

Question and Answer Session

Summary:

- Foot care is important in neuropathy because of the lack of feeling in the foot. Examine feet each night for cuts, cracks, injuries.
- Everyone should have regular internist. How to find one? Guidelines: try him/her out. Will he be willing to work with our specialists at the amyloid centers, who are always willing to work with the local doctors? Patient needs to participate by helping educate doctors. Doctor should be open, communicative and non-threatened by asking for help.
- There are no trials at present for post-transplant GI problems in amyloid.
- Eligibility for drug trials for amyloid. People with diabetes are not eligible. Must have pure disease, not other conditions that mimic amyloid symptoms. Future trails will focus on heart, but now just neuropathy.
- Manage pain of neuropathy? Amotryptalene. 25 mg at bedtime. Can be increased.

Neurontin. Next step up.

Steer clear of narcotics if possible

Warm baths, massages.

Can't sleep because of pain? Ketamine - cream based – anesthetic agent. Also capsation (sp?) which is an arthritis drug. Boston not a fan of trans electrical nerve stimulator.

- Atrial fibrillation. Use of calcium or beta blockers to control rate. Dr. Hanna uses beta blockers only occasionally to control rate. Tries to avoid. Risk of stroke from forming clots so put people on blood thinners. Pradoxal or cumadin.
- Arrhythmias. Amioderon keeps heart in rhythm but has side effects.
- Can you have amyloid in pancreas – person has diabetes. Yes, but it is a different kind and localized.
- Familial types that benefit from transplant: clearly TTR, Fibrinogen. Questionable in Apolipoprotein A1.
- Diabetic eligible for transplant? Impairment of other organs must be evaluated before transplant.
- Dialysis related amyloidosis – long term dialysis (10-20 years) can be a risk factor for amyloidosis.

- When to consider a heart, but not a liver, transplant. Jury still out on TTR. It might be reasonable but must be judged on individual cases. APOA1 will do well with just heart. African Americans with Isolucene122 may do well as it tends to be isolated to the heart.
- Boston U. philosophy toward transplants. Each case is individual. More aggressive with young person. Good to have both organs from same donor.
- Transplant list for two organs. Sometimes impossible to get. Donor contributions flat. Organ allocation system now allows filing for exception if person has amyloid. The MELD score can jump as the amyloid liver can be used for another patient.
- Age factor in domino transplant? If eligible to get liver then eligible to give liver.
- Dialysis? Saves lives, has been improved over the years. Problems: increased risk of infection
Atherosclerotic, time factor.
- Effect of amyloidosis on routine medical procedures. Dental is okay. Those with neuropathy and autonomic problems may have difficulty tolerating sedation. Also is a challenge to cardiovascular system.
- Dr. Solomon's antibody? Compounds for TTR are some time away.
- How does interfering RNA affect amyloidosis? Body will make less protein.
- Mass spectrometry. Machine analyses size of small rare particles. Powerful tool. Quick and definitive to identify amyloid protein.
- Different donors for heart and liver. Prefer same donor. But if done at different times, obviously need two different donors.
- Proposed amyloid treatments do not treat eye. Need different delivery system. There is a "blood-brain barrier" between eye (nervous system) and the rest of the body.
- Inflammation and TTR. Inflammation level goes down in TTR
- Onset of amyloid. Probably not at birth. There are changes in the body with age. Perhaps the metabolism can't handle it anymore.
- Nutritional status is one of the best indicators of successful outcome after liver transplant.
- Local physicians. Patient is his own best advocate. Ask if willing to collaborate or find a new doctor.
- Immunizations before liver transplant. Yes. But it will be done. Don't worry about it. Always get flu shot.
- Different mutations in same family. Not likely.
- Chelation .Not effective in amyloid.

- Elevated liver function. Unusual for amyloid to affect liver. Probably related to medications.
- Heart biopsy before liver transplant. In the absence of evidence of heart involvement, not necessary.
- Clinical trials listing. www.clinicaltrials.gov also www.amyloidosisupport.com
- Stabilize children with gene when drug is available? Not before symptoms evidence. They cannot give consent. Not ethical.
- Diflunisal trial. Alcohol? Excludes heavy drinkers.
- Anti-rejection medicine. Needed after transplant because your body has an immune response to a foreign object.
- Orthostatic hypotension? - Indicated by a 20mm drop from lying down to standing up.
 - Get up slowly
 - Elastic stockings
 - Meds – midodrene – 5mg, three times daily. Florinef – not as good.
 - A little more salt sometimes helps but is difficult to balance amount.
- Ankle swelling is common after transplant. Might check for kidney problems with blood test /urine test.
- Amyloid enhancing factors. Small fibrils that become big fibrils. Not in TTR.
 - -Environmental factors. Lead a healthy lifestyle. Reduce stress, watch nutrition, get exercise. Get yourself in the best shape possible in all areas including emotional and psychological.