

Протокол №

Кафедрального совещания на открытое занятие, проведенное преподавателя Эралиева Ж.М. на тему: «Hereditary aplastic anemia in children»

Повестка дня:

1. Обсуждение открытого занятия проведенного преподавателя Эралиева Ж.М. на тему: «Hereditary aplastic anemia in children»

Присутствовали: к.м.н., доцент, заф.каф. Бугубаева М.М., к.м.н., Осмонова Г.Ж., Алимова Н.А., Эндеш кызы Г. Абдикаримов У.А.

Открытый урок проведен в кабинете 109 здания Димедус. В начале занятия выступила Эралиева Ж.М. ознакомившись присутствующих с темой, структурой и целями занятия, дала мотивацию студентам. Далее урок проведен по хронометражу. Выбранная структура урока была рационально использована, студенты участвовали активно на всех этапах занятия и бурно обсуждали тему.

Выступила к.м.н., доцент, зав.каф. Бугубаева М.М.: «Урок соответствует теме и поставленным целям, программе и стандартам. Все поставленные задачи и РО удалось реализовать. Все этапы урока связаны между собой. Этапы занятия проведены по плану, был устный опрос в виде презентации, где студенты свободно участвовали в обсуждении, решали тестовый контроль и ситуационные задачи, а также продемонстрировали различные формы апластического анемии у ребенка.

В целом, урок прошел успешно, были даны замечания для улучшения качества преподавания, посредством непрерывного повышения их квалификации, компетентности, профессионализма.

Секретарь :

Абдикаримов У.А.



Рецензия

Актуальность : Апластическая анемия (АА) – заболевание системы крови, характеризующееся панцитопенией, обусловленной аплазией костного мозга, связанной с нарушением иммунных механизмов регуляции кроветворения, количественным дефицитом и функциональными дефектами стволовых кроветворных клеток. Успех иммуносупрессивной терапии (ИСТ) больших апластической анемией связан с воздействием на патогенетические звенья процесса угнетения функции костного мозга, которое происходит вследствие аутоиммунной атаки на клетки-предшественницы кроветворения этапах течения заболевания.

Aim of practical class: to be able to identify and make differentiation of Aplastic anemia in children, describe clinical features of Aplastic anemia in children. Prescribe treatment of Aplastic anemia in children according to the severity of the disease.

Plan of practical class:

1. Definition of Aplastic anemia in children. Terminology.
2. Clinical manifestations aplastic anemia in children.
3. Diagnosis. Treatment. Dispensary observation.

Control Questions:

1. Tell the definition of Aplastic anemia in children.
2. Tell the etiological factors, pathogenesis of Aplastic anemia in children.
3. Describe clinical features of Aplastic anemia in children.
4. Write the investigation methods of Aplastic anemia in children.
5. Differentiate Aplastic anemia in children from other types of anemia.

Открытый урок проводился в 109 кабинете здания Димедус, применялись новые интерактивные методы, такие как Димедус, ТВЛ. Были усовершенствованы все основные этапы занятия: изучение нового, закрепление изученного и контроль знаний студентов. На занятии преподаватель раздавал материалы для самостоятельного изучения, проводил тестирование.

Материал урока связан с темой урока, наблюдается логическое соответствие между темой урока и выбором заданий. Все это позволяет увеличить плотность урока и оптимально увеличить его темп. Преподаватель рассчитала время необходимое для выполнения заданий на каждом этапе, в заключении провести выводы, объяснить задание на дом, выставить оценку студентам за работу на занятии

Занятие было интересным не только для студентов, но и для посетивших преподавателей.

Зав. каф., к.м.н., доцент

Бугубаева М,М,

**MINISTRY OF EDUCATION & SCIENCE OF KYRGYZ REPUBLIC
OSH STATE UNIVERSITY
INTERNATIONAL MEDICAL FACULTY
DEPARTMENT "CLINICAL DISCIPLINE 2"**

«REVIEWED» -
In meeting of department CD 2
Prot. № 1 from 29.08 2023
Head of department, Assoc. Prof., MD
Bugubaeva M.M. _____

«Recommended by» -
Academic councilor of department
of Clinical Disciplines 2
Endesh kyzy.G.

