

Patient information sheet for domino liver transplantation – Receiving a liver from a patient with hereditary amyloidosis.

What is a domino liver transplant?

A domino liver transplant involves re-using a liver removed from a living patient who has a rare hereditary disease that can be treated by means of a liver transplant from a healthy donor. These hereditary diseases are also known as metabolic liver disorders.

Liver transplants are usually performed in patients with liver failure due to cirrhosis etc, but liver transplantation is also a treatment for patients with certain very rare genetic diseases in which their livers are functioning normally but are genetically programmed to produce an abnormal blood protein that can (but not always) cause disease in other parts of the body.

What is amyloidosis?

Amyloidosis is a disorder in which an abnormal blood protein builds up in various organs throughout the body, which causes progressive organ damage and ultimately organ failure. The abnormal protein deposit is called amyloid, and the disease is called amyloidosis. Hereditary amyloidosis is extremely rare, and is caused by the liver producing a genetically abnormal blood protein from birth. However, symptoms of hereditary amyloidosis usually do not begin until middle age or later, and do not ever occur in some patients who have inherited the abnormal hereditary amyloidosis gene. Liver transplantation is a treatment for hereditary amyloidosis, since the normal donor livers given to these patients does not produce an abnormal amyloid-forming protein.

The liver itself in hereditary amyloidosis is usually healthy in the sense that it functions normally. However, when such a liver is re-used and transplanted into another person, it will continue to produce an amyloid forming protein into the bloodstream, which may therefore build-up in vital organs and lead to amyloidosis after many years in the 'domino' liver transplant recipient.

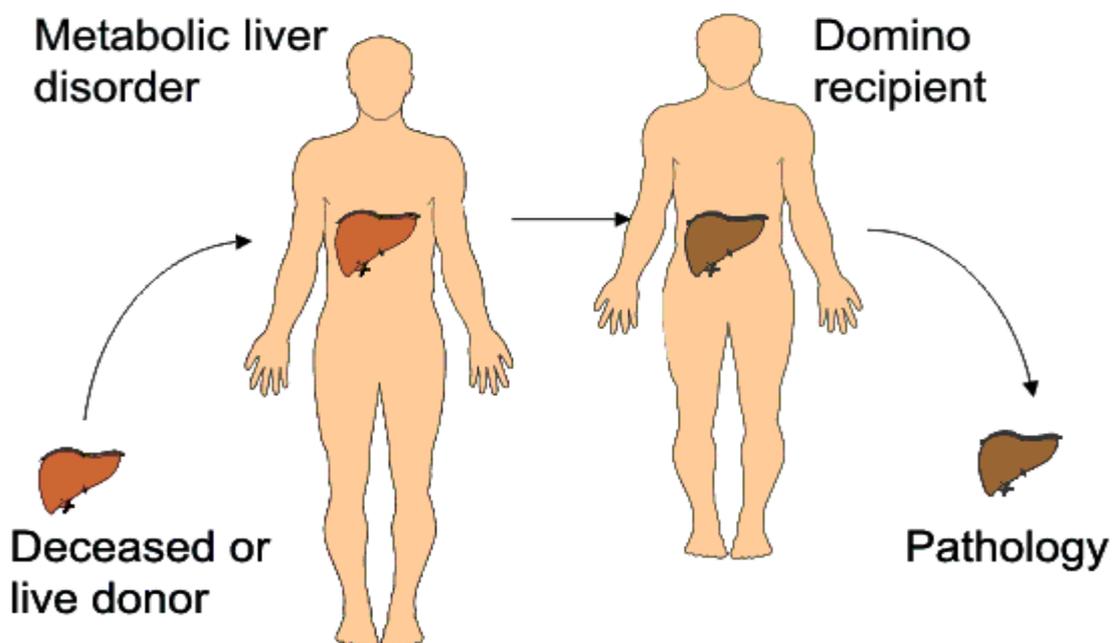
What symptoms did the donor of your liver have?

There are 2 types of hereditary amyloidosis for which liver transplants are performed.

The first is familial amyloid polyneuropathy (FAP). FAP is caused by an inherited mutation in the gene for a blood protein transthyretin. This protein is made in the liver and genetically abnormal transthyretin builds up as amyloid in the nerves and heart of patients with FAP. This typically causes wasting, muscle weakness, abnormal sensation (loss of feeling or pain), loss of appetite, and bladder and bowel problems, along with breathlessness, fluid retention and lethargy due to heart damage. Symptoms typically start after the age of 30. The livers of these patients is the source of the abnormal transthyretin protein, but continue to function normally and have only traces of amyloid within them.

The second type is hereditary fibrinogen A-alpha chain amyloidosis. This blood protein is also made by the liver, and genetically abnormal forms can build up as amyloid mainly in the kidneys. Symptoms typically start after the age of 30, and often not before age 60. This type of hereditary amyloidosis results in kidney failure. It can be treated by combined kidney and liver transplantation – the kidney transplant to restore normal kidney function, and the liver transplant to remove the source of the genetically abnormal amyloid-forming protein. The livers in patients with hereditary fibrinogen A-alpha chain amyloidosis usually contain only traces of amyloid and function normally in the general sense.

Domino liver transplantation – the Concept



What are the risks of having a domino liver transplant from a patient with hereditary amyloidosis?

There is a risk that you will eventually develop amyloidosis yourself. The 'domino' liver will continue to produce an abnormal protein which may (or may never) begin to build-up in your organs as amyloid. Several hundred of these domino transplants have been performed around the world during the past 15 years, mostly using livers from patients with familial amyloid polyneuropathy. To date, only a few of these domino liver recipients have developed amyloidosis. Only about five domino transplants have used livers from patients with hereditary fibrinogen A-alpha chain amyloidosis, and so far there has been no documented case of amyloidosis developing any of these patients.

At present, there is no drug treatment for hereditary amyloidosis, but we are optimistic that new treatments will become available within 5-10 years.

Are there any benefits in receiving a domino liver from a patient with amyloidosis?

Regrettably, about one in five patients on the waiting list for a liver transplant dies before a donor liver becomes available; use of domino liver transplants plainly increases the number of available donor livers. Domino donor livers are removed from living people, and are likely to be in excellent condition.

Further information and possibility of follow up tests for amyloidosis

The National Amyloidosis Centre is based at the Royal Free Hospital, and its team of doctors are happy to discuss any further questions or concerns you may have before or after undergoing a domino liver transplant. The National Amyloidosis Centre is pleased to offer regular check-ups regarding the possibility of developing amyloidosis as well as information on new treatments as they become available.

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