iJRASET
International Journal For Research in Applied Science and Engineering Technology

INTERNATIONAL JOURNAL FOR RESEARCH

IN APPLIED SCIENCE & ENGINEERING TECHNOLOGY

Volume: 10 **Issue:** VI **Month of publication:** June 2022

DOI: https://doi.org/10.22214/ijraset.2022.43831

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International Journal for Research in Applied Science & Engineering Technology (IJRASET)
ISSN: 2321-9653; IC Value: 45.98; SJ Impact Factor: 7.538
Volume 10 Issue VI June 2022- Available at www.ijraset.com

Pathophysiology of Blood Vein Thrombotitis Hemostasis Disorders

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Annotation: Primary or vascular platelet hemostasis disorders include thrombocytopenia, thrombocytopathy, and vasopathy. Hemorrhages are characteristic of the clinic of these diseases, with the appearance of hemorrhagic rashes on the skin as a result of increased permeability of the capillary wall, bleeding from the nose, metro and menorrhagia, resulting in the development of severe posthemorrhagic anemia. Hemorrhagic diathesis in this group and includes many nosologies acquired over a lifetime. Almost all disorders lead to life-threatening hemorrhages.
Keywords: vascular platelet hemostasis, thrombocytopenia, thrombocytopathy, vasopathy, hemorrhage

I. PURPOSE OF WORK

Interpretation of the theoretical basis of the etiology, pathogenesis, mechanisms of clinical symptoms of diseases caused by disorders of platelet hemostasis on the basis of modern literature

Theoretical part Disorders of primary or vascular platelet hemostasis include thrombocytopenia, thrombocytopathy, and vasopathy. Hemorrhages are characteristic of the clinic of these diseases, with the appearance of hemorrhagic rashes on the skin as a result of increased permeability of the capillary wall, bleeding from the nose, metro and menorrhagia, resulting in the development of severe posthemorrhagic anemia. Hemorrhagic diathesis in this group and includes many nosologies acquired over a lifetime.

The following types of hemostasis disorders are distinguished:

1) According to its origin: congenital and acquired
2) According to the mechanism of origin:
a) Disorders of platelet hemostasis
b) Disorders of coagulation hemostasis
3) According to the direction of changes: hypocoagulation and hypercoagulation

Hypocoagulation is a slowing of the blood clotting process, characterized by recurrent bleeding and hemorrhage. Hypocoagulation is caused by a deficiency of one or more of the factors or components of primary or secondary hemostasis .

Hypercoagulation is characterized by the formation of a local or massive thrombus.
Primary or vascular platelet hemostasis disorders are divided into 3 groups according to their causes:

- Thrombocytopenia is associated with a decrease in the number of platelets
- Thrombocytopathies are associated with qualitative changes in platelets.
- Vasopathies are associated with changes in the blood vessel wall .

Thrombocytopenia A decrease in the number of platelets in the peripheral blood from 150 G / l (Giga / liter) , Clinical signs of hypocoagulation, ie hemorrhage, are observed when the platelet count falls below 70 G / l. Thrombocytopenia is congenital and acquired.

Acquired thrombocytopenia is caused by three groups of factors:

- Deficiencies in platelet formation
- Too much, excessive breakdown of platelets
- Excessive platelet depletion (in thrombosis and DIC syndrome).

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Defects in platelet formation are accompanied by a decrease in the formation of platelets in the bone marrow. These include bone marrow damage (chemicals, leukemia under the influence of certain drugs (sulfonamides), metastasis of tumors to bone marrow, radiation , severe infections, uremia, hypothyroidism, vitamin B12 and folic acid deficiency.

Increased platelet breakdown is observed in splenomegaly due to mechanical damage to platelets. However, increased platelet breakdown is often caused by immune damage mechanisms (heterosexual and autoimmune thrombocytopenia).

