

# Index

- abdominal compartment syndrome, 68–9  
 abdominal insufflation  
   AVSD, 29–30  
   Fontan procedure patients, 225  
   HLHS, stage I palliation, 195  
   pulmonary hypertension and, 328–9  
   TAPVR, 175–6  
   VSD, 12–13  
 abdominal wall defects, pentalogy of  
   Cantrell, 64–5, 66, 67, 68–9  
 ABO-incompatible heart transplantation,  
   292–3  
 absent pulmonary valve syndrome (APVS).  
   *See also* tetralogy of Fallot with  
   absent pulmonary valve syndrome  
   respiratory pathology, 41, 52, 53–4, 55, 56  
   TOF with, 41, 52, 53  
   tracheobronchomalacia, 41, 53–4, 55–6  
   ventilation strategies, 53–4, 55–6  
 ACEi. *See* angiotensin converting enzyme  
   inhibitors  
 acetylcholinesterase inhibitors, post-heart  
   transplantation, 295  
 ACHD. *See* adult congenital heart disease  
 acute vasoreactivity testing (AVT), 307  
 adult congenital heart disease (ACHD)  
   anesthetic implications, 150–1, 152–3  
   atrial switch procedures in patients with,  
     146, 149–50, 156  
   echocardiography evaluation, 153–6  
   follow-up, 149–50  
 afterload, monitoring and management,  
   133–4  
 AI. *See* aortic insufficiency  
 air embolism, craniostomosis repair, 126  
 airway management  
   BDG shunt patients, 205  
   DCM, 249, 250  
   Duchenne muscular dystrophy patients,  
     244  
   lung transplantation, 287  
   Marfan syndrome, 389  
   mucopolysaccharidoses, 369–70, 371,  
     372–3  
   post-heart transplantation, 302  
   pulmonary hypertension, 312, 325, 328  
   TAPVR, 176  
   VACTERL association, 362–3, 364  
   vascular rings, 342–3  
 airway obstruction  
   APVS and, 52, 53–4, 55, 56  
   TOF/PA/MAPCAs and, 60–1  
   Alagille syndrome, 34–5, 61  
   alcohol ablation, HCM, 116  
   amiodarone, long QT syndrome, 379  
   anatomic diagnosis, cardiac  
     catheterization, 4  
   anatomic shunting, 2, 3  
   Anderson–Tawil syndrome, 376. *See also*  
     long QT syndrome  
   anemia, premature infants, 11–12  
   anesthetic risk. *See also specific lesions*  
     cardiac catheterization, 87  
     patient populations with high, 6–7, 79,  
       107, 111  
   aneurysms, coarctation of the aorta, 124  
   angina, HCM, 114  
   angiography, 5  
   angiotensin converting enzyme inhibitors  
     (ACEi), 48  
     Fontan procedure patients, 211, 222, 231  
   angiotensin receptor blockers (ARBs),  
     Marfan syndrome, 387, 388, 391  
   antibiotic prophylaxis. *See also* bacterial  
     endocarditis prophylaxis  
     heterotaxy syndrome, 173  
     VAD considerations, 274  
   anticholinergic therapy  
     DORV patients, 18  
     post-heart transplantation, 295  
   anticoagulation therapy  
     ECMO management, 266  
     Fontan procedure patients, 211, 216,  
       222, 231  
     mBT shunt patients, 18, 19  
     pulmonary hypertension, 307, 334  
     VAD use, 271–2, 274, 279, 280  
   antiemetics, long QT syndrome, 379  
   antiplatelet therapy  
     Fontan procedure patients, 211, 222, 231  
     mBT shunt patients, 18  
   aorta. *See also* coarctation of the aorta  
     TOF with overriding, 39–40  
   aortic arch. *See also* vascular rings  
     double, 338, 339–40  
     embryogenesis, 339  
     right-sided, 339, 340  
     TOF with, 41  
     VACTERL association, 360, 362  
   aortic insufficiency (AI)  
     after Ross procedure, 94–5  
     subvalvular aortic stenosis, 98–9  
   aortic root dilatation, Marfan syndrome,  
     386–7, 388  
   aortic stenosis (AS), 83, 90  
     anatomy, 90, 97–9  
     anesthetic implications  
       anesthetic risk, 87, 107, 109, 111  
       cardiac catheterization special  
         considerations, 87–8  
       ECG, 87, 100, 106, 108  
       fasting considerations, 102  
       hemodynamic management, 109  
       induction technique, 102–3, 110  
       maintenance technique, 111  
       management goals, 88, 96, 102  
       monitoring, 88, 96, 103, 110–11  
       outpatient considerations, 111  
       perioperative preparation and  
         management, 88, 96, 109  
       postoperative care, 89, 96, 103–4, 111  
       premedication, 102, 110  
       preoperative assessment, 86, 95, 101,  
         102, 108  
       procedural complications, 89, 94–5  
       subvalvular, 101–4  
       supravalvular, 107–11  
       type of anesthetic, 110  
       valvular, 94–6  
   BAV procedure and advantages, 85–6,  
     92–3  
   critical, 83, 84, 90–1  
   echocardiographic findings, 84, 87, 92,  
     100–1, 102, 108  
   epidemiology, 97  
   intervention criteria, 92  
   Konno operation, 93, 101  
   neonatal pathophysiology, 83–4, 90–1  
   Ross procedure, 93, 94–5  
   Ross–Konno procedure, 93–4, 95, 101  
   SAV procedure and advantages, 85, 86, 93  
   Shone complex, 129, 130, 131, 133–4  
   subvalvular, 90, 91, 97  
     anatomy, 97–9  
     anesthetic considerations, 101–4  
     associated lesions, 99  
     echocardiographic evaluation, 100–1,  
       102  
     electrocardiography, 100  
     interventional cardiac catheterization,  
       101  
     medical management, 101  
     natural history, 100

- subvalvular (cont.)  
