MINISTRY OF HEALTH OF UKRAINE

O.O. BOHOMOLETS NATIONAL MEDICAL UNIVERSITY

CONGENITAL DEFORMITIES AND ABNORMALITIES OF THE DEVELOPMENT OF THE HAND. CLASSIFICATION, DIAGNOSIS, TREATMENT

WORK BOOK

For independent work of students of the 5th course

Study discipline "Traumatology and Orthopedics"

direction “Medicine”

specialty “Curative care”

Department of Traumatology and Orthopedics

Authors: prof. Burianov A.A., assist. Tsygankov M.A.

Introduction

One of the effective means of organizing an independent work of students on topics of disciplines assigned for independent study is the work of a student with his/her workbook. Work with the workbook should begin with the acquaintance with the key issues on the topic. The next step includes the acquaintance with the list of sources from which the student can find the answers to the posed questions. For more deep study of the problem students can visit professional websites.

Having become acquainted with the theory, the student needs to assess his/her degree of mastering the material. In this regard, he/she resolves the proposed tasks; answers test questions on the topic. Students should pay particular attention in preparing for classes to the required minimum of practical skills to be mastered. In the relevant sections of textbooks, manuals, he must glean the information that he needs for mastering further practical skills.

Arrangement of independent work with the use of workbook is conducted as follows: tutor provides the workbook to a student in digital format (to be downloaded from website of the Department), or in printed version; later the students do the tasks at their extra-curricular time, whereupon the tutor checks and assesses them **at the initial stage of practical classes.**

Criteria for assessing the tasks in the workbook

Each task requires a separate approach when assessing the quality of its implementation under particular criteria. And yet, with a 5-point evaluation ranking for each type of tasks, one should observe the general didactic criteria, namely:

"5" is graded when the student:

1. Executed the work w/o errors and deficiencies.
2. maximum one deficiency.

"4" is graded when the student executed the work in full but made:

1. maximum one gross error and one deficiency.
2. maximum two deficiencies.

"4" is graded when the student executed at least one half of a work correctly or made:

1. maximum two gross errors or one gross and one mild errors and one deficiency.
2. maximum two mild errors or one mild error and three deficiencies.
3. In the lack of errors but when four or five deficiencies are available.

"2" is graded when the student made:

1. number of errors (deficiencies) exceeding the limit when grade "3" could be applied.
2. In case less than half of work is completed.
3. Failed to start the work.

Congenital deformities and abnormalities of the development of the hand. Classification, diagnosis, treatment.

Goal (Educational goals):

1. To determine the etiological (exo- and endogenous) factors of congenital deformities and abnormalities of the development of the hand.

2. Conduct analysis of a typical clinical picture of congenital deformities and abnormalities of the development of the hand.

3. To establish the diagnosis of congenital deformities and abnormalities of the development of the hand.

4. Differentiation of congenital deformities and abnormalities of the development of the hand with other acquired disorders

5. To substantiate and formulate a preliminary diagnosis of congenital deformities and anomalies of the development of the hand

6. To draft a plan of examination of patients with congenital deformities and abnormalities of the development of the hand

7. To analyze the data of clinical, laboratory and instrumental methods of examination of patients with congenital deformities and anomalies of development of the hand.

8. To choose the optimal method, terms and stages of treatment of patients with congenital deformities and anomalies of the development of the hand.

The student should know:

1. Etiological (exo- and endogenous) factors of congenital deformities and abnormalities of the development of the hand.

2. Classification of congenital deformities of the hand.

3. Clinical picture congenital deformities of the hand.

4. Classification of congenital deformities of the hand.

5. Diagnostic criteria for various types of congenital deformities of the hand.

6. Principles of conservative and surgical treatment of congenital deformities of the hand.

7. Prognosis of the disease, working ability and social rehabilitation of patients with congenital deformities of the hand.

8. Etiological factors of development of congenital abnormalities of the hand.

9. Classification of developmental abnormalities of the hand.

10. Clinical picture of congenital abnormalities of the development of the hand.

11. Diagnostic criteria for various abnormalities of the development of the hand.

12. Principles, terms, ways of conservative and surgical treatment of patients with congenital abnormalities of development of a brush

13. Prognosis of the disease in patients with congenital abnormalities of the development of the hand.

14. Principles and main tasks of rehabilitation of patients with congenital an abnormalities and deformities of the hand.

A student must be able to:

1. To determine the exogenous and endogenous etiologic factors of occurrence of congenital developmental abnormalities, or deformities of bones in children.

