U 1 Neurology, medical genetics

U 2 Neuromuscular disease

#SPINAL AMIOTROPHIES THIS:

+ a group of progressive neuromuscular diseases in which muscle weakness is caused by damage to the spinal cord

the same, but muscle weakness is due to primary nerve damage

a group of hereditary diseases characterized by an increase in muscle weakness and atrophy

a group of progressive neuromuscular diseases in which the focus is located in the brain stem

a group of progressive neuromuscular diseases in which muscles are primarily affected

# THE FOLLOWING DISEASES ARE SPINAL AND MYOTROPHIES:

Charcot-Marie

Dejerine-Sotta

+ Verding-Hoffmann

Kugelberg-Velander

Erba Rotta

Landusi Dejerina

# FOR VERDING-HOFFMANN'S DISEASE TYPE OF INHERITANCE:

autosomal dominant

+ autosomal recessive

X-linked

holondric

mitochondrial

# IN THE ACUTE PERIOD OF NEUROPATHIES IT IS IMPOSSIBLE TO APPLY:

electrophoresis of novocaine

+ electrical stimulation

diadynamic currents

dehydration therapy

anti-inflammatory therapy

# WITH VERDING-HOFFMAN SPINAL AMIOTROPHY, THE LEADING SYMPTOMS ARE

flaccid paresis

muscle hypotension

tendon areflexia

decreased muscle strength, gait changes

+ all of the above

# EARLY FORM OF VERDING-HOFFMANN'S DISEASE BEGINS TO BE MANIFESTED:

since birth

+ up to 1.5 years

1.5-2 years

after 5 years

after 10 years

# FORM OF VERDING-HOFFMANN'S DISEASE, FOR WHICH IN THE FIRST DAYS AFTER BIRTH, LACER PREPARES OF EXTREMITIES, LOW MUSCULAR TONUS, BULBAR DISORDERS ARE CHARACTERISTIC

early

late

+ congenital

ultrafast

short-term

# FOR THE CLINIC OF THE EARLY FORM OF WERDNIG-HOFFMANN DISEASE CHARACTERISTIC:

+ loss of previously acquired motor skills

patients retain mobility for a long time

pseudohypertrophy of the calf muscles appears

increased reflexes

pathological stop signs

# INTELLIGENCE, AS A RULE, REMAINS SAVED WITH THE FOLLOWING FORM OF HYDROCEPHALIA:

congenital form

early form

+ late form

short-term

heavy

# KUGELBERG-WELANDER'S DISEASE IS DEVELOPING:

in the first days after birth

up to 1.5 years

+ from 2 to 17 years

already in utero

after 50 years

# KUGELBERG-WELANDER'S DISEASE IS DESCRIBED:

proximal paresis

pseudohypertrophy of the calf and gluteal muscles

fascicular muscle twitching

hyporeflexia, areflexia

+ all of the above

# THE COURSE OF KUGELBERG-WELANDER DISEASE:

+ benign, patients retain the ability to move independently for a long time

independent walking ability rarely

since birth sick patients

quickly leads to death

progressing

# DURING THE BIOCHEMICAL STUDY OF BLOOD DURING KUGELBERG-WELANDER DISEASE DETECT:

+ moderate increase in creatine phosphokinase

creatine phosphokinase, aldalase normal

decreased creatine phosphokinase

bilirubin increase

decreased ALT

# TREATMENT OF ALL FORMS OF SPINAL AMYOTROPHY:

strictly specific

+ symptomatic

not carried out

anti-resident

only massage and exercise therapy

# NEURAL AMIOTROPHIES THIS:

a group of progressive neuromuscular diseases in which the weakness of the muscle apparatus is caused by damage to the spinal cord

+ same, but muscle weakness is due to primary nerve damage

a group of hereditary diseases characterized by an increase in muscle weakness and atrophy

a group of progressive neuromuscular diseases in which the pathological focus is located in the muscles

a group of progressive neuromuscular diseases in which the focus is located in the lateral columns of the spinal cord

