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**

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# RENAL AMYLOIDOSIS

## 1. Definition

Satellite nephropathy as a result of formation and depositing in the kidneys of structurally pathologic and not typical for mammals protein with great mass leading to severe structural and functional renal impairment in the glomeruli and interstitial vessels and finally to irreversible chronic renal insufficiency (CRI). The disease is called β-fibrilosis and it is biochemically characterised by formation of insoluble complex of globular and fibrilar protein connected with polysaccharide bridge.

## 2. Brief analysis of the disease stages

The students retell in details about both main diffuse forms of amyloidosis, namely – primary – genetically determined and secondary one. Some of the localised forms as carpal-tunnel syndrome in patients on haemodialysis are mentioned. The assistant emphasises the discussion of the secondary one, the most common, form of renal amyloidosis. The diseases of many years leading to secondary amyloidosis are discussed: bronchial ecstasies, pneumosclerosis (chronic pneumonia), lung or non-lung form of tuberculosis, osteomyelitis, suppurate processes of perineum and pararectally, cholecystitis, Hodgkin's and non-Hodgkin's lymphomas and neoplasm's.

An important element is the determination of the earliest symptoms connected with deposit of amyloid in the parenchyma organs – liver, spleen, suprarenal glands and kidneys and mainly hepato-splenomegaly and arterial hypotension.

## 3. Handling of a patient having renal amyloidosis

***Anamnesis:***

- Relative's deceased respectively young – up to 50 years of heart failure and renal insufficiency draw the attention to primary amyloidosis.
- Patients having long lasting suppurate or neoplastic diseases are asked in details about the commencement of the arterial hypotension and the term of oedemas.
- It is asked about unmotivated weakness, faintness, lack of appetite and sharp changes in the diuresis volume, during the day and night hours.

***Physical examination*** (status):

The students have to determine an expressed skin and mucous membrane alabaster paleness. The assistant emphasises the characteristic stages of amyloidosis nephropathy and the late term of manifestation of the nephrotic, soft and testable oedemas in the lower limbs and the body as a consequence of the high sodium ($Na^+$) loss rate from hypocorticism.

For the cardio-vascular system: it is found arterial hypotension resistant to therapy due to suprarenal insufficiency. During heart percussion it is found cardiomegaly, more often in primary amyloidosis as a result of amyloidal infiltration of myocardium. Auscultatorily it is found tachycardia, systolic noise of Erb. The arterial pressure is very low, 80/40 mm/Hg, the pulse is slightly filled.

Hepato-splenomegaly is the most characteristic in palpating the abdomen, as it might be determined comparatively early. Nephrotic cold, soft and testable oedemas in the lower limbs, the body and the body hollow parts are determined quite late in the period of massive proteinuria.

***Tests:***

- *The haematological indices* form an anaemic syndrome: (Hb < 110 g/l). The anaemia is of complex origin: iron deficiency by consumption in reticuloendothelium as in spleenomegaly hemolysis is accumulated.
- *Biochemical tests* show very severe hypoproteinaemia (total protein < 40 g/l), and hypoalbuminaemia (<20 g/l) with very high gamma globulins from the proteinogram (35-40%). The high plasma fibrinogen is especially indicative in the advanced stages > 6,0 g/l.
- In case of amyloidosis progressive development for less than a year it reaches the stage of CRI as urea increase > 8,3 mmol/l and creatine > 125 μmol/l.
  Getting the main disease under control during the initial stage of amyloid deposit in the secondary renal amyloidosis stops the unfavourable changes in the parenchyma organs.
- *From the urine tests* it becomes obvious massive up to 10 g/24 glomerular and highly non-selective proteinuria with index > 0,5 – 0,6. The sediment is clear without any cells with hyaline and granular cylinders.
- *The functional samples* give limitation of the glomerular filtration rate and later of renal plasma flow.
- *The ultrasound test* shows large kidneys with very thick parenchyma. Liver and spleen are large too.

- ❖ *Puncture renal biopsy* reveals the diagnosis 100%. Amyloid is deposited mainly around the small vessels and along the hilus in the glomerulus and it is made positive with Congo red. Biopsy of rectal and gingival mucous membrane is in 70% positive.
- ❖ Venous urography is not made.

**4. Diagnosis**

- ❑ Anamnesis for preceding disease and heredity
- ❑ Clinical manifestation with hypotension, hepato-splenomegaly, massive proteinuria, late oedemas
- ❑ Puncture renal biopsy (rectal and gingival)
- ❑ Writing in Latin of a working diagnosis

**5. Differential diagnosis:** Mainly it is discussed:

1. Chronic glomerulonephritis
   - Hypertension
   - Without hepatosplenomegaly
2. Collagenous lupus nephropathy
   - hypertension
   - anti DNA antibodies positive

**6. Therapy**

- ❑ In practice there is therapy for primary amyloidosis. The following have been used with respective effect:

*1. Resochin tabl. 250 mg S. 2 x 1 t./daily*

*2.Melphalan 2 mg/kg/daily*

- ❑ For secondary amyloidosis it is of great importance the therapy of the main disease led to the amyloid deposition.
  - The antibiotic therapy should be energetic and accompanied by surgical interventions
- ❑ Symptomatic therapy
  1. Infusions of Human albumin 18% bags 100 ml x 1 bag/daily
  2. Native plasma, bags of 300 ml, 1 b. daily for influencing hypoalbuminaemia and nephrotic oedemas
  3. Diuretic therapy

*Furosemid tabl. 40 mg S. 2 x 1 t./daily until diuresis of 2500 ml/24 h.is obtained.*

4. *Predmisolon S. Tabl. 5 mg x 1 tabl. daily in the morning*
5. Haemodialysis therapy

❖ Writing a prescription