 physical examination findings, 100  
 physiologic considerations, 99–100  
 surgical approaches, 101
- supravalvular, 90, 91, 105  
 anesthetic considerations, 107–11  
 surgical repair indications, 107  
 Williams syndrome association, 105–6  
 without WS, 106
- treatment options, 84, 92–3
- types, 90, 91
- valvular, 90, 91  
 anatomy, 90  
 anesthetic considerations, 94–6  
 echocardiography, 92  
 physiology, 90–2  
 surgical approaches, 92–4, 95
- aortic valve. *See also* aortic stenosis  
 unicuspid, bicuspid, or quadricuspid,  
 90, 91, 92
- aortic valvotomy, aortic stenosis, 84, 85,  
 86, 93
- aortic valvuloplasty. *See* balloon aortic  
 valvuloplasty
- aortopulmonary collaterals. *See also*  
 tetralogy of Fallot with pulmonary  
 atresia and multiple  
 aortopulmonary collaterals  
 acquired or persistent, 61  
 TOF with, 41, 57, 58, 60  
 unifocalization of, 58–60
- apnea, premature infants with, 11–12, 13
- APVS. *See* absent pulmonary valve  
 syndrome
- ARBs. *See* angiotensin receptor blockers
- arrhythmias. *See also specific arrhythmias*  
 after atrial switch procedure, 147  
 BDG shunt patients, 202  
 cardiomyopathy, 256–7  
 CCTGA, 160–1  
 Duchenne muscular dystrophy, 241  
 Ebstein anomaly, 71, 73, 74, 81  
 Fontan failure, 229, 231–2, 235  
 Fontan procedure patients, 221  
 HCM, 115, 120  
 long QT syndrome, 374, 379  
 Marfan syndrome, 390, 391  
 pacemaker or ICD placement  
 complications, 245  
 post-heart transplantation, 295, 301  
 after TOF repair, 46–7, 50–1  
 VAD complications, 273
- arrhythmogenic right ventricular dysplasia  
 (ARVD), 246–7, 252–3
- arterial line  
 aberrant subclavian artery  
 considerations for, 41  
 aortic stenosis patients, 103, 110–11  
 AVSD patients, 29  
 Ebstein anomaly patients, 73  
 HCM, 119  
 pericardial effusion, 348  
 VACTERL association, 364
- arterial switch operation (ASO), 17, 140, 141  
 anesthetic implications  
 anesthetic risk counselling, 142–3  
 induction considerations, 143–4  
 monitoring and invasive access  
 considerations, 143  
 outpatient considerations, 144  
 preoperative assessment, 142  
 ventilation strategy, 144  
 atrial switch procedure versus, 145  
 CCTGA, 161–2  
 long-term complications, 141
- ARVD. *See* arrhythmogenic right  
 ventricular dysplasia
- AS. *See* aortic stenosis
- ASD. *See* atrial septal defect
- ASO. *See* arterial switch operation
- aspirin, Fontan procedure patients, 211,  
 222, 231
- asplenia syndrome, 172
- atenolol, Marfan syndrome, 387
- atria, 2
- atrial fibrillation, HCM, 120
- atrial isomerism, 171. *See also* heterotaxy  
 syndrome  
 left, 172  
 right, 172
- atrial reentrant tachycardia, after TOF  
 repair, 46, 50–1
- atrial septal defect (ASD)  
 complete AVSD with, 25, 26  
 in parallel circulations, 4  
 primum, 20, 21, 22–3, 25, 26  
 restrictive, 25–6  
 secundum, 20  
 TGA with, 138  
 TGA with hypoplastic left ventricle, 217,  
 218  
 TOF with, 41  
 tricuspid atresia with, 207–8
- atrial septostomy, pulmonary arterial  
 hypertension, 308
- atrial switch procedure, 140  
 anesthetic implications  
 baffle evaluation, 153–6  
 cardiac catheterization procedures,  
 152–3  
 central venous line placement, 152,  
 153  
 high-risk patients, 151–2  
 preoperative assessment, 150–1  
 procedure location, 156  
 arterial switch operation versus, 145  
 atrial arrhythmias, 147  
 CCTGA, 161–2  
 echocardiography, 148, 149, 150–1,  
 153–6  
 follow-up, 149–50  
 historical background, 146  
 long-term sequelae, 147, 148, 149, 150  
 modern day performance of, 146  
 Mustard versus Senning procedure, 146,  
 147, 148  
 patient survival and functional status, 149  
 RV function, 148–9, 150–1
- atrial tachyarrhythmias, Fontan failure,  
 229, 231–2, 235
- atrialized ventricle, 71, 75, 76
- atriopulmonary Fontan, 227–8
- atrioventricular concordance, 217
- atrioventricular discordance, 158, 159
- atrioventricular septal defect (AVSD), 20.  
