ABSTRACT

Familial amyloid polyneuropathy is a disease that occurs in southwestern Bulgaria. It causes a severe disability. The clinical picture is accompanied by weight loss, a muscle weakness, a cardiomyopathy and damage to the peripheral nervous system. The onset of symptoms is at a relatively early age (45-50 years). Specialists from Alexandrovskia Hospital monitor patients with this type of disease.

**Purpose:** The aim of the study is to create a kinesitherapeutic algorithm for the study of patients with familial amyloid polyneuropathy.

**Methods:** The review study was undertaken in the Neurology department at Multidisciplinary hospital for active treatment- Blagoevgrad, Bulgaria. The study was conducted for the period 2015-2017, during which 10 patients with FAP were examined. In our algorithm we have included manual-musculå testing (MMT) of the upper and lower limbs, an centimeter testing to measure hypotonia and a discriminant test to detect sensory disturbances.

**Results:** The study involved 6 women and 4 men. The average age of the studied contingent is 48.0 ± 12.7 years. In the study we found that there is a change in the study of manual muscle testing. One unit increase available. There are no statistically significant differences in the study of centimeter in the lower extremities and in the study of the senses as well. Severe muscular hypotension and distal hypoesthesia persist.

**Conclusion:** Unfortunately, these patients often progress rapidly. They remain disabled. Kinesitherapy, with its methods, can improve the quality of life.

**Keywords:** physiotherapy, familial amyloid polyneuropathy, methodology.

INTRODUCTION

Because the disease is rare in recent years sought an optional method of testing and physiotherapy. In our study, we provide a specialized methodology of testing of patients with expression by the peripheral nervous system. Peripheral polyneuropathy characterized by sensorimotor disorders, in some embodiments. In TTR, V30M is primarily affected the lower limbs [1, 2]. With other mutations and other TTR I84S TTR L58H typical peripheral neuropathy, which is a leader in clinical and affect the upper limbs [3, 4, 5]. The patients complain of pain and weakness in the same. With progression of the disease leads to complete disability and need for facilities for movement. The peripheral nervous system, which is affected, can be performed with hyperalgesia and abnormal temperature sensitivity [6]. Dysfunction of the autonomic nervous system is frequent and severe and manifests as weight loss, retention or incontinence of urine and gastric emptying difficult.

**METHODS**

The review study was undertaken in the Neurology department at Multi-profile hospital for active treatment Blagoevgrad, Bulgaria. The study was conducted for the period 2015-2017, during which 10 patients with FAP were examined. In our algorithm we have included manual-musculå testing (MMT) of the upper and lower limbs, an centimeter testing to measure hypotonia and a discriminant test to detect sensory disturbances [1, 3, ]. The study started at the time of registration of the disease. Starting with methodical clinical blood to identify the type of disease, electromyography, must be done in consultation with a cardiologist and is held echocardiography. In an extreme case makes tissue biopsy. Some tests are only done once to confirm a diagnosis, while others may be repeated to monitor the disease and response to therapy. There is scant information on the conduct of physiotherapy in this type of patients. As a suggestion of physiotherapy perspective recommended manual muscle testing of the upper and lower limbs. Testing has a six-point scale. Cantimetry of hypotrophic limbs; Sensory examination; Gaits examination - gait studied by tracking the posture of the patient in the right position, the position of the limbs while walking back and forth, tracking the speed of movement, the ability to
rhythmic movements and the presence of normal synkinesis limbs. We think it is an extremely important to study of the activities of daily life. There is a large selection of tests, but in our opinion, the most comprehensive and accurate test is the Barthel index.

Methods for assessing the functional status of patients (Kinesitherapy methods):

- MMT of upper and lower limbs - testing has a 6-point scale and values from 0 to 5 (fig. 1).

Fig. 1. Manual muscle testing of m. quadriceps femoris grade 3

Fig. 2. Sensory examination - sensation of pain

• Gait examination;
  Gait studied by tracking the posture of the patient in the right position, the position of the limbs while walking back and forth, tracking the speed of movement, the ability to rhythmic movements and the presence of normal synkinesis limbs. Assessed in the following degrees:
  • 0 – can not walk with aids;
  • 1 – can walk a few steps with assistance;
  • 2 – can walk with assistance or aid in the room;
  • 3 - can walk alone or with the aid of up to 500 m.;
  • 4 - walk independently, free climbing stairs.

- Sensory examination.
  The sensory examination is one of the most difficult elements of neurological status because assessing sensory disorders is based on the subjective response of the patient. It depends on the clarity of mind of the patient, his intelligence, educational status, desire for cooperation and other factors. The examination of tactile sensitivity is achieved by a cotton swab or brush to quantify the touch feel. The patient eyes are closed. The availability of reduced sensitivity is referred to as tactile hypesthesia or anesthesia when it is completely absent. The sensation of pain is performed by a slight sting with a sharp object in the area of the lower legs (fig. 2).

- Santimetry of hypotrophical limbs;

The sensory examination is one of the most difficult elements of neurological status because assessing sensory disorders is based on the subjective response of the patient. It depends on the clarity of mind of the patient, his intelligence, educational status, desire for cooperation and other factors. The examination of tactile sensitivity is achieved by a cotton swab or brush to quantify the touch feel. The patient eyes are closed. The availability of reduced sensitivity is referred to as tactile hypesthesia or anesthesia when it is completely absent. The sensation of pain is performed by a slight sting with a sharp object in the area of the lower legs (fig. 2).
RESULTS
The average age of the contingent was 48.0 ± 12.7 years. The study involved 6 women and 4 men. In the study we found that there is a change in the study of manual muscle testing (fig. 3). One unit increase available. There are no statistically significant differences in the study of centimeter in the lower extremities and in the study of the senses as well (Chi-square test). Severe muscular hypotension and distal hypoesthesia persist.

CONCLUSION
Developing rapidly interventional medicine and the introduction of new drugs into practice opens broad prospects in neurology requiring adequate involvement by physiotherapy as part of the proactive approach in the recovery of the patient. FAP is a progressive disease. The outcome was fatal and occurs within 5-6, rarely 10 years after the occurrence of the first symptoms. Affect the area of Blagoevgrad, making it an endemic area of the country. Kinesitherapy assists the patient in making the easier of the available active movements and activities of daily life.

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Fig. 3. Manual muscle testing

![Fig. 3. Manual muscle testing](image-url)