

## Medical Treatments which have been useful in Alström syndrome

General	Maintain good mobility and exercise, eg swimming, gym, walking (with buddy or dog).
Nutrition	70 gram carbohydrate diet with modest intake of saturated fat.
Diabetes	Metformin – where cardiac and renal function are carefully followed Glitazones – if no heart problem Younger patients rarely require insulin, but some older patients may respond to high doses.
High blood fats	Mildly raised serum triglycerides may respond to fibrates and statins combined but once serum triglycerides increase above 10 mmol/l nicotinic acid as Niaspan 3g nocte may be required in adults with or without fibrate in the morning. Aspirin 75mg half an hour before the nicotinic acid will minimize flushing.
Heart	From the diagnosis of cardiomyopathy long-term ACE inhibitors are indicated. Hypertension may be treated with the usual combination of drugs.

## Surgical Treatments possible in severe forms of complications

Heart	Severe cardiomyopathy has been improved with dual chamber pacemaker, both to override subtle dysrhythmias and improve coordination of the cardiac cycle. The place of cardiac transplantation is under review
Bladder	The incoordination between bladder and urine outflow (destrusor-urethral dyssnergia) can be helped by intermittent self catheterisation of the bladder and even more rarely may require ileal diversion.
Kidney	Rarely fibrosis in the kidneys may lead to renal failure. The place of renal transplantation in this circumstances is being explored.
Liver	Fatty liver can rarely progress to significant fibrosis and portal hypertension in Alström syndrome. A very few subjects have required upper gastrointestinal endoscopy with injection of varices, and/or the TIPS procedure to relieve variceal pressure.