*See also* unbalanced common  
 atrioventricular septal defect
- anatomic defects associated with  
 subtypes, 20, 21
- anesthetic implications  
 postoperative care, 24  
 specific considerations, 23–4  
 unrepaired AVSDs, 23
- associated cardiac anomalies of AV  
 valves and LVOT, 22
- cardiac surgical considerations, 22
- characteristic AVV defect, 22
- clinical implications, 22
- complete, 16, 20–2  
 anatomic characteristics, 25, 26  
 neonatal symptoms, 27  
 physiologic issues, 25  
 pulmonary hypertension and, 315–16,  
 319–20
- echocardiographic and angiographic  
 findings, 23
- partial, 20–2
- physiologic differences between  
 subtypes, 20–2
- Rastelli classification, 26
- residual postoperative defects, 22
- transitional, 20–2
- trisomy 21 and, 22, 27, 28–9
- atrioventricular valves (AVVs). *See also*  
 mitral valve; tricuspid valve  
 AVSDs affecting, 20, 22  
 common, 25, 26, 27, 28  
 regurgitation, AVSDs with, 22–3, 25
- automatic implantable cardioverter  
 defibrillator. *See* implantable  
 cardioverter-defibrillator
- AVSD. *See* atrioventricular septal defect
- AVT. *See* acute vasoreactivity testing
- AVVs. *See* atrioventricular valves
- bacterial endocarditis prophylaxis  
 BDG shunt, 202–3  
 Ebstein anomaly, 79  
 Fontan procedure, 223, 233–4  
 guidelines, 167, 168, 202–3, 233–4  
 heart transplantation, 295  
 Marfan syndrome, 390  
 after TOF repair, 49  
 truncus arteriosus, 183
- balloon angioplasty, coarctation of the  
 aorta, 123–4

- balloon aortic valvuloplasty (BAV), 84  
 anesthetic considerations, 86–9  
 procedure, 85–6, 92–3  
 results, 89  
 SAV versus, 86, 93
- balloon atrial septostomy (BAS)  
 DORV, 15–16  
 TGA, 138, 139  
 TGA with hypoplastic left ventricle, 217, 218
- balloon valvuloplasty  
 aortic stenosis, 84  
 anesthetic considerations, 86–9  
 procedure, 85–6  
 SAV versus, 86, 93  
 pulmonary stenosis  
 anesthetic considerations, 35–8  
 complications, 37, 38  
 evaluation of, 38  
 procedure, 35, 36  
 technique, 35, 36, 85–6
- Barth syndrome, 255
- BAS. *See* balloon atrial septostomy
- BAV. *See* balloon aortic valvuloplasty
- BDG shunt. *See* bidirectional Glenn shunt
- Beck's triad, 346
- Berlin Heart EXCOR, 268–9, 270, 277  
 anesthetic implications, 271–2, 273–4, 275  
 indications, 270–1  
 outcomes, 271
- β-blockers  
 long QT syndrome, 376  
 Marfan syndrome, 387, 388, 391  
 post-heart transplantation, 295
- bidirectional Glenn (BDG) shunt  
 anesthetic implications  
 analgesic options, 205  
 cerebral perfusion pressure, 203–4  
 discharge criteria, 205–6  
 emergence considerations, 205  
 endocarditis prophylaxis, 202–3  
 hematocrit expectations, 202  
 induction method, 204  
 intraoperative management goals, 203  
 laryngeal mask airway, 205  
 monitoring, 203  
 premedication, 202  
 preoperative assessment and investigations, 201–2  
 preoperative fasting considerations, 202  
 PVR manipulation, 204  
 respiratory tract infection management, 201  
 ventilation strategy, 204–5  
 for Ebstein anomaly, 79  
 anesthetic considerations, 79, 80, 81  
 fluid management and transfusion strategy, 81  
 oxygen saturations after, 80  
 pulmonary blood flow and venous return after, 79, 80  
 ventilation strategies, 81
- endocarditis prophylaxis, 202–3  
 Fontan procedure after, 227  
 for HLHS, 191, 198  
