

# Detailed contents

## Part 1 Introduction to adult congenital heart disease

---

<b>1 Morphology and classification</b>	<b>3</b>
Introduction	4
Physiological classification	4
Sequential segmental analysis	6
Atrial arrangement	8
<b>2 Non-invasive imaging</b>	<b>11</b>
Chest X-ray (CXR)	12
Transthoracic echocardiography (TTE)	18
Transoesophageal echo (TOE)	20
Cardiovascular magnetic resonance (CMR) imaging	24
Computed tomography (CT)	30
<b>3 Physiological testing</b>	<b>33</b>
Physiological testing	34
Types of exercise tests	36
Bruce exercise testing	37
Metabolic exercise testing	38
<b>4 Cardiac catheterization</b>	<b>41</b>
Introduction	42
Indications for catheterization	44
Precatheterization care	46
Calculations	48
<b>5 Influence of pulmonary blood flow on management and outcome</b>	<b>49</b>
Introduction	50
Pulmonary vascular development in early life	50
Cyanotic heart disease and pulmonary blood flow	52

<b>6 Cyanosis</b>	<b>57</b>
Introduction	58
Non-cardiac manifestations of cyanosis	60
Checklist for inpatients and emergencies with cyanotic heart disease	62

---

**Part 2 Specific lesions**

---

<b>7 Valve and outflow tract lesions</b>	<b>65</b>
7a Left ventricular outflow tract obstruction (LVOTO)	67
Introduction	68
Subvalvar aortic stenosis (AS)	70
Bicuspid aortic valve	72
Supravalvar AS	74
7b Left ventricular inflow lesions	75
Congenital mitral valve abnormalities	76
Cor triatriatum	78
Shone syndrome	80
7c Right ventricular outflow tract obstruction (RVOTO)	81
Introduction	82
Pulmonary valvar stenosis	82
Supravalvar pulmonary stenosis	84
Pulmonary artery stenosis	84
Double-chambered right ventricle	84
7d Ebstein anomaly	85
Definition	86
Incidence	86
Associations	86
Natural history	86
Presenting features in the adult	86
Physical signs	88

---

<b>8 Septal defects</b>	<b>91</b>
8a Atrial septal defects (ASDs)	93
Introduction	94
Ostium secundum ASD	96
Ostium primum ASD	100

Sinus venosus ASD	100
Coronary sinus defect	102
Patent foramen ovale	104
8b Ventricular septal defects (VSDs)	105
Definition and incidence	106
Cardiac associations	106
Presentation	106
Physical signs	108
Investigation	108
Repair	108
8c Atrioventricular septal defects (AVSDs)	109
Definition	110
Associations	110
Incidence and recurrence	110
Clinical presentation	112
Investigation	112
Surgical management	112
Late complications post repair of AVSD	112
<hr/>	
<b>9 The Eisenmenger syndrome</b>	<b>113</b>
Introduction	114
Natural history	114
Complications and extra-cardiac manifestations	114
Physical signs	114
Investigations	116
Management	116
<hr/>	
<b>10 Aortic lesions</b>	<b>117</b>
Coarctation of the aorta (CoA)	118
Patent ductus arteriosus (PDA)	122
Aortopulmonary window	122
Common arterial trunk/truncus arteriosus	124
Marfan syndrome	126
<hr/>	
<b>11 Venous anomalies</b>	<b>127</b>
11a Anomalies of systemic venous drainage	129
Introduction	130
Superior vena cava (SVC) anomalies	130

xxii DETAILED CONTENTS

Inferior vena cava (IVC) anomalies 132

11b Anomalies of pulmonary venous drainage 133

Total anomalous pulmonary venous drainage (TAPVD) 134

Partial anomalous pulmonary venous drainage (PAPVD) 136

Scimitar syndrome 138

---

**12 Transposition complexes 141**

12a Complete transposition of the great arteries (TGA) 143

Introduction 144

Complete TGA 146

Interatrial repair—Mustard or Senning operation 148

Arterial switch operation 150

Rastelli operation 152

12b Congenitally corrected TGA (ccTGA) 153

ccTGA—AV and VA discordance 154

---

**13 Tetralogy of Fallot and pulmonary atresia with VSD 159**

Tetralogy of Fallot 160

Natural (unoperated) or shunt-palliated history 162

Operated history—follow up after radical repair 164

Pulmonary atresia with VSD 168

---

**14 Functionally univentricular hearts and  
Fontan circulation 171**

The functionally univentricular heart 172

Staged approach to achieve definitive palliation 174

Fontan circulation 176

Hypoplastic left heart syndrome (HLHS) 186

---

**15 Rare conditions presenting in adulthood 189**

Coronary anomalies 190

Sinus of valsalva aneurysm 192

**Part 3 General management issues of adult  
congenital heart disease**

---

**16 Emergencies 197**

Introduction 198

Haemoptysis in a cyanotic patient 198

Other emergencies and pitfalls to avoid in cyanotic heart disease	200
Haemoptysis or haematemesis in patient with repaired coarctation	202
Tachyarrhythmias	204
<hr/>	
<b>17 Contraception and pregnancy</b>	<b>207</b>
Introduction	208
General principles	208
Contraception	210
Preconception	214
Pregnancy and delivery	218
Post-partum	220
<hr/>	
<b>18 Endocarditis</b>	<b>221</b>
Endocarditis prophylaxis	222
<hr/>	
<b>19 Lifestyle issues</b>	<b>227</b>
Exercise	228
Diving	230
High-altitude travel	232
Flying	232
Insurance	234
<hr/>	
<b>Appendix 1 List of operations for congenital heart disease</b>	<b>235</b>
List of operations	236
<hr/>	
<b>Appendix 2 List of syndromes</b>	<b>241</b>
List of syndromes and their associations	242
<hr/>	
<b>Glossary of terms</b>	<b>245</b>
<b>Index</b>	<b>249</b>