# TO THE GROUP OF NEURAL AMIOTROPHIES RELATED:

+ Charcot-Marie,

Dejerine-Sotta

Kugelberg-Velander

Verding-hoffman

Erba Rotta

Landusi Dejerina

# FOR DISEASES OF SHARCO-MARI TYPE OF INHERITANCE:

+ autosomal dominant

autosomal recessive

linked to the X chromosome

mitochondrial

holondric

# MOST DISEASE CHARCOT-MARIE DEVELOPS

up to 3 years

+ at school age

in adulthood

already in utero

at 4 - 5 years old

# THE BASIC CLINICAL SYMPTOMS IN HEREDITARY POLYNEUROPATHIES ARE:

distal atrophy of the feet, bones

polyneuritic type of sensitivity disorder

decrease in speed of peripheral nerves (according to EMG)

gait change

+ all of the above

# THE BASIC DIAGNOSTIC CRITERIA OF WILSON-KONOVALOV DISEASE ARE:

copper metabolism defect

enlarged liver, Kaiser-Fleischer ring around the iris

decrease in total serum copper and serum ceruloplasmin levels

extrapyramidal disorders

+ all of the above

# THE BASIC DIAGNOSTIC CRITERIA OF A DEFORMING MUSCULAR DYSTONIA ARE:

+ torsion hyperkinesis of the muscles of the trunk, limbs, head

muscle dystonia

hyperreflexia of knee, Achilles reflexes, pathological reflexes

nystagmus

cerebellar disorders

# PROGRESSING MUSCULAR DYSTROPHY (TYPICAL FORM) IS DESCRIBED:

+ the first symptoms, as a rule, appear by the end of the 1st year of life by a delay in the rate of motor development

the first symptoms appear over the age of 2 years

first symptoms appear at school age

the first symptoms appear from 20 to 25 years

the onset of progression is still in utero

# FOR SHARCO-MARI DISEASE CHARACTERISTIC:

+ distal paresis, distal atrophy

proximal paresis

proximal atrophy

mosaic atrophy

all of the above

# MYOPATHY:

a group of progressive neuromuscular diseases in which the weakness of the muscle apparatus is caused by damage to the spinal cord

the same, but muscle weakness is due to primary nerve damage

+ a group of hereditary diseases characterized by an increase in muscle weakness and atrophy. Pathological focus in the muscles

group of diseases in which the focus is located in the channels of the membranes

group of diseases in which the focus is located in the cerebral cortex

# IN MYOPATHY, PRIMARY INFLUENCE OF:

cells of the anterior horns of the spinal cord

peripheral nerve trunks

+ skeletal muscle

craniocerebral nuclei

back columns of the spinal cord

# CHARACTERISTICS OF PATOMORPHOLOGICAL CARD OF PROGRESSING MYOPATHIES ARE:

muscle destruction, proliferation of nuclei

replacement of muscle fibers with connective tissue

replacement of muscle fibers with adipose tissue

the number of mitochondria in the muscle fiber, muscle hypotension

+ all of the above

# PROGRESSING MYOPATHIES RELATED TO:

Charcot Marie

Verding-hoffman

Kugelberg-Velander

Degerina Sotta

+ Landusi-Dejerine, Erba-Rotta

# TYPE OF INHERITANCE FOR DYSHENNA DISEASE:

autosomal dominant

autosomal recessive

+ linked to the X chromosome

mitochondrial

holondric

# IF FREEDREICH'S DISEASE TAKES PLACE:

+ recessive type of inheritance

dominant type of inheritance

sex-linked (via the X chromosome)

holondric type of inheritance

mitochondrial inheritance

# SUFFICIENT CLINICAL SIGNS IN THE DIAGNOSTICS OF SYRINGOMELIA ARE:

+ segmental dissociated, the presence of dysraphic features of the structure of the musculoskeletal system