Heteroimmune thrombocytopenia is more common in children. Platelet breakdown is explained by the formation of antibodies to platelets due to changes in platelet antigenic properties. The antigenic structure of platelets is altered by viruses (measles, adenovirus), haptens (quinidine, sulfonamides, rifampicin), and vaccines. Heteroimmune thrombocytopenia is of good quality and can be completely cured if the cause is eliminated.

Autoimmune thrombocytopenia is severe. It is caused by a lack of immune tolerance to platelet antigens. Example: Chronic autoimmune idiopathic thrombocytopenia (idiopathic thrombocytopenic purpura or *Verlgof disease*). The incidence of Idiopathic Thrombocytopenia in the world is 1.6-3.9 per 100 thousand population per year. Among adults and children, the prevalence ranges from 4.5 to 20 per 100,000 population. ITP does not have geographical features of distribution. Men get sick 3-4 times less often than women, in reproductive age this difference is even greater - 5-6 times. Most often, people aged 20 to 40 years (54%) get sick, in 20% of cases - aged 40 to 60 years, rarely the disease develops in patients older than 70 years (2%) and younger than 20 years The clinic of Verlhof's disease was recorded 100 years before the discovery of platelets (1735). The disease produces antibodies to glycoprotein receptors on the surface of platelets. The amount of IgG on the surface of platelets increases. Given that IgG is mainly synthesized in the spleen, **splenectomy** surgery is included in the treatment of the disease. Corticosteroids and immunosuppressants are also used.

However, to date, the disease has not been completely cured. The chronic form of the disease occurs mainly in young women.

Increased platelet breakdown is caused by antiplatelet antibodies. Immune thrombocytopenia also includes drug-induced allergic thrombocytopenia. Many antibacterial drugs (erythromycin, isoniazid, tetracycline, penicillin, streptomycin, sulfanilamides), antiepileptic drugs (carbamazepine , trimethadone), nonsteroidal anti-inflammatory drugs (aspirin, indomethacin, quinidine, diarcetamide, paracetamol), diarcetamide, diuretics (diuretics). hypoglycemic drugs (insulin, chlorpropamide), psychotropic drugs (aminazine, barbiturates, diazepam)) cause allergic-type thrombocytopenia.

Chronic lymphocytic leukemia, hyperthyroidism, Evans syndrome, thrombocytopenia are also observed in systemic lupus erythematosus.

Some thrombocytopenia develop on the basis of non-immune mechanisms. A common symptom of such thrombocytopenia is low platelet viability. The most important of them are:

Thrombocytopenia in Wiskott- Aldrich syndrome, eczema , and recurrent infections are often associated with otitis. The disease is recessively inherited from the X chromosome and is passed down from generation to generation. Boys with the disease die before the age of 6 from infections, hemorrhages, or malignant tumors.

Gram-negative sepsis. Endotoxins damage the endothelial layer of large blood vessels. Many platelets are used to regenerate the vascular endothelium, leading to thrombocytopenia .

In uremic hemolytic syndrome (Gasser's disease), thrombocytopenia is accompanied by hemolytic anemia and renal failure.

- Extracorporeal thrombocytopenia is caused by platelets adhering to a foreign surface.
- Alcoholic thrombocytopenia occurs after large amounts of ethanol.
- Thrombocytopenia in splenomegaly is caused by the accumulation of large numbers of platelets.

Thrombocytopathies - includes all hemostasis disorders that occur due to qualitative change or dysfunction of platelets.

Thrombocytopathies constitute a large group of hemorrhagic diatheses. Many microcirculatory types of hemorrhages (petechiae, ecchymosis, epistaxis, menorrhagia) are associated with thrombocytopathy.

While platelets are not functionally complete, their basic properties of adhesion and aggregation properties are impaired. The following groups of congenital (hereditary) thrombocytopathies are distinguished in the functional classification:

- Degranulation reaction. intact thrombocytopathies. Example: Glansman-Negeli thrombasthenia . In this case, due to the absence of specific glycoproteins IIb and IIIa on the surface of platelets, the interaction of platelets is lost. Aggregation is also not observed in interaction with fibrinogen. Clinical signs are petechiae and hemorrhages (bleeding from the nose, uterus).

