "protein-malnourished" when his blood protein (such as his serum *albumin*) measures low. The danger of this lies in the impaired capacity to ward off infection and maintain normal body fluids. It has been well documented that low serum albumin goes hand and hand with increased mortality in hospitalized patients.

   A diet that is deficient in protein over a period of time would result in kwashiorkor type of malnutrition.

3. Marasmus-Kwashiorkor or Protein-Calorie Malnutrition: is basically a combination of the first two and is usually the hardest to treat, especially if it is far gone. It is often the result of prolonged inadequate intake of nutrients.

Therefore, a well balanced diet that is adequate in both macro as well as micronutrients cannot be overemphasized if one is to stay in good health or to recover from illness when it ensues.

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14. Glossary

Adipose: fat, fatty

Albumin: a protein in tissues and body fluids soluble in water and coagulated by heat; principal protein in blood regulating osmotic pressure and its level in the blood is often indicative of the individual's protein nutrition status.

Amino Acids: these are the building blocks of proteins, i.e. eggs, fish or meat are all made up of amino acids combined in different configuration to give them their particular characteristics.

Anemia: deficiency in the circulatin hemoglobin, red blood cells or packed cells volume.

Antioxidant: a substance that prevents deterioration by hindering oxidation as in tocopherols preventing oxidation and rancidity of fats.

Arachidonic acid: a 20-carbon fatty acid with four double bonds, it is the physiologically active fatty acid.

Ataxia: loss of ability of muscular coordination.

Basal metabolism: energy expenditure of the body at rest in the post-absorptive state.

Beriberi: a deficiency disease caused by lack of thiamin and characterized by extreme weakness, polyneuritis, emaciation, edema and cardiac failure.

Biologic value: a measure of the effectiveness of a nutrient, such as protein, in the living organism.

Biotin: a vitamin of the B complex; participates in fixation of carbon dioxide in fatty acid synthesis.

Blood brain barrier: the barrier that prevents or delays the entry into brain tissue of certain substances in the blood. Presumably, it consists of the walls of the blood vessels of the central nervous system and the surrounding glial membrane.

Calcification: hardening of tissue by a deposit of calcium and also magnesium salts.

Calorie: a unit of heat measurement; in nutrition, the kilocalorie is the amount of heat required to raise the temperature of 1Kg of water 1 degree Centigrate.

Carotene: precursor of Vitamin A; yellow plant pigments occurring abundantly in dark green leafy and deep yellow vegetables.

Cellulose: the structural fibers of plants; an indigestible polysaccharide.

Ceruloplasmin: copper-containing protein in blood plasma.

Cholesterol: A sterol fond in animal foods and made within the body; a constituent of gallstones and of atheroma.

Choline: A nutrient found in many animal and some vegetable tissues. Required for the synthesis of acetylcholine. Synthetic preparations of choline derivatives are used as parasympathetic stimulants and act to increase the heart rate, contract smooth muscle tissues and increase secretion of most glands.

Chylomicrons: large molecules of fat occurring in lymph and plasma after a fat rich meal; consist of triglycerides attached to a small amount of protein.

Cobalamine: compound containing cobalt grouping found in B-12.

Coenzyme: prosthetic group of enzyme; for example, a vitamin that conjugates with a protein molecule to form an active enzyme.

Diaphragmatic: pertaining to the diaphragm which is a strong dome shaped muscle that separate the thoracic and the abdominal cavities and is involved in breathing.

Dietary fiber: plant fibers that include cellulose, hemicellulose, lignin, mucilages and gums, and pectin.

Diglyceride: a fat containing 2 fatty acid molecules.

Disaccharide: a carbohydrate that yields 2 simple sugars upon hydrolysis; sucrose, maltose and lactose.

Duodenum: first portion of the small intestine, extending from the sphincter between the stomach and the intestine to the jejunum.

Dysgeusia: perverted sense of taste; bad taste.

Dysphagia: difficulty in swallowing.

Enrichment: the addition of thiamin, riboflavin, niacin and iron to refined cereal and bread products.

Enzyme: an organic compound of protein nature produced by living tissue to accelerate metabolic reactions.