 blood flow pattern after, 199–201  
 hemi-Fontan versus, 199, 200  
 oxygen saturation expectations, 201  
 physiologic goals, 199  
 procedure, 198, 199, 200  
 reason for staging, 198–9  
 indications, 199  
 oxygen saturations, 219  
 physiologic goals, 199  
 pulmonary vascular resistance, 219  
 respiratory infection in patients with, 80  
 for TGA with hypoplastic left ventricle, 219  
 for tricuspid atresia, 208–9
- Blalock-Taussig shunt (BTS). *See also* modified Blalock-Taussig shunt  
 occlusion, 18–19
- blood flow  
 effective, 2  
 total, 3
- blood loss  
 craniostomosis repair, 125–6, 127  
 ECMO bleeding risk, 266  
 Fontan procedure patients, 214–15  
 transfusion guidelines, 127  
 VAD bleeding risk, 274, 280
- bradycardia  
 oculocardiac reflex, 18  
 trisomy 21 patients, 29
- bronchiolitis obliterans, 283, 284–5
- bronchomalacia. *See* tracheobronchomalacia
- bronchopulmonary dysplasia, 12, 323–4, 325  
 anesthetic implications, 326–9  
 anesthetic risk, 327
- bronchoscopy, complications, 288
- bronchospasm, BDG shunt, 205
- BTS. *See* Blalock-Taussig shunt
- calcineurin inhibitors, anesthetic agent  
 interactions, 296–7
- calcium channel blockers, and pulmonary hypertension, 307, 320
- cardiac arrest  
 heart transplantation failure, 301  
 VAD complications, 273  
 Williams syndrome, 107
- cardiac catheterization. *See also specific procedures*  
 aortic stenosis, 84  
 anesthetic considerations, 86–9  
 pressure gradient measurement, 92  
 procedure, 85–6, 92–3  
 results, 89  
 SAV versus, 86, 93  
 subvalvular, 101
- AVSD, 23
- diagnostic, 3–5  
 coarctation of the aorta, 122–3
- normal data, 4, 5  
 pulmonary hypertension, 306–7, 308–9, 317, 318, 319, 324–5, 334  
 pulmonary stenosis, 33  
 TAPVR, 174  
 TOF, 42–3  
 Williams syndrome, 108
- endotracheal intubation, 302
- interventional, 3–4  
 aortic valvuloplasty, 84, 85–9  
 after atrial switch procedure, 152–3  
 balloon angioplasty, 123–4  
 coarctation of the aorta, 123–4  
 HCM, 116  
 pulmonary valvuloplasty, 35–8  
 stent placement, 123–4
- laboratory special considerations, 87–8
- pulmonary stenosis, 33  
 anesthetic considerations, 35–8  
 complications, 37, 38  
 evaluation of, 38  
 procedure, 35, 36  
 surgical procedures for unsuccessful, 38
- TOF/PA/MAPCAs, 61
- cardiac index (CI), 4, 5
- cardiac magnetic resonance imaging (CMRI), 6  
 anesthetic techniques, 356  
 Duchenne muscular dystrophy, 241  
 Ebstein anomaly, 72–3  
 HCM, 115, 116–17  
 Kawasaki disease, 354–5  
 monitoring, 356–7  
 TOF, 49  
 Williams syndrome, 108
- cardiac output (CO)  
 after BDG shunt, 80  
 cardiac catheterization data on, 4, 5  
 Fontan procedure patients, 221, 224, 226  
 HLHS, 192, 193, 194–5, 196  
 pulmonary hypertensive crisis treatment, 310–11
- cardiac pressures  
 cardiac catheterization data, 4, 5  
 echocardiography data, 6
- cardiac tamponade  
 definition and hemodynamic consequences, 345–6  
 diagnosis, 346  
 echocardiographic findings, 346
- cardiac transplantation. *See* heart transplantation
- cardiomyopathy, 252, 259–60. *See also* dilated cardiomyopathy; hypertrophic cardiomyopathy; restrictive cardiomyopathy  
 anesthetic implications  
 distraction techniques, 258, 259  
 intraoperative goals and plan, 257, 258  
 preoperative evaluation, 256–7

- cardiomyopathy (cont.)  