PLAN – PRACTICAL CLASS
DISCIPLINE: "Hospital Pediatrics" (5 course)
For students studying in specialty: (560001) - «General medicine» (GM)

TOPIC №7. Hereditary aplastic anemia in children.
Etiopathogenesis, clinic, Diagnosis, Differential diagnosis, Treatment, Prevention

Osh 2023

TOPIC №7. Hereditary aplastic anemia in children. Etiopathogenesis, clinic, Diagnosis, Differential diagnosis, Treatment, Prevention

Type of class – practical.

Class time – 2 hours (90 minutes).

Plan of practical class:

1. Definition of Aplastic anemia in children. Terminology.
2. Clinical manifestations aplastic anemia in children.
3. Diagnosis. Treatment. Dispensary observation.

Control Questions:

1. Tell the definition of Aplastic anemia in children.
2. Tell the etiological factors, pathogenesis of Aplastic anemia in children.
3. Describe clinical features of Aplastic anemia in children.
4. Write the investigation methods of Aplastic anemia in children.
5. Differentiate Aplastic anemia in children from other types of anemia.

Aim of practical class: to be able to identify and make differentiation of Aplastic anemia in children, describe clinical features of Aplastic anemia in children. Prescribe treatment of Aplastic anemia in children according to the severity of the disease.

Form of class: practical (sub groups)

Type of class: practical class

Equipments used in class: markers, board, slides, posters, laptop

Interdisciplinary correlation: Pediatrics, Embryology, Anatomy, Physiology.

Topics correlation: practical classes №1,2,4,5,6,8,9.

Educational Result (ER) and competencies formulated in the process of studying the discipline "Hospital pediatrics"

During studying the discipline, the student will achieve the following Educational Result(ER) and will have the appropriate competencies:

Code of LO in GEP and its wording	Competencies of GEP	Code of LO of the discipline (LOd) and its wording
LO - Knows how to apply basic knowledge in the field of diagnostic activities to solve professional problems	PC-11 - capable and ready to make a diagnosis based on the results of biochemical and clinical studies, taking into account the course of pathology in organs, systems and the body as a whole;	LOd-1 capable and ready to make a diagnosis based on clinical manifestations and standard methods of examination of childhood
LO - Knows how to apply basic knowledge in the field of medical practice to solve professional problems	PC-15 - is able to prescribe adequate treatment to patients in accordance with the diagnosis.	LOd-2: is able to prescribe adequate modern treatment and carry out basic therapeutic measures depending on the age of the children
LO - knows how to apply basic knowledge in the field of research activities to solve professional problems	SPC-3 - capable of analyzing medical information based on the principles of evidence-based medicine PC-27 - ready to study scientific and medical information, domestic and foreign experience on the research topic	LOd-3: able to study scientific and medical literature on the subject, prescribe treatment based on the principles of evidence-based medicine

At the end of PRACTICAL class students are:

- Able to identify and make differentiation of Aplastic anemia in children in children,
- Able to describe clinical forms of a Aplastic anemia in children
- Able to prescribe treatment according to causative agent and clinical form of Aplastic anemia in children