Essential amino acid: an amino acid that must be present in the diet as the body cannot manufacture it, ex: linoleic acid.

Exogenous: originating or produced from the outside.

Extrapyramidal: relating to motor control centers other than the motor command area (pyramidal).

Fatty acids: open-chain monocarboxylic acids containing only carbon, hydrogen and oxygen.

Fluoridation: the addition of fluoride to water to reduce tooth decay.

Folacin: folic acid; a vitamin of the B complex.

Folic acid: A vitamin of the B complex necessary for the maturations of red blood cells and synthesis of nucleoproteins; also known as folacin.

Fortification: the addition of a nutrient or a group of nutrients to a food that were not there in the first place, ex: vitamin D to milk.

Fructose: a 6-carbon sugar found in fruits and honey; also obtained from the hydrolysis of sucrose; fruit sugar.

Galactose; a single sugar resulting from the hydrolysis of lactose.

Glossitis: inflammation of the tongue.

Glucose: a single sugar occurring in fruits and honey; also obtained by the hydrolysis of starch, sucrose, maltose, and lactose; the sugar found in the blood ; dextrose; grape sugar.

Glyceride: organic ester of glycerol; fats of esters of fatty acids and glycerol.

Glycogen: polysaccharide produced from glucose by the liver or the muscle; “animal” starch.

Glycogenesis: formation of glycogen from glucose by the liver or muscles.

Glycolysis: the anaerobic conversion of glucose to pyruvic and lactic acids; an energy yielding process.

Goiter: enlargement of the thyroid gland.

Guar: a water soluble fiber.

Hemicellulose: indigestible polysacchrides that form the cell wall of plants.

Hemolysis: separation of hemoglobin from the red blood cells.

Hemorrhage: loss of blood from the vessels; bleeding.

Hemorrhagic: pertaining to hemorrhage.

Hepatic; pertaining to the liver.

Homeostasis: tendency to maintain equilibrium in normal body states.

Hormone: substance produced by an organ to produce a specific effect in another organ.

Hydrogenation: the addition of hydrogen to a compound, such as unsaturated fatty acid to produce a solid fat.

Hydrolysis: the splitting up of a product by the addition of water.

Hyper: a prefix meaning above, beyond or excessive.

Hypercalcemia: abnormally high calcium level in the blood.

Hypocupremia: low copper blood levels.

Hypoglycemia: a lower than normal level of glucose in the blood.

Ileum: lower portion of the small intestine extending from the jejunum to the colon.

Inositol: a 6-carbon alcohol found especially in cereal grains; combines with phosphate to form phytic acid.

Insulin: hormone secreted by a specialized area of the pancreas and which promotes utilization of glucose and lowers blood sugar.

Jaundice: a condition characterized by elevated bilirubin level of the blood and deposit of bile pigments in skin and mucous membranes.

Jejunum: middle portion of small intestine; extends from the duodenum to the ileum.

Ketogenesis: formation of ketones from fatty acids and some amino acids.

Ketone: any compound containing a ketone grouping.

Ketcsis: condition resulting from incomplete oxidation of fatty acids, and the consequent accumulation of ketones, acetone, beta-hydroxybutyric acid, and acetoacetic acid.

Kwashiorkor: deficiency disease related principally to protein lack and seen in severely malnourished children.

Lactic acid: 3-carbon acid produced in milk by bacterial fermentation of lactose; also produced during muscle contraction by anaerobic glycolysis.

Lactose: a disaccharide composed of glucose and galactose; milk sugar.

Lecithin: a phospholipid that contains choline and is found in many animal tissues especially nerve tissues, semen and egg yolk.

Limiting amino acid: that amino acid that is in shortest supply in relationship to synthetic needs and therefore limits synthesis.

Linoleic acid: a fatty acid considered essential and is found mostly in oils.

Linolenic acid: a fatty acid considered essential and is also found in vegetable oils.

Lipid: a term for fats including neutral fats, oils, fatty acids, phospholipids, cholesterol.

Lipogenesis: formation of fat.

Lipolysis: the splitting of fat for energy production.

