ABOUT POLYCYSTIC LIVER DISEASE (PLD)

PLD Facts
Polycystic liver disease (PLD) is a genetic disease in which cysts develop in the liver.

- The most common occurrence of PLD is associated with autosomal dominant polycystic kidney disease (ADPKD), where patients have cysts in both the liver and kidneys. However, there is a form of PLD where patients only have cysts in the liver. Both forms are hereditary.
- The majority (80 percent or more) of ADPKD patients also have PLD. It occurs more often in women than men.
- Most patients with both ADPKD and PLD never have symptoms from their liver condition. About five to 10 percent of patients will have more significant symptoms that may impact their health.
- PLD cysts may cause pain, but typically do not affect liver function. Severe PLD is uncommon.
- Most cysts do not need to be treated. If cysts do affect liver function or become too painful to function, surgery may be needed to drain or remove the cysts.

Symptoms of PLD
- Abdominal pain/bloating
- Early satiety (feeling full early during a meal) and heartburn
- Bile duct dilation, bile duct cancer and congenital hepatic fibrosis are rare, but can occur
- A combination of fever, abdominal pain/bloating and feeling unwell could indicate cyst infection and medical help should be sought.

Complications Related to PLD
- Gastric reflux
- Cyst infection
- Cyst hemorrhage
- Cyst rupture

Options to Treat PLD
- Surgical draining or removal of cysts
- Partial liver resection (removal of part of the liver with cysts)
- Liver transplant in severe cases

Health Management
- Maintain a healthy diet
- Avoid fatty foods
- Avoid caffeine
- Avoid soy-based diet (soy contains estrogen, which has been implicated in liver cyst growth)
PLD Frequently Asked Questions

Q. How is PLD diagnosed?
A. Your doctor will physically examine you and check your liver and kidney function. Tests such as an ultrasound, CT scan or MRI can be used to check the presence, size and position of the cysts in the liver. Discuss your liver imaging with your doctor especially if you are having abdominal symptoms, are female, had prior pregnancies or have been on estrogen therapy.

Q. Do I have PLD as a result of my PKD?
A. Most of the time, this is the case. There is another hereditary condition, autosomal dominant PLD, where there are only liver cysts and few or no kidney cysts and normal kidney function.

Q. What type of doctor do I see for PLD?
A. If you are having trouble with your liver, your doctor may refer you to a liver specialist called a hepatologist, to a nephrologist or to a specialty center with expertise in PLD. You will also continue seeing your nephrologist and, ideally, the two doctors would work together to create a treatment plan.

Q. Will I need a liver transplant?
A. A liver transplant is only needed in rare cases of massive PLD, or when it is causing extreme pain, a significant decrease in liver function, severe physical disability, fatigue or clinically-advanced malnutrition.

Q. How do I manage pain associated with PLD?
A. There are no specific medical treatments that are used in PLD. Prescription pain medicine may be required and in those situations, patients should seek evaluation with a nephrologist with experience in PKD and PLD.

Q. Are there medications that I should avoid because I have PLD?
A. Estrogen therapy should be avoided because it is implicated in liver cyst growth. Always consult with your doctor before starting a new medication.

Q. How will PLD affect my health?
A. Most patients never have symptoms from their PLD. But about five to ten percent of patients will have more significant symptoms that may impact their health. Typically these symptoms relate to liver enlargement causing abdominal bloating, pain and distention, heartburn and early satiety.

Q. Is there a special diet for people with PLD?
A. PLD patients should avoid fatty foods and too much caffeine (more than one 8 oz coffee or equivalent per day). In addition, a soy-based diet should be avoided. Soy contains estrogen, which has been implicated in liver cyst growth.

Q. Is PLD hereditary?
A. PLD is a hereditary disease and is most commonly seen in association with ADPKD. However, there is a form of PLD where patients only have cysts in the liver.

Q. What is liver resection?
A. The removal of part of the liver with cysts.

Q. Is there living donation for livers?
A. Yes, organ donors can give a portion of their liver to be transplanted into a recipient.

For more information, visit pkdcure.org.