

PRIMARY SCLEROSING CHOLANGITIS AND LIVER TRANSPLANTATION

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Primary Sclerosing Cholangitis

- Definition
 - Chronic cholestatic liver disease
 - Destruction of biliary tract
 - Associated with inflammatory bowel disease
 - Usually presents 3rd –4th decade of life
 - Male predominance

Clinical Manifestations

- Pruritis
- Jaundice
- Abdominal pain
- Fatigue

Progression to cirrhosis

As disease progresses symptoms are secondary to portal hypertension

- Development of ascites
- Variceal bleeding
- Hepatic encephalopathy

Cholangiocarcinoma

- Most feared complication of PSC
- Incidence of 1.5% per year after diagnosis is made
- Survival of less than 1 year
- Liver transplant contraindicated
 - Rapid recurrence
 - High mortality

Diagnosis

- Endoscopic findings
- Liver biopsy

Treatment

- Medical management
 - Variety of medications
 - Endoscopic management
- Surgical Management
 - Liver resection
 - Biliary reconstruction

Referral for Liver Transplant



Indications

Failure of medical
management

Poor quality of life

Debilitating fatigue

Intractable ascites

Severe muscle wasting

Work-up for Liver Transplant

- Psycho-social evaluation
- Surgical evaluation
- Medical evaluation
- Endoscopy
- Cardiac work-up
- Radiological work-up

Listing



- Case presented at listing meeting
- MELD score
PT/INR, creatinine, bilirubin,
- Wait for appropriate cadaveric liver
?months to years

Life after transplant

- Improvement of quality of life
- Concerns

Rejection

Some form of immunosuppression for a lifetime

Susceptibility to infection

Prophylactic antibiotics

Recurrence of primary disease

Live Donor Liver Transplant



Advantages of Live Donor Transplant

- Alternative option
- Surgery is scheduled
- Known quality of organ
- Lower cold ischemia time
- Surgery is not a race against time

Workup for Live Donation

- Similar to workup for transplant
- Evaluation by surgeon, hepatologist
- CT scans, U/S, cardiac w/u,....
- Liver biopsy

Surgery

- 2 teams: Donor and Recipient
- For adult to adult live donor liver transplant right lobe is used
- Right lobe is usually 60-70% of liver by weight

Live Donor Program

- 122 adult to adult live donor liver transplants
- 21% pts transplanted for PSC
- Patient survival, and graft survival is 92%