POLYCYSTIC KIDNEY DISEASE

DEFINITION:

Polycystic kidney disease is an inherited disorder in which clusters of cysts develop primary within your kidneys, causing your kidneys to enlarge and lose function over time. Cysts are non cancerous ,round sacs containing fluid. The cysts vary in size , and they can grow very large. Having many cysts or large cysts can damage your kidneys.

Polycystic kidney disease is a group of monogenic disorder that results in renal cysts development. The morbidity associated with the most common forms , autosomal dominant PKD [ADPKD]and auto recessive PKD [ARPKD], is largely limited to the kidney and liver and extends from neonates to old age.

HISTORY:

Polycystic kidney disease in one of the most common inherited kidney diseases causing end stage renal disease . Although it has been in existence with humanity , it was defined in 18th century. The most detailed observations on PKD have been written after disease of STEPHEN BATHORY, THE KING OF POLAND. He had FATIGUE AND CHEST PAIN accompanied by UNCONSCIOUSNESS within few days after hunting trip , and died within 9days ,at the age of 53yearsin 1586.

Surgeon JAN ZIGULITZ described the cysts in his kidneys as large like those of bull, with an uneven and bumpy surface during the mummification . Based on available information ,347 years later ,a group of physicians and historians in Krakow concluded that the cause of kings death was PKD and uremia.

In 1888, FLIX LEJARS used the term polycystic kidney for the first time, and stressed that these cysts were bilateral and causing clinically identifiable symptoms .At the end of 19th century , the basic clinical signs, and genetic basis of disease have been better defined .However, the inheritance pattern could only be understood long years later.

GENERAL PHYSICAL EXAMINATION :

Patients with PKD usually are in discomfort. Physical examination of patients with polycystic kidney disease is usually remarkable for hypertension, jaundice ,pallor, icteric sclera, palpable nodular hepatomegaly , palpable flank

mass.

APPERANCE OF THE PATIENT :

Patients with PKD usually are in discomfort .

Vital signs : hypertension

Skin : jaundice , pallor

HEENT : icteric sclera.

Neck ,lung ,heart, genitourinary and neuromuscular examinations of the patient with PKD were usually normal .

Abdomen : a palpable abdominal mass in lumbar quadrant , palpable nodular hepatomegaly.

Back : palpable mass in flank , vertebral angle tenderness.

Extremities : pitting edema of upper /lower extremities.

PRESENTATION :

Pain in the abdomen , flank or back is most common initial complaint, and almost universally present in patients with autosomal dominant PKD [ ADPKD] pain may be caused by any of the following :

* Enlargement of one or more cysts
* Bleeding which may be confined inside the cyst , or lead to gross hematuria with passage of dots or peri nephric hematoma.
* Urinary tract infections
* Nephrolithiasis and renal colic

Rarely, a coincidental hypernephroma

In addition patients with ADPKD may have abdominal pain . Dull aching an uncomfortable sensation of heaviness. Patient with ADPKD may be at a higher risk of developing thoracic aortic aneurysms , abdominal aneurysms are not increased among these patients. pain may also develop for reasons completely unrelated to the underlying disease ; thus abdominal pain in patients with ADPKD ,may be diagnostic challenge. others include fatigue ,breathlessness,

Weakness and malaise .

CAUSES :

A genetic mutation causes PKD. In most cases, this means that the condition passes from parent to child in the DNA. Sometimes , a person can develop the gene mutation that causes PKD without receiving it form a parent. Scientists call this a spontaneous gene mutation , and it is rare.

PKD affects males and females equally . Age, Race, Ethnicity don’t seem to influence a person’s chances of having the disorder . However, people who have a blood relative with PKD are more likely to have the condition than those who don’t.

TREATMENT :

The severity to PKD varies from person to person even among members of same feeling. Often people with PKD reaches end stage kidney disease between age 55 and 65. But, Some people with PKD have disease and might never progress to end stage kidney disease.

Treatment of PKD involves dealing with the following signs, symptoms and complications in their early stage.

KIDNEY CYST GROWTH :

Tolvaptan therapy may be recommended for adults at risk of rapidly progressive ADPKD. It is a pill that you take by mouth that works to slow the rate of kidney cyst growth and the decline in how well your kidneys work.

There’s a risk of serious liver injury when taking tolvaptan , and it can react with other medicines you take .

HIGH BLOOD PRESSURE:

Controlling high blood pressure can delay the progression of the disease and slow further kidney damage. Combing a low-sodium , low –fat diet that’s moderate in protein and calorie content with not smoking, increasing exercise and reducing stress may help control high blood pressure.

However, medications are usually needed to control high blood pressure medications called ACEI’S and ARB’S are often used to control high blood pressure.