 arrhythmogenic right ventricular dysplasia, 246–7, 252–3  
 definition, 246  
 heart transplantation, 290, 299  
 incidence, 253  
 left ventricular noncompaction, 246–7, 252–3, 255  
 pathophysiology, 258  
 subtypes, 246–7, 252–3
- cardiopulmonary resuscitation (CPR), Fontan procedure patients, 235
- cardiovascular implantable electronic devices (CIEDs). *See also* implantable cardioverter-defibrillator; pacemaker  
 defibrillation or cardioversion considerations, 384  
 definition, 377  
 dependence, 381  
 electromagnetic interference, 164–5, 232, 382  
 interrogation, 381–2  
 long QT syndrome therapy, 377  
 magnet use, 165, 166–7, 383–4  
 perioperative management, 164–5, 166–7, 381  
 preoperative evaluation, 381, 382  
 rate adaptive functions, 383  
 reprogramming, 166–7, 382–3
- catheterization. *See* cardiac catheterization
- Catheterization Risk Score for Pediatrics (CRISP), 87
- CAV. *See* coronary allograft vasculopathy
- CAVSD. *See* complete atrioventricular septal defect
- CCTGA. *See* congenitally corrected transposition of the great arteries
- cefazolin, bacterial endocarditis prophylaxis, 183
- central line  
 aortic stenosis, 96  
 atrial switch procedure patients, 152, 153  
 AVSD, 29  
 BDG shunt patients, 203  
 Fontan procedure patients, 212–13, 223, 233  
 pericardial effusion, 348  
 VAD considerations, 274
- central venous pressure (CVP), Fontan procedure patients, 209–10, 219, 223
- CentriMag, 268, 278, 279
- cerebral autoregulation, 332, 333
- cerebral perfusion, 333  
 anesthetic agent effects, 336  
 SCPA effects, 203–4
- CHARGE syndrome, 48
- CHD. *See* congenital heart disease
- chest radiography  
 aortic stenosis, 87  
 TOF, 42–3
- TOF/APVS, 54
- CHF. *See* congestive heart failure
- chronic allograft dysfunction (CLAD), 284–5
- CI. *See* cardiac index
- CIEDs. *See* cardiovascular implantable electronic devices
- circulation  
 parallel, 3, 4  
 series, 3, 4
- CLAD. *See* chronic allograft dysfunction
- CMRI. *See* cardiac magnetic resonance imaging
- CO. *See* cardiac output
- coarctation of the aorta, 121  
 anatomy and physiology, 121, 122  
 anesthetic implications  
 hemodynamic goals, 125  
 induction and maintenance strategies, 126, 127  
 monitoring, 126  
 postoperative recovery, 127  
 preoperative evaluation, 125  
 residual coarctation, 126, 127  
 transfusion issues, 127
- associated cardiac defects, 121
- balloon angioplasty, 123–4
- critical, 123
- diagnosis, 122
- echocardiographic and catheterization data, 122–3
- failed repair, 332–3
- hypertension, 122, 124, 125, 127
- recurrence, 124, 125, 332–3
- repair, 123–4
- Shone complex, 121, 129, 130, 131
- surgical approaches, 123–4
- complete atrioventricular septal defect (CAVSD), 16, 20–2  
 anatomic characteristics, 25, 26  
 neonatal symptoms, 27  
 physiologic issues, 25  
 pulmonary hypertension and, 315–16, 319–20  
 Rastelli classification, 26
- complete heart block  
 CCTGA, 160–1  
 Fontan failure, 232
- complex shunts, 3
- computed tomography (CT), 6  
 Marfan syndrome, 388  
 TOF/APVS, 54  
 Williams syndrome, 108
- computed tomography angiography (CTA), TOF/PA/MAPCAs, 61–2
- conduction system anomalies. *See also* arrhythmias  
 CCTGA, 160–1  
 heterotaxy syndrome, 169, 174  
 congenital diaphragmatic hernia. *See* diaphragmatic hernia
- congenital heart disease (CHD). *See also* adult congenital heart disease; *specific lesions*  
 heart failure onset and, 228  
 heart transplantation, 290, 299  
 NSQIP-P severity classification, 166  
 outcomes, 361  
 pathophysiologic variability, 2  
 physiologic approach, 2–3  
 preoperative analysis, 2  
 basic concepts and terminology, 2–3, 4  
 high-risk patient populations, 6–7, 79, 107  
 imaging, 3–6  
 pulmonary hypertension and, 314, 315–16  
 anesthetic risk, 316, 318–19  
 segmental approach, 2  
 trisomy 21, 27
- congenital rubella syndrome, 34–5
- congenitally corrected transposition of the great arteries (CCTGA), 137–8, 158  
 anatomic characteristics and variations, 158–60  
 anesthetic implications  
 bacterial endocarditis prophylaxis, 167, 168  
 discharge, 167–8  
 induction and maintenance methods, 167  