No	Stages	Aim of Practical class	Actions of teacher	Actions of students	Methods	Results of study	Equipments used	Time
1	Organizational part	Greeting, check for absentees, check students' appearance and readiness for practical class, organizing attention, introduction with new topic and its questions	Introduction	Writing the topic & its questions	Introduction	Get attention of students for class	Board, markers	5 min.
2	Inquiring of material on prerequisites of practical class	Make general information from students knowledge about the materials studied and establish a link with a new topic.	Asking questions	Selectively answer questions one by one.	Questions-answers	Recalling materials of prerequisites, contributing to self preparation	Board, markers	10 min.
3	Motivation for new topic	Enhance students' mental activity, develop critical thinking	Demonstration of a case	Ask interested questions about topic	Show the video	Focusing students on the issues, participating in team discussions, freely express their opinions	Board, markers	5 min.
4	Discussion and interview of new topic	Giving students' knowledge on a new topic, to form the skills of the ability to use them in practical classes	Control questions	Participation in 2 different teams	Divide students into groups	A theoretical base of knowledge and skills for use in practical exercises.	Board, markers	40 min.
5	Conclusion of new topic and summing up	Definition and analysis of the material covered, making changes to its content	Give questions about topic	Participate in discussion, the ability to work in a team	Schedule a case of child with Aplastic anemia in children	Independently use the knowledge gained on the topic, forming competence	Board, markers	15 min.
6	Checking the students and their knowledge	Teach students to self-esteem	Checking knowledge of students	Give exact and correct answers	Giving marks	Make assessment of students how much knows the topic	Pen	10 min.
7	Homework	To increase quality of knowledge	Describe the homework	Sign the homework	Give text of next topic	To increase quality of knowledge	Board, markers	5 min.

Attachment №1

General Overview

Aplastic anemia is a life threatening blood disorder that affects a large number of children each year in India. Exposure to toxic agents like benzene, radiation and viruses like hepatitis A have been implicated in its etiology. The incidence in developing countries is far higher compared to developed countries. Investigating a child with pancytopenia is more complex than that of an adult as numerous inherited bone marrow failures can also present with aplastic anemia without any obvious somatic features. Precise etiological diagnosis is therefore mandatory in children before embarking on any therapy.

Aplastic anemia (AA) is a rare hematologic disease and a distinctive example of bone marrow failure syndromes. AA is characterized by diminished or absent hematopoietic precursors in the bone marrow, most often due to injury to the pluripotent stem cell. The designation "aplastic anemia" is a misnomer, because the disorder is characterized by pancytopenia rather than anemia. The disease is estimated to occur in two to four individuals per million populations every year. Paul Ehrlich introduced the concept of aplastic anemia in 1888 when he studied a case of a pregnant woman who died.

What is bone marrow?

Bone marrow is a substance found in the spongy center of bones and is where blood cells are formed. The bone marrow forms 'stem cells' which develop into any of the three types of blood cell – red blood cells, white blood cells and platelets.

Normally, the bone marrow controls the number of blood cells formed and released into the blood stream, so the body remains healthy. Too many or too few of any of the blood cells can cause problems.

The number of blood cells is often referred to as a blood count and is often separated into the different types of blood cells.

Red blood cells – about three million red blood cells are produced by the bone marrow every second. They carry a protein called 'haemoglobin' which carries oxygen to all parts of the body providing energy.

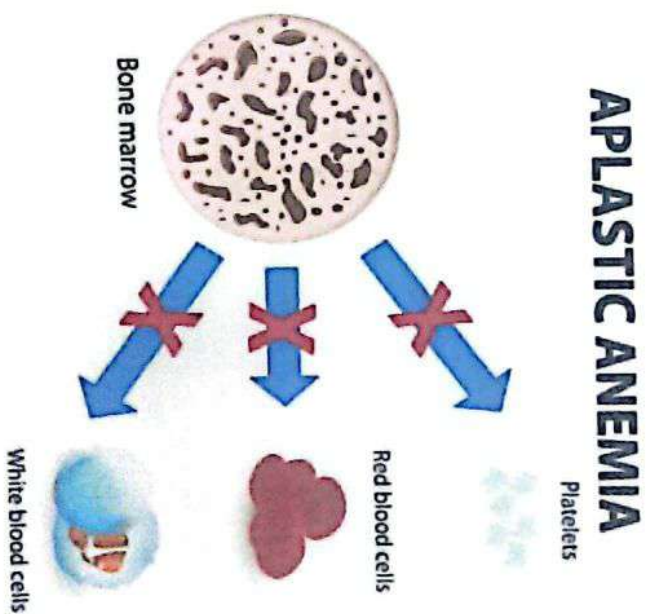
The normal level of haemoglobin in a child's blood varies with age but is around 10 to 12 grams per decilitre (g/dl). Another name for red blood cells is erythrocytes.