2. Analyze a typical clinical picture of congenital deformity and abnormality of development of the hand.

3. To substantiate and formulate a preliminary diagnosis of congenital deformity and anomaly of development of the hand, according to the existing classification.

4. To draft a plan for examining a patient with congenital deformity or an anomaly of development of the hand, involving clinical, laboratory and instrumental methods of research.

5. To be able to interpret the results of instrumental research methods with anomalies of development and congenital deformities of the hand.

6. To conduct a differential diagnosis of congenital malformations and deformities of the hand with other diseases

7. To define tactics of conservative and surgical treatment of the patient, conduct preparation for surgical treatment, plan rehabilitation measures.

8. To forecast the course of the disease and prospects for workability and social adaptation of the patient.

Basic terms of the topic:

Term	Definition

Congenital anomalies	Various forms of impaired development of organs and tissues. there are 3 groups of congenital anomalies based on the etiology: - hereditary, resulting from inherited or spontaneous mutations; hereditary anomalies can be divided into genomic, chromosomal and gene; - exogenous, caused by infectious or toxic teratogenic damage to the embryo or fetus (for example, intrauterine infection: toxoplasmosis, syphilis, rubella, cytomegalovirus, herpes simplex virus, HIV) - multifactorial.
Syndactyly	Abnormality of development: complete or partial fusion between two or more adjacent fingers or toes. It constitutes 1/2 of all anomalies of the upper limbs. Syndactyly (dermal) - with the presence of a thick web of skin and soft tissues. Syndactyly (dermal membranous) is often incomplete, with the presence of a thin dermal membrane. Syndactyly (ending) - only at the level of terminal phalanges. Syndactylynia (bony) - fusion of phalanges of fingers.
Madelung disease (congenital subluxation of the wrist)	Developmental abnormality, which is the local dysplasia of the radius; causes a lag in its growth in length. The ulnar bone grows normally and there are distortions of the forearm, the inclination of the wrist and hand towards the radius with dislocation of the ulnar bone head. Treatment - surgical.
Amniotic Finger Bracing	It is rare congenital defect. Usually it accompanies syndactyly or brachydactyly. Bracing could be superficial and deep. The latter can cause disturbances in lymph- and blood circulation followed by further trophic disorders.
Focomelia or Amelia	Developmental abnormality, when the upper limb is absent completely, or the shoulder and forearm are absent, and the hand moves away from the underdeveloped shoulder and resembles the seal's fin. Sometimes instead of a hand from an underdeveloped shoulder such underdeveloped shoulder ends with only one finger. This condition is called peremelia.
Ectromelia	Complete absence of one or two limbs or part of a limb.
Hemimelia	Abnormality of development, when there is no lower part of the limb (hand or hand and forearm), while the upper parts are developed normally.
Polydactyly	Congenital abnormality characterized by the availability of "extra" fingers on the hands or on the feet. Along with polydactyly, there is also adactyly.
Brahidactyly	Brachydactylia is an underdevelopment of distal phalanges of fingers; ectrodactyly cleavage of the hand or pincer hand. Absence or underdevelopment of middle fingers or metacarpal bones.

Oligodactyly	It is a developmental defect, when there is no part of the fingers on the hands or feet. The radial form is the absence of the thumb and other fingers on the radial side of the hand. The ulnar form is characterized by the absence of 5 fingers and other fingers on the ulnar side of the hand.
Macrodactyly	Congenital developmental anomaly characterized by a disproportionate increase in the size of one or more fingers of the hand
Hypoplasia of the first finger of the hand	Congenital abnormality of the hand's development characterized by underdevelopment or lack of the first finger of the hand.
Congenital manus vara	It occurs in the lack of development of muscles on the forearm, or - underdevelopment of the radial or ulnar bone.. The ulnar *manus vara* develops, in case of lack of development of muscles of the elbow part of the forearm; the ulna is missing completely or partially. There may be a simultaneous underdevelopment of 4-5 fingers of the hand, parts of metacarpal bones and wrist from the ulnar side. The wrist under this condition is sharply turned to the ulnar side (outward), its function is disrupted. Radial stranded. This abnormality develops in the absence (underdevelopment) of the radius. The defect can be two-sided. I.e. simultaneously: the underdeveloped muscles of the forearm from the radial side, the first finger of the wrist, bones of the wrist from the radial side. The wrist is inclined towards the radius (inside), its function is severely disrupted, and due to the absence of the first finger of the wrist, it is impossible to capture objects.
Camptodactyly	Congenital or family contracture of the fingers of the hand can develop on both sides in isolation on the 5^{th} finger. This is an unwilled contraction, uncompensated. It is associated with changes in the tendon sheath, causes a shortening of the tendon of one or more fingers or feet. It is necessary to differentiate it with the acquired contracture of Dupuytren, the ischemic contracture of Volkmann, the neurogenic contracture after trauma to the ulnar nerve.
Clinodactyly	Congenital defect of finger development. Outwardly manifested in their curvature and altered position (deformation) relative to the limb axis (fingers oblique medially or laterally). In addition to the deformation of the phalanges of the fingers, there is a disorder of the joint surfaces between themselves. It progresses in the course of puberty period