Lipoprotein: a conjugated protein that incorporates lipids to facilitate transportation of the lipids in an aqueous medium.

Marasmus: extreme protein-calorie malnutrition marked by emaciation, especially severe in young children.

Metabolism: physical and chemical changes occurring within the organism; includes of biologic materials and breakdown of substances to yield energy.

Micelle: a microscopic particle of lipids and bile salts.

Mineral: any homogeneous inorganic material found in the earth's crust. In this test it refers to elements that are found in very small amounts in food and which are essential to the human body.

Monosaccharide: a single sugar not affected by hydrolysis; includes glucose, fructose, galactose.

Mucosa: membrane lining the gastrointestinal, respiratory, and genitourinary tracts.

Myelin: lipid-protein complex surrounding nerve fibers.

Myo: prefix meaning muscle.

Neuroleptic: an agent that produces symptoms resembling those of nervous system disorders.

Neuropathy: disease of the nervous system.

Neurotransmitter: a chemical substance that relays nerve impulses, thus enabling cells to communicate; dopamine, nerepinephrine, serotonin.

Niacin: a water-soluble B-complex vitamin required for cell respiration; antipellagra factor.

Nicotinic acid: niacin.

Nutrient: chemical substance in foods which nourishes, e.g. amino acid, carbohydrates, vitamin A.

Oleic acid: an 18-carbon fatty acid containing one double bond.

Oligosaccharide: a carbohydrate that yields more than 2 simple sugars upon hydrolysis.

Osteo: prefix meaning bone.

Osteomalacia: softening of the bones, chiefly in adults.

Osteoporosis: reduction of the quantity of bone, occurring principally in women after middle age.

Pantothenic acid: one of the B-complex vitamins.

Pectin: polysaccharide found in many fruits and having gelling properties.

Pellagra: a deficiency disease of the skin, gastrointestinal tract, and nervous system caused by lack of niacin.

Peristalsis: the rhythmic, wavelike movement produced by muscles of the small intestine to move food forward.

Periodontal: around a tooth.

Phospholipid: a fatlike compound that contains a phosphate and another group such as a nitrogen base in addition to glycerol and fatty acids, e.g. lecithin, cephalin.

Phytic acid: a phosphoric acid ester of inositol found in seeds; interferes with absorption of calcium magnesium, iron, zinc.

Plasma: fluid portion of the blood before clotting has taken place.

Poly: prefix meaning much or many.

Polyneuritis: inflammation of a number of nerves.

Polypeptide: a compound consisting of more than three amino acids; an intermediate state in protein digestion.

Polyunsaturated fatty acid: fatty acids containing two or more double bonds; linoleic, linolenic, and arachidonic acids.

Postsynaptic: occurring after the synapse is crossed (see synapse).

Pyridoxine: one of the forms of vitamin B-6.

Pyruvic acid: a 3-carbon ketoacid; an intermediate in glucose metabolism.

Regurgitation: the backward flow of food; casting up of undigested food.

Relapse: the return of a disease weeks or months after its apparent cessation.

Renal: pertaining to the kidney.

Riboflavin: heat-stable B-complex vitamin.

Rickets: a deficiency disease of the skeletal system caused by a lack of vitamin D or calcium or both, and often resulting in bone deformities.

Satiety: feeling of satisfaction following meals.

Saturated: a state in which a substance holds the most of another substance that it can.

Scurvy: a deficiency disease caused by lack of ascorbic acid and leading to swollen bleeding gums, hemorrhages of the skin and mucous membranes, and anemia.

Sucrose: cane or beet sugar; a disaccharide that yields glucose and fructose when hydrolysed.

15. Recipes

The following recipes were especially designed for therapeutic use and they are classified according to specific indications. They require minimal physical efforts and their ingredients are economical. Please note that "T" refers to tablespoon while "t" refers to teaspoon.