White blood cells – these are larger than red blood cells and have different functions.

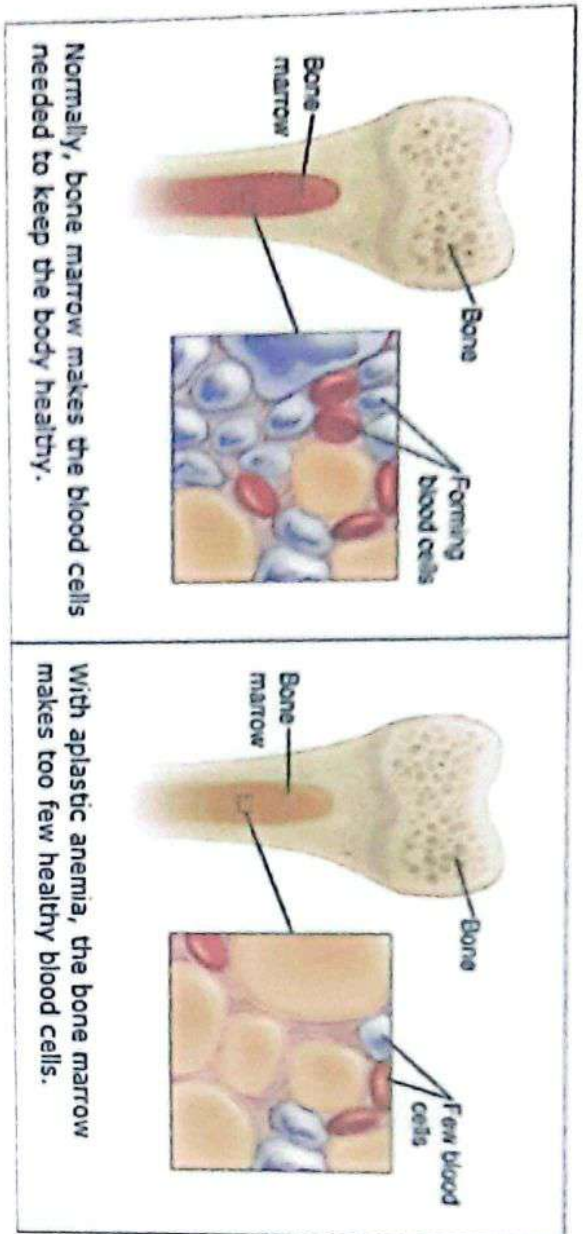
The two main white blood cells are neutrophils – which fight bacterial infections – and lymphocytes – which help fight viruses like chicken pox and measles and other non-bacterial infections.

The normal level of all white blood cells in a child's blood is about six to $16 \times 10^9/l$.

The normal level of neutrophils is between 1.5 and $8.5 \times 10^9/l$ and lymphocytes between 2 and $9.5 \times 10^9/l$.



Platelets – these are much smaller than red blood cells and help the blood clot by sticking together. The normal level of platelets is between 150 and $400 \times 10^9/L$.