- Degranulation reaction. thrombocytopathies with impaired autosomal recessive type. The reason is a violation of the activity of SOG (cyclooxygenase) and a decrease in the activity of platelet contact protein, the pathogenesis of the disease is the lack of aggregation of platelets in the interaction with collagen and the release of biologically active substances from the granules. The main clinical signs are in the form of petechiae and hemorrhages (bleeding from the nose, uterus).
- Thrombocytopathy with impaired accumulation and excretion of substances in the granules - Herdjmansky-Pudlak disease is transmitted from generation to generation in the autosomal recessive type. The reason is that ADF, adrenaline, serotonin, Ca^{2+} are stored in dense granules.

The disorder of assembly is characterized by the absence of aggregation in the interaction with collagen in the pathogenesis of this hypocoagulation, the inseparability of granulation retention, resulting in varying degrees of bleeding.

Thrombocytopathy with platelet adhesion and aggregation includes the following diseases:

- Willebrand Jurgens syndrome - an autosomal recessive hereditary disease - occurs due to a deficiency of Willebrand factor.
- Bernard Sule 's disease (autosomal recessive) deficiency of glycoproteins in the platelet membrane. This pathology is characterized by capillary bleeding. This can lead to life-threatening bleeding during adolescence and childbirth: menorrhagia and metrorrhagia.
- Platelet-related factor 3 deficiency - **Boue and Ovek disease**
- Thrombocytopathies associated with other inherited anomalies.

In acquired disorders of platelet adhesion, a thin layer is formed on the platelet mega receptors, and this molecular layer is a barrier wall to the receptor interaction with the Willebrand factor.

Congenital pathologies:

- in leukemias
- When the amount of IgM increases
- B12 is observed in vitamin hypovitaminosis.

Vasopathies is characterized by hemorrhagic diathesis, which occurs as a result of insufficient development of the vascular wall functionally and morphologically. There is a congenital and acquired type of this pathology.
Congenital vasopathies include Rendu-Osler disease (otherwise known as telangiectasia), Fabry disease (diffuse angiokeratoma), and hereditary thrombocytopenic microangeomatosis. Congenital hereditary vasopathies are caused by an inherited disorder of the development of the vascular subendothelial layer.
The main feature of such vessels is thinning of vessels, dilation of microtubule cavities, low collagen content in the subendothelium, rapid vascular injury, clinically observed bleeding from the nose, lungs and bronchi, gastrointestinal tract.

Acquired vasopathies include vascular pathologies in various diseases:

- Idiopathic angiopathy - characteristic of Kaposi's sarcoma (etiology unknown)
- Dymoplastic vasopathy - in Klots and Favra-Rakusho dermatitis. The cause is chronic heart failure, local venous stasis.
- Dystrophic-steroidal purpura (in treatment with corticosteroids that reduce collagen synthesis and adrenal hyperfunction)
- Scorpio (Vitamin C deficiency)
- Schonlein - Henorrhagic hemorrhagic vasculitis caused by damage to the blood vessels by the immune complex
- Neurogenic

II. CONCLUSIONS

In summary, in the pathogenesis of vascular thrombocyte hemostasis disorders are the main pathogenetic link and diseases associated with hemorrhage occur mainly in children and young people. Depending on their origin, they are hereditary and acquired. It occurs as a result of qualitative and quantitative disruption of platelets or structural disorders of the vascular wall. Today, it is these diseases that cause disability and pathological deaths

REFERENCES

[1] Gresele P; Subcommittee on Platelet Physiology of the International Society on Thrombosis and Hemostasis. Diagnosis of inherited platelet function disorders: guidance from the SSC of the ISTH. J Thromb Haemost. 2015 Feb;13(2):314-22. doi: 10.1111/jth.12792. Epub 2015 Jan 22. PMID: 25403439.

[2] Melazzini F, Zaninetti C, Balduini CL. Bleeding is not the main clinical issue in many patients with inherited thrombocytopaenias. Haemophilia. 2017 Sep;23(5):673-681. doi: 10.1111/hae.13255. Epub 2017 Jun 8. PMID: 28594466.