 open versus laparoscopic approach, 166  
 perioperative considerations, 166–7  
 postoperative care, 167
- associated cardiac anomalies, 161
- blood flow, 159, 160
- clinical presentation, 160–1
- conduction system, 160–1
- coronary artery anatomy, 160
- embryologic development, 158–9
- heart failure assessment, 163–4
- long-term sequelae of uncorrected, 162–3
- nomenclature, 159–60
- PA banding indications, 163
- pacemaker/implanted device  
 management  
 dependency determination and precautions, 164  
 electromagnetic interference, 164–5  
 magnet use, 165, 166–7  
 perioperative management, 164–5, 166–7  
 preoperative evaluation, 164, 165  
 reprogramming, 166–7
- surgical interventions, 161–2  
 double switch procedure, 161–2, 163  
 long-term sequelae, 163  
 Mustard procedure, 161–2  
 physiologic repair, 162, 163  
 physiologic versus anatomic repair, 163  
 Senning procedure, 161–2  
 Senning-Rastelli procedure, 162, 163

- single ventricle palliation, 162
  - congestive heart failure (CHF)
    - AVSDs, 22, 25, 27
    - CCTGA, 162–4
    - DCM, 246, 247
    - DORV, 15–16, 17
    - signs and symptoms, 10
    - VSDs, 10, 12, 13
  - coronary allograft vasculopathy (CAV), 291, 292
  - coronary arteries
    - anomalies
      - DORV, 14–15
      - Kawasaki disease, 353
      - truncus arteriosus, 180, 181
      - Williams syndrome, 105–6, 107, 108, 111
    - CCTGA anatomy, 160
  - corticosteroids
    - anesthetic agent interactions, 296–7
    - stress dose, 296
  - CPR. *See* cardiopulmonary resuscitation
  - craniosynostosis
    - hemodynamic goals, 125
    - induction and maintenance anesthetic agents and strategies, 126, 127
    - inotropic or vasoactive infusions, 127
    - intraoperative concerns, 125–6
    - monitoring, 126
    - pathophysiology, 124
    - postoperative recovery, 127
    - preoperative evaluation, 125
    - preoperative patient communication, 125
    - repair, 125–6, 127
    - transfusion issues, 127
  - CRISP. *See* Catheterization Risk Score for Pediatrics
  - CT. *See* computed tomography
  - CTA. *See* computed tomography angiography
  - CVP. *See* central venous pressure
  - cyanosis. *See also* Tet spells
    - aortic stenosis, 90–1
    - DORV, 15–16, 17–19
    - Ebstein anomaly, 71, 75–6
    - Fontan failure, 229
    - laparoscopy considerations, 195
    - VAD complications, 273
  - cyclosporine, 296–7
  - DA. *See* ductus arteriosus
  - DAA. *See* double aortic arch
  - DCM. *See* dilated cardiomyopathy
  - dependent (nonrestrictive) shunts, 3
  - dexmedetomidine, 63
    - BDG shunt patients, 81
    - DORV patients, 17–18, 19
    - HLHS patients, 194
    - long QT syndrome, 378
    - pulmonary hypertension effects, 312, 321
    - PVR and cerebral perfusion effects, 336
  - after TOF repair, 49
  - dextro (D)-transposition of the great arteries (D-TGA), 137–8, 145. *See also* transposition of the great arteries
    - hypoplastic left ventricle with, 217–18, 219–21
    - subtypes, 138–9
  - dextrocardia, 159–60
  - diaphragmatic hernia
    - associated anomalies, 262
    - classification, 261
    - ECMO role in management, 262
    - outcomes, 261
    - physiologic implications and prognostic indicators, 261
    - pulmonary hypertension, 261
    - surgical repair, 262
    - ventilation strategy, 262
  - differential cyanosis, aortic stenosis, 90–1
  - DiGeorge syndrome, 14–15
    - anesthetic implications, 48
    - TOF and, 48
    - truncus arteriosus with, 181, 182
  - dilated cardiomyopathy (DCM), 246, 252–3
    - anesthetic implications
      - induction and maintenance approaches, 249, 250
      - intraoperative hemodynamic management, 249, 250
      - postoperative disposition, 250
      - preoperative evaluation, 248–9
    - anesthetic risk, 249, 250
    - clinical presentation, 247
    - definition, 246
    - diagnostic modalities, 248
    - Duchenne muscular dystrophy, 240
    - hemodynamic considerations, 248, 249, 250
    - incidence, 253
    - mechanical circulatory support, 248
    - other cardiomyopathy phenotypes versus, 246–7
    - pathophysiology, 246, 258
    - pharmacologic therapies, 248
