# Key Takeaways for Clinicians from the KDIGO 2025 Clinical Practice Guideline for the Evaluation, Management, and Treatment of ADPKD: Polycystic liver disease

## 1 Polycystic liver disease (PLD)

PLD is the most common extrarenal manifestation of ADPKD. It is a hereditary disease characterized by the presence of >10 fluid-filled cysts scattered throughout the liver (Figure 1). PLD most often causes no symptoms and does not impact the synthetic or secretory capacity of the liver. In some cases, symptoms may develop due to mass effects causing abdominal fullness, distension and mechanical back pain, or compression of other organs such as lungs and stomach, or of veins such as the hepatic or portal veins or inferior vena cava, or of bile ducts. Symptoms may also occur with cyst hemorrhages and infections.

## 2 Abdominal imaging

People with ADPKD should have abdominal imaging, using ultrasound, CT, or MRI, to evaluate both liver and kidney phenotype. When liver cysts are found, patients should be advised of the likely outcomes and possible symptoms.

## 3 Women with PLD

PLD develops earlier and is more severe in women than in men. Women with PLD, should be counselled to minimize or avoid sex hormone therapy, as appropriate depending on the extent of the liver disease. Observational studies have shown that exposure to estrogen-containing oral contraceptives is associated with a 15.5% greater liver volume for each decade of use. The growth of polycystic livers decreases after menopause, but increases again if estrogen replacement therapy is initiated.

## 4 Treatment of PLD

Most patients with PLD have no symptoms and require no treatment; however, people with PLD who experience cyst-related symptoms negatively impacting their quality of life or who have severe disease likely to develop symptoms should receive treatment. The choice of treatment in people with symptomatic PLD should be based on symptoms, liver cyst characteristics, total liver volume, and treatment availability. Treatment may involve interventional radiology, surgery, or liver transplantation and should be done at centers of expertise if possible.

## 5 Somatostatin analogues

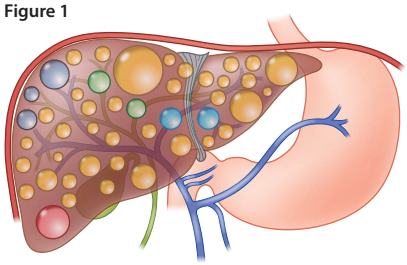
Long-acting somatostatin analogues should be given to people with ADPKD and markedly enlarged polycystic liver with volume-related symptoms to complement other therapies or when other therapies are not available. Long-acting somatostatin analogs are usually well tolerated but some adverse effects are possible (e.g., gallstones, bradycardia). Premenopausal women, who experience a faster liver growth than postmenopausal women, have a better response. Liver volume and disease-specific symptom questionnaires, such as PLD-Q and POLCA, may serve as measures to assess treatment outcomes.

## 6 Liver cyst infection

Liver cyst infections should be suspected in the presence of a triad of fever, localized abdominal pain and marked elevation of C-reactive protein or leukocytosis, supported by imaging consistent with infection, sometimes requiring 18FDG-PET-CT scanning, and confirmed by diagnostic features in at least 2 categories, such as clinical factors and microbiology. Treatment should be initiated promptly with a 3rd-generation intravenous cephalosporin with or without a fluoroquinolone; cyst drainage is required in severe (i.e., sepsis, immunosuppression) or refractory cases. Antibiotics (intravenous or oral) should be continued for ≥4 weeks (Figures 2 & 3).

ADPKD, autosomal dominant polycystic kidney disease; CT, computed tomography; 18FDG-PET-CT, 18F-fluorodeoxyglucose integrated with positron emission tomography/computed tomography; MRI, magnetic resonance imaging; PLD-Q, Polycystic Liver Disease Questionnaire; POLCA, Polycystic Liver Disease Complaint-specific Assessment

Figure 1



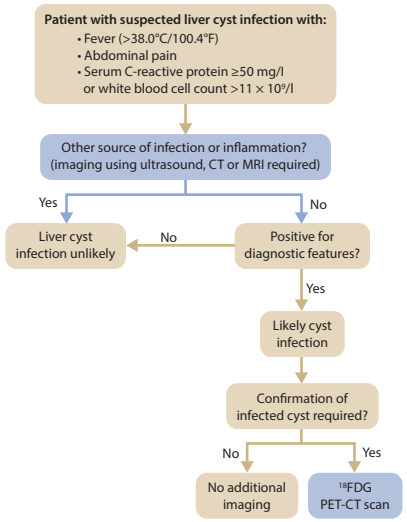
**Pressure against diaphragm and lungs**
1. Shortness of breath
2. Fatigue

**Pressure against the stomach**
1. Lack of appetite or early satiety
2. Acid reflux
3. Nausea and vomiting
4. Involuntary weight loss

**Cyst complications**
1. Intracystic
   a. Recurrent cyst infection
   b. Recurrent cyst hemorrhage
2. Extracystic
   a. Jaundice
   b. Hepatic venous outflow obstruction and portal hypertension

**Overall liver size**
1. Abdominal fullness
2. Limited mobility
3. Fatigue
4. Anxiety
5. Concern or dissatisfaction with abdomen size
6. Problems with intercourse
7. Mechanical back pain

Figure 2




**Diagnostic features**
**Diagnostic features considered positive in the presence of at least two items from at least 2 categories:**

**Clinical factors**
1. Pain presenting as acute pain or tenderness in liver area
2. History of cyst infection
3. Recent instrumentation of biliary tract
4. Immune compromised patient (including patients on dialysis)

**Microbiology**
5. Positive blood culture
6. Positive cyst fluid culture

**Imaging**
7. Imaging showing changes before and after onset of symptoms (ultrasound, CT or MRI)
8. Intracystic gas (ultrasound, CT or MRI)
9. Pericystic inflammation (CT or MRI)
10. Fluid-fluid levels in a cyst (MRI)
11. Thickened cyst wall (CT or MRI)
12. Contrast enhancement in the lining of cyst walls (CT or MRI)
13. Diffusion weighted imaging showing increased cyst density compared to normal cysts
14. Single-photon emission CT with Ga-67 abnormal uptake by a cyst
15. $^{111}$indium-white blood cell scan showing accumulation in a cyst

**Treatment**
16. Clinical response to antibiotic treatment

Figure 3