[3] Broos K, Feys HB, De Meyer SF, Vanhoorelbeke K, Deckmyn H. Platelets at work in primary hemostasis. Blood Rev. 2011 Jul;25(4):155-67. doi: 10.1016/j.blre.2011.03.002. Epub 2011 Apr 14. PMID: 21496978.

[4] Lippi G, Favaloro EJ. Laboratory hemostasis: from biology to the bench. Clin Chem Lab Med. 2018 Jun 27;56(7):1035-1045. doi: 10.1515/cclm-2017-1205. PMID: 29455188.

[5] Narzulaeva, U. R., Samieva, G. U., & Hasanov, F. R. THE IMPORTANCE OF A HEALTHY LIFESTYLE IN ELIMINATING RISK FACTORS IN THE EARLY STAGES OF HYPERTENSION.Assessment of neonatal platelet adhesion, activation, and aggregation / SM BakerGroberg, S. Lattimore, M. Recht, OJ McCarty, et al. // J Thromb Haemost. - 2016. - Vol. 14, № 4. - p. 815-827

[6] Poloxov, D.M. Modern approaches in the laboratory diagnosis of thrombocytic hemostasis / D.M. Poloxov, M.A. Panteleev // Hematology. Transfusiology. Eastern Europe. - 2016. - №2. - s. 270-290.

[7] Germanskogo-Pudlaka syndrome: features of differential diagnostics / Demina I.A., Zozulya N.I., Lixacheva E.A., Vasilev S.A. and dr. // Hematology and transfusiology. - 2015. - № 4. - p. 41-44.

[8] Juraeva, D. N., Narzulaeva, U. R., & Aslamovna, K. G. (2022). GENDER DIFFERENCES IN THE PARACLINICAL FEATURES OF THE COURSE OF TRIGEMINAL NEURALGIA. World Bulletin of Public Health, 8, 186-190.