    - physical exam findings, 247
    - prognosis, 247, 248
  - disordered automaticity, Duchenne muscular dystrophy, 241
  - diuretic therapy
    - perioperative issues, 12, 176, 231
    - pulmonary arterial hypertension, 307
  - DMD. *See* Duchenne muscular dystrophy
  - DORV. *See* double-outlet right ventricle
  - double aortic arch (DAA), 338, 339–40
  - double discordance, 159, 160, 163. *See also* congenitally corrected transposition of the great arteries
  - double switch procedure, 161–2
  - double-outlet right ventricle (DORV), 14
    - anatomy, 14, 15, 16
  - anesthetic implications
    - intraoperative considerations, 18–19
    - mBT shunt precautions, 18
    - perioperative concerns and risks, 17
    - perioperative monitoring, 18
    - postoperative recovery concerns, 19
    - premedication, 17–18
    - preoperative assessment, 17
    - specific anesthetic considerations, 18
    - sudden oxygenation and hemodynamics changes, 19
  - associated cardiac defects, 14–15
  - clinical presentation, 14, 15–16, 17
  - incidence of, 14
  - physiologic subtypes, 17
    - TGA-type, 15–16, 17
    - TOF-type, 15–16, 17
    - VSD-type, 15–16, 17
  - surgical treatment options, 17
  - VSD relationship to great arteries, 14, 15, 16
- doubly committed ventricular septal defect, 15–16
- Down syndrome
  - anesthetic concerns, 29
  - associated anomalies, 27
  - AVSDs with, 22, 27, 28–9
  - comorbidities, 319, 320
  - DORV with, 14–15
  - pulmonary hypertension and, 28–9, 314, 315–16, 319–20
  - TOF with, 41, 47–8
- d-TGA. *See* dextro (D)-transposition of the great arteries
- D-transposition of the great arteries. *See* dextro (D)-transposition of the great arteries
- Duchenne muscular dystrophy (DMD), 239
  - anesthetic implications
    - complications of pacemaker or ICD placement, 245
    - ICD insertion considerations, 242–3
    - induction and maintenance considerations, 243, 244
    - intraoperative hemodynamic changes, 244
    - intraoperative respiratory management, 244
    - intubation concerns, 244
    - malignant hyperthermia risk, 242
    - neuromuscular blocking drug use, 242, 244
    - postoperative care, 245
    - preoperative discussion, 242
    - preoperative evaluation, 241, 242
    - vascular access and monitoring, 243
  - anesthetic risk, 241, 242–3, 245
  - cardiac implications, 240, 241
  - DCM, 240
  - gastrointestinal implications, 242
  - medical therapy, 240
  - pathophysiology underlying, 239

- Duchenne muscular dystrophy (cont.)  
 perioperative medication  
   considerations, 240  
 pulmonary function, 240, 241, 242  
 symptom presentation and progression, 239
- ductal stent placement, 366
- ductal-dependent physiology  
 aortic stenosis, 83, 84  
 HLHS, 187–8
- ductus arteriosus (DA)  
 closure of, 33  
 PGE<sub>1</sub> reopening of, 35
- dural ectasia, Marfan syndrome, 389
- dynamic obstructions, 3  
 RVOTO, 40
- dyspnea, HCM, 114
- Ebstein anomaly (EA), 70, 75  
 anatomic and clinical presentations, 70, 71, 75–6  
 anesthetic implications  
   anesthetic and surgical risk, 79  
   arrhythmia risk, 81  
   arterial line monitoring, 73  
   BDG shunt physiology, 80  
   emergence and postoperative concerns, 81  
   endocarditis prophylaxis, 79  
   fluid management and transfusion strategy, 81  
   imaging review, 72–3  
   induction concerns, 73  
   management considerations, 74  
   mechanical ventilation concerns, 73–4  
   noncardiac surgery after BDG shunt, 80, 81  
   outpatient considerations, 82  
   oxygen saturations after BDG shunt, 80  
   post-anesthesia ECG abnormalities, 74  
   postoperative admittance, 74  
   premedication, 73  
   preoperative evaluation, 72, 79  
   recovery location, 81  
   respiratory infection considerations, 80  
   ventilation strategy, 73–4, 81  
 arrhythmias, 71, 73, 74, 81  
 associated defects, 70  
 BDG shunt, 79  
 cyanosis, 71, 75–6  
 inability to wean from PGE<sub>1</sub>, 77–8  
 long-term strategy, 78–9  
 medical management and decision making for newborns, 77  
 natural course and survival, 76  
 prevalence, 71  
 pulmonary atresia, 71, 72, 75–6, 77–8  
 pulmonary blood flow, 75–6, 77, 81  