I. Recipes for patients with swallowing difficulties

Apple Farm Cottage Cheese

¾ C low fat cottage cheese
1 C unsweetened applesauce
1 T honey (when allowed on your individual prescription)
2 T nonfat skim milk powder

- Mix together.
- Serve as a main breakfast dish or a supplemental dessert

Creamed Chicken

2 C cooked chicken parts, shredded
¼ C diced green onions
½ C sliced fresh mushrooms
2 T soy sauce
¼ C chicken broth
1/8 t salt

1/8 t pepper
2 T cooking sherry
1 C plain yogurt

- Combine soy sauce, chicken broth, salt, pepper, and sherry in a skillet, heat to boil
- Add chicken, green onions, and mushrooms
- Simmer at low heat 20-25 minutes
- Transfer to serving platter
- Add yogurt while hot
- Let stand for a few minutes before serving.

Pureed Chicken Fantastique

1 C cooked shredded chicken
¾ C chicken broth
Dash salt/pepper
1 T minced onions
1/8 t chili powder
2 T plain yogurt

- Combine all ingredients in a blender
- Blend at medium speed until pureed
- Heat and serve

Egg Salad a l'Italian

4 hardboiled eggs
2 C finely chopped ripe tomatoes
1/8 t salt
1/8 t pepper

- Mix together
- May use as sandwich filling

Low Calorie Tuna Salad

1 C water packed tuna, drained
¼ C low calorie buttermilk salad dressing
¼ C dried celery

- Mix together
- Serve on a bed of lettuce

Soup-a-Meal

½ C of your favorite cream soup, undiluted
½ C pureed meat
2 T pureed vegetables

- Mix together
- Eat with a spoon

Meal on the Run

½ C mashed potatoes
½ C pureed meat
2 T gravy

- Mix together, eat with a spoon

II. High Protein High Calorie Recipes

Protein Powerhouse*

1 C fruited yogurt
½ banana
¼ C nonfat skim milk powder
¼ C milk

- Blend together, eat with a spoon

Applecheese*

½ C applesauce

½ C cottage cheese
2 T skim milk powder

- Mix with a fork
- Serve as a dessert

Powerhouse Deep Dish Dessert

2 C cooked yams
4 large ripe bananas, sliced
1 ½ C diced pineapples
½ C pineapple juice
½ C chopped nuts
1 C pina colada flavored yogurt

- Preheat oven 275 degrees
- Grease a 2-quart casserole dish
- Spread yams in 1 layer
- Spread banana slices in a 2nd layer
- Spread diced pineapple in a 3rd layer
- Pour ½ C pineapple juice
- Bake for 15 to 20 minutes
- Mix yogurt and nuts together and pour over above while hot

Astronaut's Milkshake*

¼ C walnut
½ C fruited yogurt
½ C milk
½ C ice cream
½ C applesauce
2 T skim milk powder

- Blend at a high speed, may eat with a spoon

III. Low-Calorie Recipes

Cottage Cheese Delight

2 C low fat cottage cheese
¼ C dried parsley
¼ C chopped green onions
1 large tomato, chopped
½ C diced celery
½ C diced carrots
1 t Worcestershire sauce

- Mix all ingredients
- Serve as a main dish or over a bed of lettuce as salad

Chicken au Beure

1 C shredded leftover chicken
1 t butter
½ C chicken broth
salt and pepper to taste

- Heat butter in a skillet
- Add chicken and heat over medium heat while stirring constantly
- Add chicken broth, cover and simmer for 10 minutes

Garlic Snapper

3 cloves of fresh garlic, minced
¼ C lemon juice
1/8 t salt
pepper to taste
1 lb fresh red snapper filet

- Preheat oven to 250 degrees
- Combine garlic, lemon juice, salt, and pepper
- Place fish filet in bottom of baking dish
- Pour lemon juice mixture, cover with foil
- Bake for 15 to 20 minutes

Tasty Liver

2 C sliced onions
1 t Worcestershire sauce
1/8 C wine vinegar
1 T lemon juice
2 t cumin powder
salt and pepper to taste
1 lb sliced beef liver or chicken livers

- Combine vinegar, lemon juice, Worcestershire sauce, salt, pepper, and cumin in a skillet
- Heat to a boil
- Add onions, cook at medium heat until transparent
- Push onions to one side of the skillet and add liver
- Brown at medium heat (both sides of each slice)
- Turn heat to low, simmer for 10 minutes

