

Table of Contents

1. Introduction, History of Aortic Diseases	9
1.1 Anatomy of the aorta	13
1.2 Embryology and histology of the aorta	23
1.3 Pathophysiology and etiology of aortic diseases	28
 2. Imaging Modalities	 35
2.1 Echocardiography	37
2.2 "Classic" Aortography with contrast material	38
2.3 Computer Tomography	39
2.4 Intravascular Ultrasound	41
2.5 Magnetic Resonance Imaging	41
 3. Aortic Aneurysm	 45
3.1 Definition and classification of aortic aneurysms	47
3.2 Pathophysiology of the aortic aneurysm	51
3.3 Etiology of the aortic aneurysm	69
3.3.1 Aneurysms from a genetic etiology	69
3.3.2 Aneurysms with positive family history (hereditary)	73
3.3.2.1 Marfan syndrome	75
3.3.2.2 Loeys-Dietz syndrome	80
3.3.2.3 Turner syndrome	85
3.3.2.4 Alagille syndrome	88
3.3.2.5 Bicuspid aortic valve	90
3.3.2.6 Ehlers-Danlos syndrome	95
3.3.2.7 Sphrintzen-Goldberg syndrome	97
3.3.2.8 Osteogenesis imperfecta	98
3.3.2.9 Beals syndrome-CCA	100
3.3.2.10 Polycystic kidney disease	101
3.3.2.11 Noonan syndrome	101

3.3.2.12	Klippel-Feil syndrome	102
3.3.2.13	Pseudoxanthoma elasticum	105
3.3.2.14	Klinefelter syndrome	106
3.3.3	Aneurysms with non-infectious inflammatory etiology	106
3.3.3.1	Giant cell arteritis – Horton’s disease	107
3.3.3.2	Takayasu arteritis	115
3.3.3.3	Behcet disease	120
3.3.3.4	Wiskott-Aldrich syndrome	122
3.3.3.5	Collagen diseases (systemic lupus, Bechterew disease)	123
3.3.3.6	Inflammatory bowel disease – ulcerative colitis	125
3.3.3.7	Cogan’s syndrome	126
3.3.3.8	Relapsing polychondritis	127
3.3.3.9	Reiter syndrome	129
3.3.3.10	Churg-Strauss syndrome	130
3.3.3.11	Sarcoidosis	132
3.3.3.12	Idiopathic isolated aortitis	133
3.3.3.13	Ormond’s disease (idiopathic retroperitoneal fibrosis)	133
3.3.4	Aneurysms in arterial hypertension and atherosclerosis	134
3.3.5	Aneurysms with infectious etiology	134
3.3.5.1	Tuberculosis	136
3.3.5.2	Salmonellosis	139
3.3.5.3	Syphilis	139
3.3.5.4	Staphylococcal infection	142
3.3.5.5	Clostridial infection	143
3.3.5.6	Streptococcal infection	143
3.3.5.7	Chagas disease	144
3.3.5.8	Acquired Immune Deficiency Syndrome	145
3.3.5.9	Other infectious agents that may cause aneurysm of the aorta	147
3.4	Clinical symptoms of aortic aneurysm	147
3.5	Aortic Aneurysm Diagnostic Methods	149
3.6	Aortic Aneurysm Treatment	149
3.6.1	Surgical Treatment of Aortic Aneurysm	150
3.6.2	Endovascular Treatment of Aortic Aneurysm	154
3.6.3	Pharmacologic Treatment of Aortic Aneurysm	156
3.7	Conclusion	156
4.	Dissection and Transection of the Aorta	159
4.1	Definition and classification of Aortic Dissection	161
4.2	Aortic Dissection Pathophysiology	162
4.3	Aortic Dissection Etiology	164

4.4 Aortic Dissection Clinical Symptomatology 166

4.5 Diagnostic modalities of Aortic Dissection 167

4.6 Aortic Dissection Treatment 170

4.7 Conclusion 175

4.8 Aortic Transection 176

 4.8.1 Definition and Clinical Symptomatology 176

 4.8.2 Diagnostic modalities of Aortic Transection 178

 4.8.3 Aortic transection treatment 178

 4.8.4 Conclusion 181

5. List of Abbreviations 183

6. References 187