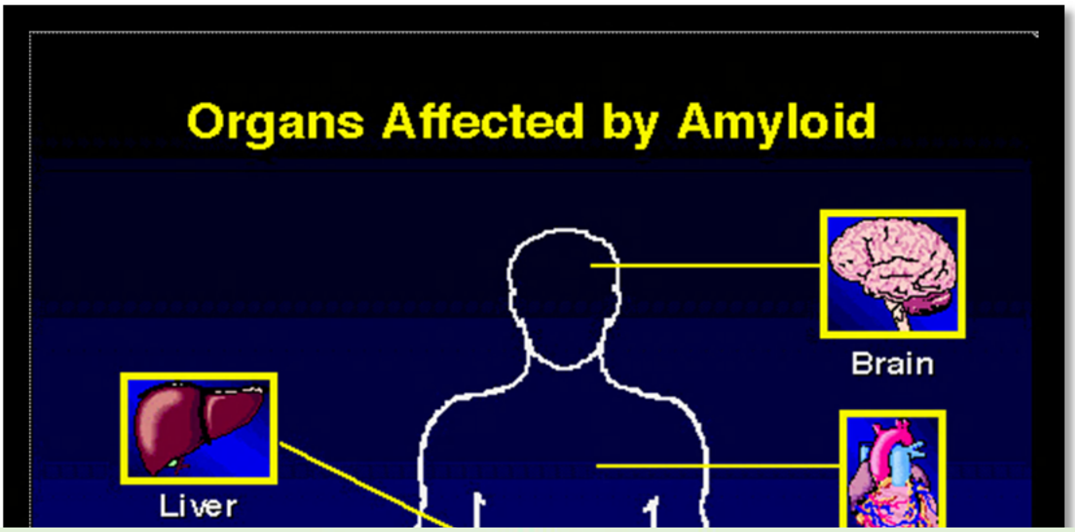
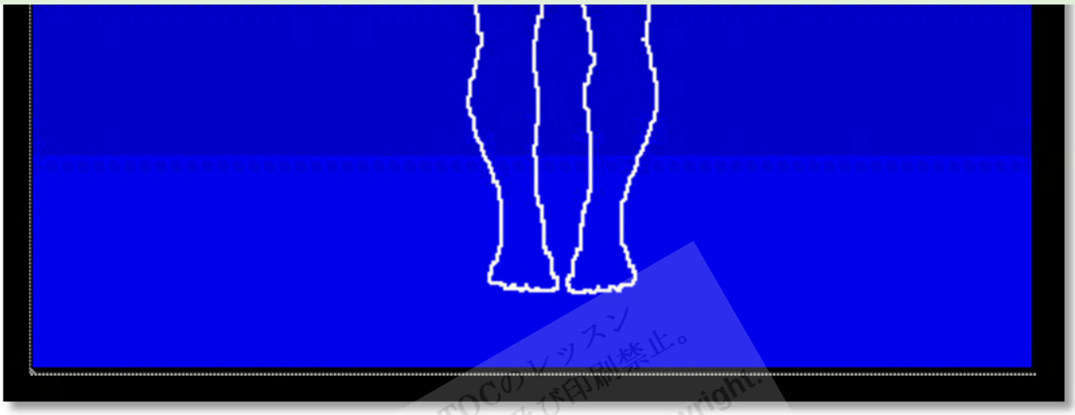
Amyloidosis





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What is amyloidosis?

Amyloidosis is a group of diseases that result from the abnormal deposition of a particular protein, called amyloid, in various tissues of the body. Amyloid protein can be deposited in a localized area and may not be harmful or only affect a single tissue of the body. This form of amyloidosis is called localized amyloidosis. Amyloidosis that affects tissues throughout the body is referred to as systemic amyloidosis. Systemic amyloidosis can cause serious changes in virtually any organ of the body.

Amyloidosis can occur as its own entity or "secondarily" as a result of another illness, including multiple myeloma, chronic infections (such astuberculosis or osteomyelitis), or chronic inflammatory diseases (such asrheumatoid arthritis and ankylosing spondylitis). Amyloidosis can also be localized to a specific body area from aging. This localized form of amyloidosis does not have systemic implications for the rest of the body. The protein that deposits in the brain of patients with Alzheimer's disease is a form of amyloid.