Hearty Vegetable Beef Dinner

1 lb lean round steak cut up in cubes
1 T olive oil
½ C diced onions
2 cloves of garlic, minced
2 C hot water
½ c fresh green beans
½ C chopped celery
½ C chopped carrots
½ C sliced zucchini
½ C sliced mushrooms
1 T flour dissolved in ¼ C cool water

- Brown beef in oil in a large pot
- Add onions and garlic
- Add water and bring to a boil
- Simmer at low heat for 1 hour
- Add celery, carrots and green beans
- Simmer for 30 minutes
- Add zucchini and mushrooms
- Simmer for 20 minutes
- Add flour and water mixture and stir constantly at medium heat until thickened

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Chicken Salad

6 oz cooked chicken, shredded
3 T reduced calorie buttermilk salad dressing
½ C minced celery
1 t parsley flakes
2 T finely chopped green onions

- Mix all ingredients together; use as a sandwich filling or serve on a bed of lettuce

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Low Calorie Vegetable Soup

1 ½ C water
1 ½ C tomato juice
1 T Worcestershire sauce

¼ C chopped green onions
½ C dried onion
1 C chopped celery
1 C chopped cabbage
1 C chopped carrots
1 C sliced zucchini
1 C sliced mushrooms

- Bring water, tomato juice, and Worcestershire sauce to boil
- Add onions, celery, carrots, and cabbage
- Bring to a boil; cook at medium heat for 20 minutes
- add remaining ingredients; cook for an additional 15 minutes

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Onion Dip

1 C plain yogurt
1 T dried Lipton onion soup mix

- Mix well
- Refrigerate for at least 1 hour before serving
(Recommended for fresh vegetable, celery and carrot sticks)

Marinated Cabbage Salad

1 C shredded cabbage
½ C diced celery

½ C diced carrots
2 T chopped green onions
1/8 C vinegar
1/8 C soy sauce
1/8 C olive oil
1 clove minced garlic
1 T lemon juice

- Mix the first 4 ingredients; set aside
- Blend remaining ingredients together
- Mix both mixtures and refrigerate overnight in a tight covered container

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Vegetable Gelatin Salad

Knox unflavored gelatin
Diced celery
Diced carrots
Salt to taste

- Mix Knox unflavored gelatin as indicated on the package
- Add celery, carrots and salt
- Refrigerate to gel

(Use as a complimentary salad or as in-between meals snack)

Deep Dish Apple Delight

2 C cooking apples cut up in bite sizes
2 T lemon juice
2 T apple juice
1 t flour
1 C apple flavored yogurt
1 T brown sugar

- Preheat oven at 350 degrees
- Lightly spray the bottom of a 2 quart casserole dish with “no stick” cooking spray
- Place apples in casserole dish
- Add lemon juice, apple juice and flour
- Bake for 30 to 35 minutes; cool for 10 minutes
- Mix together yogurt, brown sugar and flour
- Pour yogurt mixture over baked apples; serve warm

Yogurt au Chocolat

8 oz nonfat plain yogurt
1 T low calorie hot cocoa mix
1 pkg of Nutrasweet or ¼ to ½ pkg of Sweet and Low

- Mix all ingredients together
- Chill to set

(A great substitution for chocolate ice cream)

IV. Low Calorie Beverages

The following beverages can be used instead of coffee or tea as excellent appetite suppressant following a meal.

Summer Cooler

Crushed ice
Ripe fruit

- Blend at high speed; drink with a straw

Tomato Warm-up

½ C hot water
½ C tomato juice or V8 juice

- Stir together; serve in a mug

Gil's Hot Apple Juice

½ C hot water
½ C apple juice

- Stir together; bring to a boil; serve in a mug

All Purpose Chicken Broth

Bring to a boil 1 whole chicken in 2 quarts of water
Add 1 peeled whole onion

- Simmer at low heat for 1 hour
- Let it cool
- Remove chicken and use for other recipes
- Discard onion
- Place chicken broth in refrigerator over night
- The next day, skim off all fat
- Use this fat-free broth as a beverage with or in between meals