Acquired hemophilia – pathophysiology, diagnosis

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Acquired hemophilia (AH) is a rare autoimmune disorder, with a reported incidence of 1-2:1000000/year. Affected individuals are likely undiagnosed or misdiagnosed in real-world clinical practice, making it difficult to determine the real frequency of the disorder in general population. AH is characterized by potentially severe, life-threatening hemorrhages that occur in patients who lack personal and family history of bleeding. AH is predominately a disease of elderly. Less than 50% of patients have an identifiable underlying clinical condition while the others have an idiopathic form. Although both congenital and acquired hemophilia involve deficiency of the same clotting factors, the bleeding pattern and the treatment are different. The laboratory hallmark for the diagnosis of AHA is a prolonged activated partial thromboplastin time (APTT), not corrected by normal plasma (mixing test), with a normal prothrombin time (PT). Treatment, which consists of hemostatic management (controlling and preventing bleeding), eradication of the inhibitors, and treatment of the underlying disease (if apliccable), can be challenging to manage. Few data are available to guide the management of AHA-related bleeding and eradication of the disease-causing antibodies. Delayed diagnosis and concomitant medical issues are the main causes of unfavorable outcome.

Hemofilia dobandita – etiopatogenie, diagnostic

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Hemofilia dobandita este o boala autoimuna rara, cu o incidenta raportata in populatia generala de 1-2:1000000 locuitori pe an. Adeseori, diagnosticul este eronat sau ratat, ceea ce face dificila determinarea incidentei reale a bolii in populatie. Hemofilia dobandita se caracterizeaza prin hemoragii severe, potential amenintatoare de viata, ce apar la indivizi fara istoric personal sau familial de sangerare. Afecteaza cu precadere varstnicii. Mai putin de 50% din cazuri au o boala subiacenta identificabila, restul au forma idiopatica. Desi atat hemofilia congenitala cat si cea dobandita implica deficienta acelorasi factori ai coagularii, atat tipul manifestarilor hemoragice cat si tratamentul sunt diferite. Testele esentiale pentru diagnostic sunt timpul partial de trombolpastina activat (APTT) prelungit, care nu se corecteaza in amestec cu plasma normala (testul de mixing), in prezenta unui timp de protrombina (PT) normal.Tratamentul vizeaza controlul si preventia sangerarilor, eradicarea inhibitorilor si controlul bolii subiacente, cand aceasta exista, si este adeseori dificil. Ghidurile de tratament pentru aceasta afectiune rara lipsesc. Diagnosticul tardiv si comorbiditatile adeseori intalnite la aceasta grupa de pacienti sunt principalele cauze ale esecului terapeutic.