Systemic amyloidosis has been classified into three major types that are very different from

spontaneously overproduces a particular protein portion of an antibody called the light chain. (This is why it is coded as AL.) The deposits in the tissues of people with primary amyloidosis are AL proteins. Primary amyloidosis can occur with a bone marrow cancer of plasma cells called multiple myeloma. Primary amyloid is not associated with any other diseases but is a disease entity of its own, conventionally requiring chemotherapy treatment. Researchers at the Mayo Clinic in Rochester, Minnesota, and Boston University in Boston, Massachusetts, have demonstrated benefits from stem-cell transplantation, harvesting patients' own stem cells to treat primary amyloidosis.

Secondary amyloidosis

When amyloidosis occurs "secondarily" as a result of another illness, such as multiple myeloma, chronic infections (for example, tuberculosis or osteomyelitis), or chronic inflammatory diseases (for example, rheumatoid arthritis and ankylosing spondylitis), the condition is referred to as secondary amyloidosis or AA. The amyloid tissue deposits in secondary

amyloidosis are AA proteins. The treatment of patients' secondary amyloidosis is directed at treating the underlying illness in that particular patient.

Familial amyloidosis

Familial amyloidosis, or ATTR, is a rare form of inherited amyloidosis. The amyloid deposits in familial amyloidosis are composed of the protein transthyretin, or TTR, which is made in the liver. Familial amyloidosis is an inherited autosomal dominant in genetics terminology. This means that for the offspring of a person with the condition, there is a 50% chance of inheriting it.

Beta-2 microglobulin amyloidosis

Beta-2 microglobulin amyloidosis occurs when amyloid deposits develop in patients on dialysis with longstanding kidney failure. The amyloid deposits are composed of beta-2 microglobulin protein and are often found around joints.

the kidney leads to "nephrotic syndrome," which is characterized by severe loss of protein in the urine and swelling of the extremities.

How is amyloidosis diagnosed?

The diagnosis of amyloidosis is made by detecting the characteristic amyloid protein in a biopsy specimen of involved tissue (such as mouth, rectum, fat, kidney, heart, or liver). A needle aspiration biopsy of fat just under the skin of the belly (fat pad aspiration), originally developed at Boston University, offers a simple and less invasive method to diagnose systemic amyloidosis. Pathologists can see the protein in the biopsy specimen when it is coated with a special dye, called Congo red stain.

How is amyloidosis treated?

Initial treatment of amyloidosis involves correcting organ failure and treating any underlying illness (such as myeloma, infection, or inflammation). The disease is frequently discovered after significant organ damage has already occurred. Therefore, stabilization of organ function is an

initial target of treatment. The most frequent cause of death in systemic amyloidosis is kidney failure.

Sephardic Jews and Turks inherit a genetic disease called Familial Mediterranean Fever, which is associated with amyloidosis and characterized by episodes of "attacks" of fever, joint, and abdominal pains. These attacks can be prevented with the medication colchicine. Armenians and Ashkenazi Jews also have a higher incidence of Familial Mediterranean Fever attacks but do not suffer amyloid deposition disease. Other reports of amyloidosis in families are extremely rare.

Researchers are currently enrolling patients with primary amyloidosis in clinical trials using a cancer chemotherapy medication (melphalan[Alkeran]), in conjunction with bone-marrow stem-cells transplantation. The results have been promising, and this combination treatment is offered to eradicate the amyloidosis in selected patients, provided that the underlying medical condition of the patient is adequate. These aggressive treatment options with stem-cell transplantation and high doses of chemotherapy are a true breakthrough in the treatment of these patients.

Familial amyloidosis can now be cured with liver transplantation. This option requires an accurate diagnosis of the specific protein that causes the disease.

Amyloidosis at a Glance

- Amyloidosis is a disorder resulting from abnormal protein (amyloid) deposits in body tissues.
- Amyloidosis can occur as an isolated disease or as a result of another illness.
- Symptoms in patients with amyloidosis result from abnormal functioning of the particular organs involved.
- Diagnosis of amyloidosis is made with a biopsy of involved tissue.
- Treatment options for amyloidosis depends on the type of amyloidosis and involves correcting organ failure and treating any underlying conditions.

Reference:

http://www.medicinenet.com