conductor type disturbances on the opposite side

progressive muscle atrophy in areas corresponding to segmental dysfunctions

lower spastic paresis

coordination violations

# SEVERE STAGE OF DUSHEN'S DISEASE CHARACTERIZED:

the impossibility of independent movement

the rapid development of contractures

skeletal deformity

severe motor impairment

+ all of the above

# IF ERB-ROTT'S DISEASE FIRST, MUSES ARE AFFECTED:

distal extremities

faces

+ pelvic girdle, shoulder girdle

proximal limbs

backs

# FOR SPINAL AMYOTROPHY WERDNIG-HOFFMANN CHARACTERISTIC

increased serum creatinine kinase

congenital disorders in the structure of muscle fiber

+ damage to the spinal cord motor neuron

hypertonicity

increased reflexes

# FOR NEUROFIBROMATOSIS CHARACTERISTIC:

the disease always proceeds for a long time, but benign

the disease can be detected in one of the parents of proband, the presence of multiple pigmented nevi

type of inheritance - autosomal dominant

neurofibromatosis gene expression variable even within the same family

+ all of the above

# CHARACTERISTIC SYMPTOMS OF ERBA-ROTT DISEASE ARE:

atrophy of the muscles of the shoulder and pelvic girdle

pterygoid scapula

wasp waist

hypomimia, the face of the "myopath"

+ all of the above

# THE COURSE OF ERBA-ROTT'S DISEASE:

+ relatively favorable

quickly leads to immobilization

quickly fatal

ultrashort

often recurring

# TYPE OF INHERITANCE FOR LANDUSI-DEGERIN DISEASE:

+ autosomal dominant

autosomal recessive

X-linked

holondric

mitochondrial

# FOR LANDUZI-DEGERIN'S DISEASE, DEFEAT PREVENTS:

distal extremities

+ faces

shoulder girdle

shoulder and pelvic girdle

backs

# UNLIKE MYOPATHY DURING ATONIC FORM OF CP:

movement disorders, gait changes

increased tendon reflexes

persistent pathological signs

improvement of motor functions

+ all of the above

# FOR FORECASTING POSSIBLE REPEATED BIRTH OF A SICK CHILD IN A FAMILY WITH A NEUROMUSCULAR DISEASE, IT IS NECESSARY TO CARRY OUT:

+ genetics consultation

neurologist consultation

orthopedic consultation

pediatrician consultation

optometrist consultation

# TOMSON'S DISEASE ARISES

after infection

+ congenital disease

after severe injury

during the epiprush

as a complication of rickets

# MYASTENIA IS CHARACTERIZED:

+ pathology of the synaptic apparatus

damage to the cells of the anterior horns of the muscles of the spinal cord

peripheral nerve damage

damage to the lateral horns of the spinal cord

damage to the posterior columns of the spinal cord

# FOR THE CLINIC OF MIASTENIA CHARACTERISTICALLY:

+ decrease in volumes and strength of movements with repeated contractions of muscles

inability to relax muscles after contraction

bone deformities

epiprules

coordination violations

# BY THE DEGREE OF PREVALENCE OF MOTOR DISORDERS IN MIASTENIA, IDENTIFY:

+ generalized

local forms

only bulbar

generalized only

ophthalmoplegic form

# FOR WHAT FORM OF MYASTENIA DYSARTRY IS CHARACTERISTIC , CHANGE OF VOICE, SURFACE:

with damage to the oculomotor muscles

+ bulbar

mimic

generalized

born

# AT MYASTENIA TENDON REFLEXES:

promoted

+ quickly depleted

are absent

with the expansion of reflexogenic zones

with clonus stop

# TO CONFIRM DIAGNOSIS OF MYASTENIA CONDUCT

+ proserin test

amidopyrine test

caffeinated sample

diazepam test

sweat test

# ACUTE MYASTENIC CRISES CHARACTERIZE:

generalized muscle weakness

respiratory failure

bulbar disorders

cardiac impairment

+ all of the above

# THE BASIS OF TREATING MYASTENIA IS APPLICABLE:

+ anticholinesterase drugs

antiviral drugs

antibiotics

antiepileptic drugs

hormonal drugs

# ANATOMICAL AREA, WHICH MOSTLY AFFECTS DURING SPINAL AMIOTROPHY OF VERDING - Hoffmann:

central motor neuron

+ front-horn motor neuron

peripheral nerve

horn of the spinal cord

lateral horn of the spinal cord