Contents

Section 1: Disorders of Red Cells

1. Anemias of Impaired Red Cell Production 3

Anemia 3
Red cell indices 4
Iron deficiency anemia 5
Megaloblastic anemia 8
Pernicious anemia 11
Aplastic anemia 13

2. Hemolytic Anemias Due to Red Cell Membrane and Enzyme Defects 16

Hemolytic anemia 16
Hereditary spherocytosis 17
Glucose-6-phosphate dehydrogenase deficiency 20

3. Thalassemia Syndrome 22

Classification of hereditary defects in hemoglobin 22
Thalassemia syndrome 22
β-thalassemia 22
β-thalassemia major 23
β-thalassemia minor/trait 27
α-thalassemia 28

4. Sickle Cell Disease 29

Sickle cell disease 29
Sickle cell anemia 29
Sickle cell trait 34

5. Other Anemias 36

Immunohemolytic anemias 36
Hemolytic disease of the newborn 36
Antiglobulin (Coombs) test 39
Autoimmune hemolytic anemia 40
Fragmentation syndrome 41
Paroxysmal nocturnal hemoglobinuria 41
Anemias of blood loss 41
Sideroblastic anemias 42

Section 2: Disorders of White Cells

6. Quantitative and Qualitative Disorders of Leukocytes 45

Normal differential leukocyte count (DLC) 45
Quantitative disorders of leukocytes 45
Qualitative disorders of leukocytes 50
Infectious mononucleosis (Glandular fever) 51

7. Acute Leukemia 52

Acute leukemia 52
Acute lymphoblastic leukemia/lymphoma 55
Acute myelogenous leukemia 57
Myeloid sarcoma 59

8. Myelodysplastic Syndromes 60

Myelodysplastic syndromes 60

9. Myeloproliferative Neoplasms 62

Myeloproliferative neoplasms (MPN) 62
Polycythemia or erythrocytosis 63
Polycythemia vera 63
Essential thrombocythemia 65
Primary myelofibrosis 66

10. Chronic Myelogenous Leukemia 68

Chronic myelogenous leukemia 68
Natural history of chronic myeloid leukemia 70

11. Chronic Lymphocytic Leukemia/Small Lymphocytic Lymphoma 73

Chronic lymphocytic leukemia 73
Hairy cell leukemia 75

12. Plasma Cell Neoplasms 76

Plasma cell myeloma (multiple myeloma) 76
Plasmacytoma 80
Immunoglobulin deposition disease 80
Monoclonal gammopathy of uncertain significance (MGUS) 80

13. Lymphoid Neoplasms 81

Classification of lymphoid neoplasms 81
Follicular lymphoma (FL) 82
Diffuse large B cell lymphoma (DLBCL) 83
Burkitt lymphoma (BL) 83
Mature T cell and NK cell neoplasms 85

14. Hodgkin Lymphomas 87

Definition 87
Classification 87
Morphology of neoplastic cells 88
Classical Hodgkin lymphoma 88
Nodular lymphocyte predominant Hodgkin lymphoma (NLPHL) 92
Etiology and pathogenesis of Hodgkin lymphoma 93

Laboratory findings 93
Staging of Hodgkin lymphoma 94
Differences between Hodgkin lymphoma and non-Hodgkin lymphoma 94

15. Langerhans Cell Histiocytosis/Histiocytosis X **95**
Morphology 95
Laboratory findings 95

Section 3: Disorders of Hemostasis

16. Disorders of Primary Hemostasis **99**
Normal hemostasis 99
Classification of hemostatic disorders 99
Bleeding disorders caused by vessel wall abnormalities 99
Bleeding disorders due to abnormalities of platelet 100
Thrombocytopenia 100
Immune thrombocytopenic purpura 102
Thrombocytosis 104
Qualitative platelet disorders 104

17. Bleeding Disorders: Due to Abnormalities of Coagulation/Clotting Factor **105**
Classification of coagulation disorders 105
Hereditary coagulation disorders 106
Hemophilia 106
Hemophilia A (Factor VIII deficiency) 106
Hemophilia B (Christmas disease, factor IX deficiency) 108
von Willebrand disease (vWD) 108
Acquired coagulation disorders 109
Disseminated intravascular coagulation 110

18. Thrombotic Disorders: Hypercoagulable State **113**
Hypercoagulable state (Thrombophilia) 113
Inherited hypercoagulable states 114
Acquired hypercoagulable states 114

Section 4: Clinical Scenario

19. Clinical Scenario **119**
Symptoms and signs that suggest a blood disease 119
Patterns strongly suggestive of a blood disease 120

Appendix ***127***
Bibliography ***133***
Index ***135***