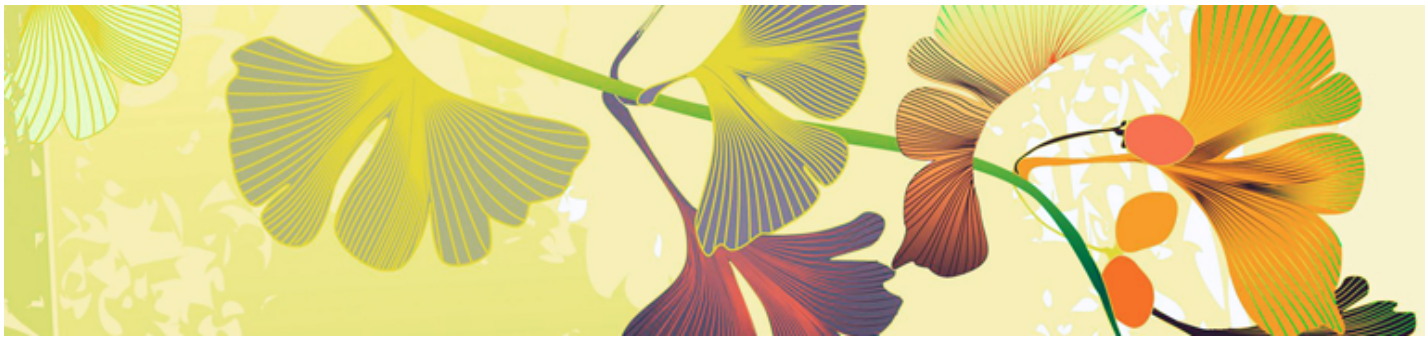

Polycystic Kidney Disease 多囊腎(英文)

What is Polycystic Kidney Disease?

Polycystic kidney is characterized by the growth of cysts on the kidneys. Most patients belong to autosomal dominant polycystic kidney disease. The cyst formation mainly involves kidney, followed by liver and pancreas. About 85% of cases are inherited from their parents. Their children can have about 50% of the incidence to get this disease. Once the disease is diagnosed, their children and siblings should receive further evaluation as well. Besides, 15% of all cases are caused by genetic mutation.

Clinical features

This disease has developed since fetus. However, most patients don’ t have any clinical features at young age. Presenting symptoms mainly include flank pain and/or abdominal pain. Besides, it also causes hematuria, polyuria, headache, hypertension, and abdominal discomforts.

Sudden onset of flank pain may be caused by cystic hemorrhage, stones, obstruction, or infection. These signs approximately appear at age of 40. With the age increases, symptoms get worse. Fifty percent of these patients will progress to renal failure after 50 years old. At present, patients with polycystic kidney disease account for 10% of hemodialysis patients.

Other extra-renal problems include enlargement of liver, intracranial aneurysm, colonic diverticulosis, and mitral valve prolapse.

Diagnosis

1. The diagnosis is confirmed if there is evidence of multiple renal cysts plus family history of polycystic kidney disease. Renal ultrasound is the best tool for diagnosis and follow-up. Patients are suggested to receive renal ultrasound once a year. The indications for CT scan

include cystic hemorrhage, abscess, renal stones, and suspicion of cancer.
2. Genetic study including sequence analysis and deletion/duplication analysis for PKD1 and PKD2 has been developed.

Treatment and Prevention

1. Diet: If patient has hypertension, it is necessary to maintain on low sodium diet. These patients should limit protein intake the same as patients with chronic renal failure stage 3-5.
2. Avoid violent exercise.
3. Avoid kidney infection: Avoid urethral catheter if possible. It is necessary to see nephrologists once you have flank pain, fever, and hematuira. Prolonged duration of antibiotic treatment (4-6 weeks) is recommended.
4. Renal stone: drink enough water every day to maintain urine output at least 2 liters per day. Consider surgery if you still can’ t excrete the stone.
5. Control blood pressure below 130/85 mmHg.
6. Consider excision of renal cysts if recurrent abdominal pain is caused by polycystic kidney disease.

Prognosis

The earlier the cysts appear, the worse the prognosis will be. In general, the disease slowly progresses. Although more than half patients over 50 years old may need to receive dialysis, there are patients with normal renal function until 80 years old. The risk factors of renal failure include male, larger kidney size, hypertension, and multigravida.

Some of the best ways to avoid renal failure is to prevent infection, control blood pressure, receive regular follow-up, and cooperate with doctors.