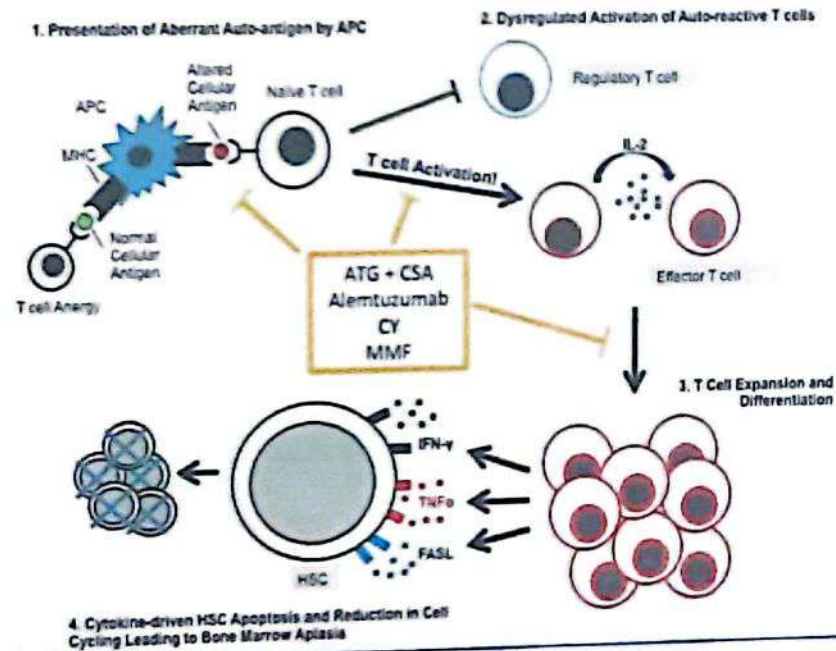


Etiology:

- Infectious Hepatitis-associated, typically seronegative
- Epstein-Barr Virus
- Cytomegalovirus
- Parvovirus
- Mycobacterial Infections
- Human Immunodeficiency Virus
- Human Herpes Virus 6
- Varicella Zoster Virus
- Measles
- Adenovirus
- Nutritional
 - Vitamin B12
 - Folic acid
- Drugs
 - Toxic Non-steroidal anti-inflammatory drugs
 - Antibiotics
 - Anticonvulsants
 - Sulfonamides
 - Gold Salts
 - Many additional agents rarely associated with aplastic anemia
 - Chloramphenicol
 - Idiosyncratic
 - Chemicals
 - Benzene
 - Insecticides
 - Pesticides
 - Solvents
 - Radiation
- Inflammatory and autoimmune (e.g. systemic lupus erythematosus)
- Graft-versus-Host-Disease

Pathogenesis:

For many years, an immune-mediated pathogenesis has been postulated for AA because Immunosuppressive Therapy (IST) is often successful in the treatment of AA and bone marrow lymphocytes from AA patients can suppress normal bone marrow *in vitro*.⁷ Results from numerous laboratories have demonstrated increased cytokine expression, low CD4 T regulatory cells, oligoclonal CD8 cytotoxic T cells, and to a lesser extent, expansion of specific CD4 cell populations in the bone marrow of AA patients. Coupled with the recent finding of acquired copy number neutral loss of heterozygosity of the short arm of chromosome 6 (6pLOH), representing a likely genetic signature of immune escape, these findings have strengthened the belief that bone marrow aplasia in acquired AA is immune-mediated, replacing the conventional term “idiopathic AA” with “immune-mediated AA.”



Clinical Presentation:

These are the most common symptoms of aplastic anemia. Symptoms may include:

- From decreased red blood cells:
 - Headache
 - Dizziness
 - Shortness of breath
- Lack of energy or tiring easily (fatigue)
 - Pale skin
 - Chest pain

- Irregular heart beat
- Enlarged heart
- From too few white blood cells:
 - Fevers
 - Mouth sores
 - Infections
- From too few platelets:
 - Easy bruising
 - Nosebleeds
 - Bleeding gums
 - Blood in the stool
 - Heavy bleeding with menstrual periods
- Other symptoms:
 - Nausea
 - Skin rashes

Diagnosis:

Features Suggestive of IBMFS in a Patient with Pancytopenia

Clinical History	<ul style="list-style-type: none"> ● Failure to thrive ● History of cytopenia, easy bruising and frequent infections ● Malabsorption/maldigestion ● Developmental Delay
Family History	<ul style="list-style-type: none"> ● Family members with cytopenias, MDS or leukemia ● Cancer of the breast, lung, esophagus, head and neck in multiple family members ● Pulmonary fibrosis, liver fibrosis, early osteoporosis ● Family members with congenital anomalies associated with IBMFS
Physical Examination	<ul style="list-style-type: none"> ● Short Stature, congenital anomalies and dysmorphologies ● Abnormal skin pigmentation, birth marks ● Nail abnormalities ● Limb (especially forearm) abnormalities ● Other skeletal abnormalities ● Renal and GU abnormalities ● Cardiac abnormalities ● Eye abnormalities ● Cleft lip/palate

	<ul style="list-style-type: none"> • Hair or teeth abnormalities • Developmental delay
	<ul style="list-style-type: none"> • Increased chromosomal breakage after exposure to cross-linking agents • Very short telomere lengths in lymphocytes • Macrocytosis • Increased fHb

Treatment:

Treatment for aplastic anemia also depends on the cause. For mild aplastic anemia, treatment may not be needed. Treatment may include:

- Blood transfusions
- Platelet transfusions
- Antibiotics
- Hormones or other medicines (to stimulate the bone marrow to produce cells)
- Immunosuppressive medicine
- Stem cell transplant

Complication:

With proper treatment, most children with aplastic anemia have no complications.

Without treatment, complications of aplastic anemia include the following:

- Medicine used to treat anemia may cause side effects
- Problems with growth and development
- Cancers
- Heart failure
- Uncontrolled bleeding
- Severe infections

Prognosis:

Untreated, severe aplastic anemia has a high risk of death. Modern treatment, by drugs or stem cell transplant, has a five-year survival rate that exceeds 85%, with younger age associated with higher survival

Survival rates for stem cell transplant vary depending on age and availability of a well-matched donor. Five-year survival rates for patients who receive transplants have been shown to be 82% for patients under age 20, 72% for those 20–40 years old, and closer to 50% for patients over age 40. Success rates are better for patients who have donors that are matched siblings and worse for patients who receive their marrow from unrelated donors.

Older people (who are generally too frail to undergo bone marrow transplants), and people who are unable to find a good bone marrow match, undergoing immune suppression have five-year survival rates of up to 75%.

Relapses are common. Relapse following ATG/ciclosporin use can sometimes be treated with a repeated course of therapy. In addition, 10-15% of severe aplastic anemia cases evolve into MDS and leukemia. According to a study, for children who underwent immunosuppressive therapy, about 15.9% of children who responded to immunosuppressive therapy encountered relapse.

Attachment №2

MCQ:

1. Unusual in aplastic anemia
 - Infection on presentation
 - Lymphadenopathy
 - Splenomegaly
 - **All of the above**
2. Severe aplastic anemia is defined by
 - Absolute neutrophil count < 500/ μ l
 - Platelet count < 20,000/ μ l
 - Absolute reticulocytes count < 60,000/ μ l
 - **All of the above**
3. Initial oral dose of cyclosporine in treatment of aplastic anemia in adults is
 - 2 mg/kg per day
 - 8 mg/kg per day
 - **12 mg/kg per day**
 - 20 mg/kg per day
4. In aplastic anemia peripheral blood smear shows all except
 - **Decreased mcv**
 - Few or absent reticulocytes
 - Normal lymphocyte number
 - Reduced platelets and granulocytes
5. Shwachman-diamond syndrome include all except
 - Pancreatic insufficiency
 - Malabsorption
 - **Eosinophilia**
 - Risk of aplastic anemia
6. Regarding fanconi's anemia all are true except
 - **Autosomal dominant disorder**
 - Short stature, café au lait spots
 - Type A is due to mutation in FANCA
 - increased risk of malignancy
7. Cyclosporine in treatment of aplastic anemia should be between
 - **150 and 200 ng / mL**
 - 250 to 400 ng / mL
 - 450 to 600 ng / mL
 - 650 to 800 ng / mL
8. most common early symptom in aplastic anemia
 - **bleeding**
 - infection
 - weight loss
 - jaundice