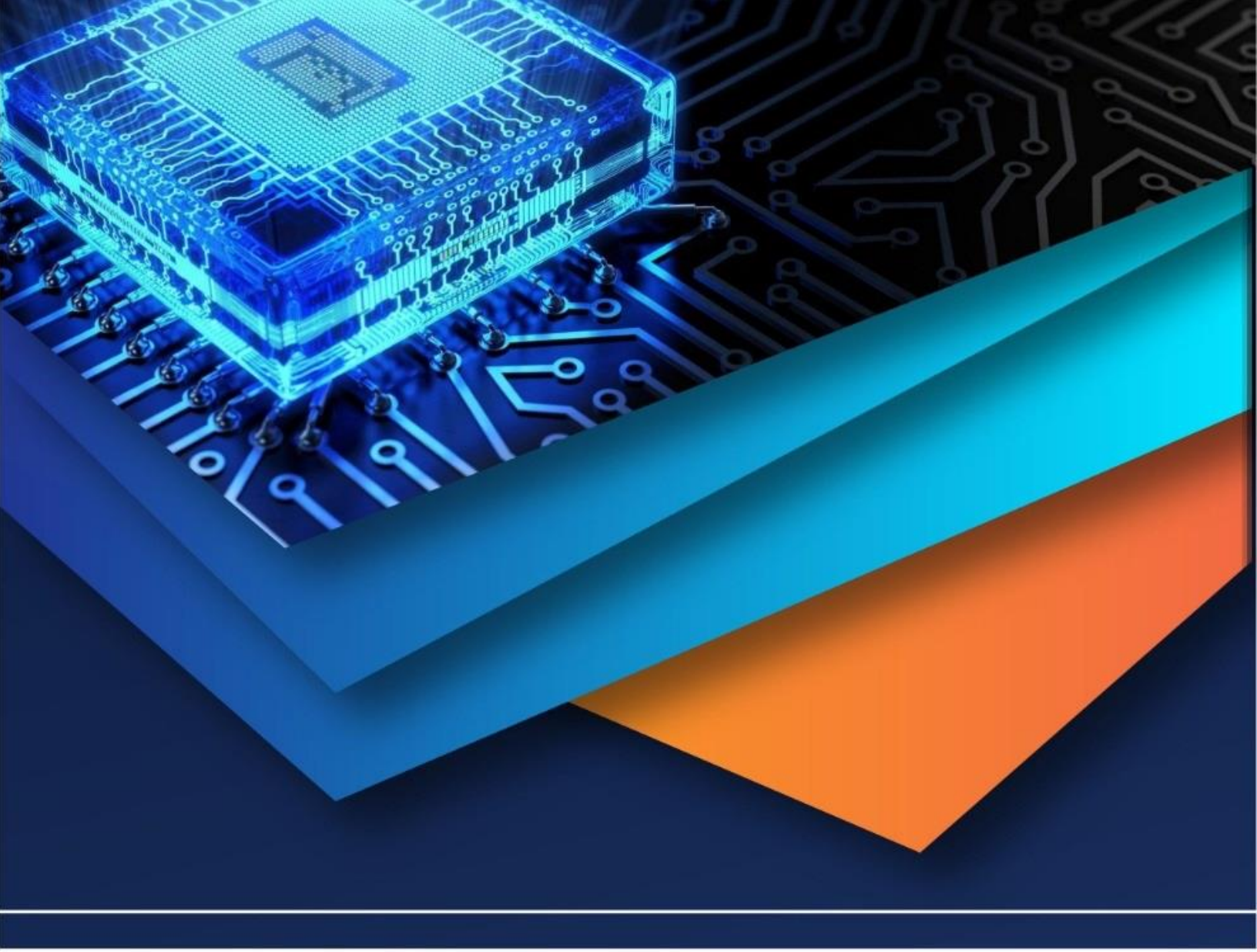
[9] Reconstructing and deconstructing agonist-induced activation of integrin alphaIIbbeta3 / J. Han, CJ Lim, N. Watanabe, A. Soriani, et al.// Curr Biol. - 2006. - Vol. 16, № 18. -p. 1796-1806.

[10] Demina, I.A. Thrombocytopathy / I.A. Demina, M.A. Kumskova, M.A. Panteleev // Rossiyskiy zhurnal detskoy hematologii i onkologii (RJDGiO). - 2015. - № 1. - p. 54-60.

[11] Нарзулаева, У. Р., Самиева, Г. У., & Насирова, Ш. Ш. (2021). ИССИҚ ИҚЛИМДА КЕЧУВЧИ ГИПЕРТОНИЯ КАСАЛЛИГИНИНГ БОШЛАНҒИЧ БОСҚИЧЛАРИДА ГЕМОРЕОЛОГИК БУЗИЛИШЛАР. ЖУРНАЛ БИОМЕДИЦИНЫ И ПРАКТИКИ, 6(1).

[12] International consensus report on the investigation and management of primary immune thrombocytopenia / D. Provan, R. Stasi, AC Newland, VS Blanchette, et al. // Blood. - 2010. - Vol. 115, № 2. - p. 168-186.

[13] Melikyan A.L., Egorova E.K., Pustovaya E.I., Kolosheinova T.I., Volodicheva E.M., Kaporskaya T.S., Ilyasov R.K., Shelekhova T.V., Fedorova N.A., Zotova I.I., Sycheva T.M., Kontievsky I.N., Shestopalova I.L., Kurkina N.V., Syrtseva E.B., Tarasenko E.V., Meresiy O.V., Cherkashenko Z.V., Chabaeva Yu.A., Kulikov S.M. INTERIM RESULTS OF EPIDEMIOLOGICAL STUDY OF IDIOPATHIC THROMBOCYTOPENIC PURPURA IN ADULTS IN THE RUSSIAN FEDERATION: INTERMEDIATE RESULTS. Russian journal of hematology and transfusiology. 2019;64(4):436–446. (In Russ.)



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