MEDICAL UNIVERSITY - PLEVEN
FACULTY OF MEDICINE

DISTANCE LEARNING CENTRE

DEPARTMENT OF "NEPHROLOGY, HEMATOLOGY AND GASTROENTEROLOGY"

PRACTICAL EXERCISES – THESES

FOR E- LEARNING IN NEPHROLOGY

ENGLISH MEDIUM COURSE OF TRAINING

SPECIALTY OF MEDICINE

ACADEMIC DEGREE: MASTER

PROFESSIONAL QUALIFICATION: DOCTOR OF MEDICINE

PREPARED BY DEPARTMENT OF NEPHROLOGY

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AUTOSOMAL DOMINANT POLYCYSTIC KIDNEY DISEASE

A. **Aim of these exercise.**The students have to learn how to make a diagnosis of autosomal dominant polycystic kidney disease, and how to manage it.

B. PKD is a subject of this topic because of its **importance**. PKD is spread throughout the world whit frequency between 1 in 200 and 1 in 1000 people. It is one of the most frequent hereditary diseases. It usually progresses to chronic renal failure, and it is one of the most usual causes of end-stage renal failure and haemodialysis as method of its treatment.

C. Diagnostic approach includes:

I. Taking an anamnesis.

II. Physical examination of the patient.

III. Laboratory data and data resulting from imaging techniques.

The students have to work whit an appropriate patient in about 40-min. time. They have to:

I. Take an anamnesis of the patient:

1. ***Present history*** - The present patient's may complain of:

a. **Pain** - It can have variable characteristic with regard to origin, severity, location, duration, character etc.

b. **Hematuria** - Approximately 30-50% of patients have micro- or macroscopic hematuria with or without inciting moment.

c**. Hypertension** - 50-60%. It is a frequent symptom too. Hypertension results from disturbance of water-salt balance.

d**. Abdominal tumor formation** - Some patients can find their self the enlarged kidneys as tumor formation in their abdomen.

e. Sign of some **complications of the disease**.

- **Pyelonephritis and inflammation of the cysts** - Patients have high temperature, lumbar pain, with/without dysury, etc.

- **Nephrolithiasis** - history of crisis with or without elimination of the calculi.

- **Mechanical complication** - Rupture of the cyst/s/ as a result of sport, transport etc.

f. Symptoms of coexisting abnormalities:

- History of cerebral hemorrhage - PKD can be associated with **intracranial aneurysms**. The aneurysms greater than 1cm in diameter are considered to have high risk for rupture.

- **Cardiovascular abnormalities** - Abnormalities of aorta, aortic or/and mitral valves - palpitation, atypical chest pain etc.

g. Features of **chronic renal failure.**

2. ***Past history*** - It can include the same complains as present history.

3. ***Family history*** is very important for diagnosis of PKD.

II. Physical examination - When the students have taken down a thorough anamnesis they have to examine their patient. The students have to try to find out signs of the disease and its complications. The careful physical examination is essential.

Enlarged kidneys with bumpy surface are important for the diagnosis.

III. The students have to consolidate all symptoms they have found by conversation with patient and its inspection into syndromes.

IV. The next step is to approach a **working diagnosis**.

V. The students must have opinion about the **differential diagnosis**.

VI. The students suggest ***laboratory and imaging examinations***.

They have notice that each examination has its indications and different possibilities of giving information about condition of renal and other systems. The data resulting from these various examinations must help them to confirm or reject a diagnosis, complications of the disease, etc.

1. **Laboratory data**.

a. **Blood examination**- Hb, Er, Hct, Leuco, serum urea, creatinine, and uric acid, serum electrolytes.

b. **Urinalysis**- proteinuria less than 1.0 g/l, hematuria.

c. **Functional analysis of urinary system**- clearance of creatinine, renal concentrating capacity /specific gravity of urine is an accurate expression of urine concentration/, isotope renogram, etc.

2. Imaging techniques:

a. **Ultrasonography** is the imaging method of choice. Typical marks of PKD include normal or enlarge kidneys, and multiple areas with varying size diffusely localized in renal cortex and medulla.

b. **Urography** - it is less sensitive than ultrasonography. Radiographic findings are: enlarged kidneys with irregular contour, distortion and splaying of the pyelon and calyces, hydronephrosis, calcification of cysts walls, etc.

c. **Computed tomography-** Because of its small advantages over US in sensitivity, CT is not preferred to initial screening. It is indicated for patients with an unusual form of PKD /ADPKD with small cysts less than 0,5 cm in diameter /or with complications of PKD /e.g. inflamed cysts, renal carcinoma, etc/.

3. **Genetic diagnosis** /Gene linkage analysis/- Blood test from patients or their family members can be used to detect /with a high degree of accuracy/ the carriers of the pathologic PKD-gene. Genetic diagnosis is a duty of the nephrologist or the genetic counselor.

D. The students have to discuss and sum up all data resulting from examination of their patient and must confirm diagnosis of PKD.

There are mean and second **diagnostic criterions**:

Main- Renal cysts

- Positive family history

Secondary - Liver cysts

- Enlarge kidneys
- Chronic renal failure
- Intracranial aneurysms.
- Others

Two main standards give diagnosis in 100%.

One main and one secondary criterion give diagnosis in high percent.

The students write down diagnosis. The diagnosis consists of:

1. Name of the disease /in Latin or English language/.
2. Any complications of the PKD.
3. Coexisting diseases etc.
4. The state of the renal function.

H. The students have to describe patient's problems and suggest what should be done as management.

Therapy of PKD.

1. There is not an etiologic and pathogenic treatment.
2. The treatment of the complications of PKD is essential with the goal to save the renal function and to protect the kidneys from further damage.

a. Treatment of acute or chronic pyelonefritis.

b. Treatment of the inflammation of the cysts.

- percutaneous or endoscopic puncture and drainage

- nephrectomy – for those patients in who prolonged therapy with appropriate antibiotics is unsuccessful. .

c. Treatment of arterial hypertension. – Various antihypertensive drugs can be administered.

d. Treatment of chronic renal failure**.**

- Conservative.
- In condition of end- stage renal disease – renal replacement therapy - dialysis methods.
- Transplantation.

The students have to know drugs- their general properties, indications and contraindications, application forms and dosages, unwanted effects. The students must write prescription for their patient.

The students should not forget that the correct diagnosis of the PKD requires a **consultation with a specialist of nephrology and with a genetic counselor**. Patients who suffer from PKD need to be traced by specialist.

J. In the end of the exercise the students may ask some questions if they have any.