Reference literature:

1. Nguyen M.P. A case report of bilateral mirror clubfeet and bilateral hand polydactyly / M.P. Nguyen, E.A. Lawler, J.A. Morcuende // Iowa Orthop. J. – 2014. – Vol. 34. – P. 171-174.

2. Senes F.M. Correction of forearm deformities in congenital ulnar club hand: one-bone forearm / F.M. Senes, N. Catena // J. Hand. Surg. Am. – 2012. – Vol. 37, № 1. – P. 159-164.

3. Symmetrical upper limb peromelia and lower limb amelia associated with persistent omphalomesenteric duct: a case report / S. Puvabanditsin, J. Savla, E. Garrow [et al.] // Clin. Dysmorphol. – 2011. – Vol. 20, № 2. – P. 102-106

4. Агранович О.Е. Врожденная гипоплазия I луча кисти / О.Е. Агранович // Дет. хирург. – 2009. – № 3. – С. 42-46.

5. Скляренко Є.Т. Травматологія і ортопедія. – К.: "Здоров'я ", 2005. – 386с..

Tasks for unsupervised work.

To be answered in written.

Variant 1

Task 1.

What are the main external factors affecting the development of congenital anomalies of the development of the hand:

1 Ionizing radiation

2 Magnetic field

3 Solar irradiation

4 Viral diseases of the pregnant woman

5 Environmental factors

Task 2:

Indicate the timing of intrauterine development, in which the hand is formed

1 8-10 weeks

2 3-4 weeks

3 6-7 weeks

4 10 to 12 weeks

5 2-3 weeks

Task 3:

What are the typical abnormalities of the development of the hand

1 Adactyly

2 *manus vara*

3 Ectrodactyly

4 amniotic constrictions

5 Camptodactyly

Task 4:

What are the typical types of grip?

1 Cylindrical

2 Direct (indirect)

3 Plucked

4 Finger (2, 3 fingers)

5 Spherical

Task 5:

What are the forms of syndactyly

1 Skin

2 Tendonous

3 Bone

4 Articular

5 Congenital

Task 6:

What are the principles of surgical treatment of congenital anomalies in the development of the hand

1 Restoration of finger grips

2 Fixing the acquired skills of using the hand

3 Social rehabilitation of a child in society

4 Psychological assistance to parents and children

5 Development of compensatory mechanisms

TEST QUESTIONS (choose one correct answer)

1. Patient B. 3 months old. Physiological genera. Fetal head presentation. Parents are healthy. On the right forearm on the back surface in the lower third - an oblique hypertrophic scar to 3 cm, there is a "drawing in" of the skin. The skin of the back surface of the hand is cyanotic, there is a slight deficit of extension of 2,3,4 fingers. Parents deny the trauma of the child. Establish the diagnosis:

A Child abuse

B Old open lesion of extensor tendons

C Amniotic constriction

D Keloid scar of right forearm

E Desmogenic contraction of the fingers of the hand

2. Patient E. 5 years. The development is harmonious, corresponds to the age norms. 2nd and 3d fingers of the left hand fixed with each other, nail plates separate on 2nd and 3d fingers, flexion and extension of fingers, the skin is not changed. Diagnosis: skin syndactyly of the 2^{nd} and 3d fingers of the left hand. Surgical treatment is planned. What is the sequence of actions:

A Radiography, separation of syndactyly, gypsum immobilization

B Ultrasonography, separation of syndactyly

C Separation of Syndactyly, Skin Plastic

D Radiography, separation of syndactyly, skin plastic

E Conservative treatment up to 7 years

3. The patient of 15 complains of absence of the 1^{st} finger of the right hand. After X-ray examination 1 metacarpal bone is visualized, complete absence of phalanges of the 1^{st} finger. Choose treatment tactics:

A Surgical deepening of the first interstitial space

B Skin plastics

C Aesthetic prosthetics

D Transplantation of the 2nd finger from the foot to the wrist into the position of the 1st finger

E Functional rehabilitation

4. Patient B of 4. Parents noticed a deviation of the hand towards the ulnar side, progressing with age. It was found that the child had a fractured clavicle during delivery. The movements of the fingers of the hand are preserved, the weakness of flexion of 4 and 5 fingers. Establish a diagnosis:

A Pathological fusion of the bones of the forearm

B Obsolete damage to the brachial plexus

C Elbowed *manus vara*

D Madelung disease

E Radiant *manus vara*

5. Child of 12, male. Established diagnosis: focomelia. The wrist is located on an underdeveloped shoulder, the length of the humerus is 15 cm, the shoulder joint is preserved, the forearm is absent, blood circulation is compensated, the fingers of the hand are functioning. The child reaches out to the face, head, neck, draws and writes with this hand. Choose treatment tactics:

A Skin plastic, staged lengthening of the limb

B physiotherapy, massage, physical rehabilitation

C Observation in dynamics

D Elongation of the shoulder according to Ilizarov's technique

E Amputation of the hand, bionic prosthetics of the forearm and hand

Variant 2

Task 1.
What are the main endogenous factors affecting the development of congenital of the hand:

1 Ionizing radiation

2 Chromosomal mutation

3 Solar irradiation

4 Genetic predisposition

5 Environmental factors

Task 2:

What are the diseases that can lead to the development of a congenital anomaly of the hand

1 acute respiratory infection

2 Syphilis

3 Diabetes mellitus

4 Herpes

5 HIV

Task 3:

What are the typical congenital deformities of the hand

1 Adactyly

2 *manus vara*

3 Ectrodactyly

4 Amniotic Bracing

5 Camptodactyly

Task 4:

What are the forms of forms of oligodactyls

1 Radial

2 Complicated

3 Congenital / acquired

4 Elbow

5 Progressive

Task 5:

What are the forms of *manus vara*

1 Arthrogenic

2 Tendonous

3 Bone

4 Beam

5 Elbow

Task 6:

What are the principles of surgical treatment of congenital anomalies of the development of the hand

1 Restoration of bone structures

2 Fixing the fingers in the physiological position

3 Social rehabilitation of a child in society

4 Aesthetic correction of the appearance of the limb

5 Maximum restoration of hand function

TEST QUESTIONS (choose one correct answer)

1 Patient S. 5 months old. Physiological delivery. Fetal head presentation. Parents are healthy. 1st finger of the left hand is reduced in size, 2nd and 3d fingers are missing. Establish a diagnosis:

A Adactyly of the 2nd and 3d fingers

B Congenital amputation of fingers

C Amniotic constriction

D Radial oligodactyly

E Hypoplasia of the 1st finger of the left hand

2 Patient E. of 5. The development is harmonious, corresponds to the age norms. 2nd and 3d fingers of the left hand fixed with each other, nail plates are fused on 2nd and 3d fingers, flexion and extension of fingers is friendly, the skin is not changed. Diagnosis: Bone syndactyly of 2nd and 3d fingers of the left hand. Surgical treatment is planned. Name the sequence of actions:

A Radiography, separation of syndactyly (osteotomy), skin plastics, gypsum immobilization

B Ultrasonography, separation of syndactyly

C Separation of Syndactyly, Skin Plastics

D Radiography, separation of syndactyly, skin plastics

E Conservative treatment till 7 years

3 A patient of 15 years complains about an additional finger of the right hand. Under the results of X-ray examination, the additional finger is full. Choose treatment tactics:

A Surgical removal of "extra" finger

B Surgical removal of "extra" finger. Skin plastics

C At the request of the patient (removal / observation)

D Removing the "extra" finger and metacarpal bone

E Functional rehabilitation

4 Patient B. 4 years old. Parents noticed the deviation of the wrist in the radial direction, progressing with age. It was established that the child had a fractured clavicle during delivery. The movements of the fingers of the hand are preserved, the weakness of flexion of the 1st and 2d fingers. Establish a diagnosis:

A Pathological fusion of the bones of the forearm

B Obsolete damage to the brachial plexus

C Elbowhandedness

D Madelung disease

E Radial manus vara

5 Child of 15, male. Splitting of the wrist to the level of the middle third of the metacarpal bones between 2nd and 3d fingers. Establish a diagnosis, suggest treatment tactics.

A Acromegaly. Skin plastic.

B Ectrodactyly. Amputation of 2nd and 3d fingers

C Syndactyly

D Ectrodactyly. Sealing the defect, skin plastic.

E Physiotherapy, massage.