   after BDG shunt, 79, 80
- PVR, 75–6, 77, 81  
 surgical approaches, 77–8  
   for inability to wean from PGE<sub>1</sub>, 77–8  
   indications, 77  
   one-and-a-half-ventricle palliation, 72, 77  
   single ventricle palliation, 72, 77, 78–9  
   Starnes procedure, 72, 77, 78–9  
   two-ventricle repair, 72, 77  
 symptomatology, 71
- EC. *See* ectopia cordis
- ECG. *See* electrocardiogram
- echocardiography, 5–6  
 aortic stenosis, 84, 87, 92, 100–1, 102, 108  
 atrial switch procedure, 148, 149, 150–1, 153–6  
 AVSDs, 23, 29  
 cardiac tamponade, 346  
 coarctation of the aorta, 122–3  
 DCM, 248  
 Duchenne muscular dystrophy, 241  
 Ebstein anomaly, 72–3  
 Fontan procedure patients, 223  
 HCM, 115, 116–17  
 HLHS, 192  
 Kawasaki disease, 354  
 Marfan syndrome, 388, 389  
 pulmonary hypertension, 306–7, 309, 316–17, 324  
 pulmonic stenosis, 33, 35–6  
 TAPVR, 174  
 TOF, 42–3, 49  
 TOF/PA/MAPCAs, 61–2  
 VACTERL association, 362
- ECMO. *See* extracorporeal membrane oxygenation
- ectopia cordis (EC), 65, 66, 67
- EF. *See* ejection fraction
- effective blood flow, 2
- Eisenmenger syndrome, 10, 11, 20–2
- ejection fraction (EF), 4, 6
- elastin deficiency, Williams syndrome, 105
- electrical alternans, 346
- electrocardiogram (ECG)  
 aortic stenosis, 87, 100, 106, 108  
 BDG shunt, 202  
 Duchenne muscular dystrophy, 241  
 Ebstein anomaly, 71, 74  
 heterotaxy syndrome, 174  
 Marfan syndrome, 388  
 pericardial effusion, 346  
 post-heart transplantation, 295  
 TOF, 42–3, 49
- electromagnetic interference (EMI), 164–5, 232, 382
- endocardial cushion defects, 20, 25. *See also* atrioventricular septal defect
- endocarditis prophylaxis  
 Ebstein anomaly, 79  
 after TOF repair, 49
- endothelin receptor antagonists, 307–8, 334
- end-tidal CO<sub>2</sub> (ETCO<sub>2</sub>), 88
- epinephrine  
 HCM, 119  
   intraoperative hypotension, 30
- esmolol, for tet spell management, 43–4
- esophageal atresia  
 classifications, 359–60  
 diagnosis, 362  
 incidence, 359–60  
 surgical repair  
   airway and ventilation management, 362–3, 364  
   cardiorespiratory compromise, 364  
   complications, 365  
   ductal stent placement, 366  
   extubation, 365  
   hypoxemia management, 365–6  
   induction and intubation technique, 363  
   monitoring, 364  
   open TEF repair, 364–5  
   outcomes, 361  
   postoperative care, 365  
   preoperative bronchoscopy, 363  
   preoperative workup, 362  
   thoroscopic, 364
- esophageal foreign bodies, 73
- ETCO<sub>2</sub>. *See* end-tidal CO<sub>2</sub>
- extracorporeal membrane oxygenation (ECMO), 261  
 anesthetic implications  
   coagulopathy, bleeding, and anticoagulation management, 266  
 complications, 266  
 intensive care unit surgery, 267  
 intraoperative hemodynamic instability, 266–7  
 intraoperative monitoring, 265, 267  
 management plan, 265  
 personnel, 267  
 preoperative assessment, 265  
 anticoagulation management, 266  
 circuit components, 263, 264  
 definition, 262  
 diaphragmatic hernia management, 262  
 heart transplantation failure, 301  
 HLHS, 192, 196, 197  
 monitoring, 265, 267  
 VAD versus, 264, 276  
 veno-arterial, 263, 264, 266–7  
 veno-venous, 263, 264, 266–7, 281–2  
 ventilator management, 264–5
- fibromuscular ridge, subvalvular aortic stenosis, 97–8
- fixed obstructions, 3  
 RVOTO, 40
- fluid management  
 Ebstein anomaly, 81  
 Fontan failure, 234  
 lung transplantation, 288  
 pulmonary hypertension, 313  
 pulmonary stenosis considerations for, 37

- repaired TOF considerations for, 50
- TAPVR, 175
- Fontan procedure, 207, 217
- anesthetic implications
  - bacterial endocarditis prophylaxis, 223, 233–4
  - blood loss and transfusion, 214–15
  - discharge disposition, 236
  - emergent surgery considerations, 235–6
  - extubation, 216
  - fasting instructions, 221–2
  - hemodynamic goals, 234
  - hypoxemia management, 214
  - induction method, 211, 224, 232–3
  - intraoperative fluid management, 212
  - intraoperative hypotension management, 234, 235
  - intraoperative temperature management, 213
  - laparoscopic surgery considerations, 225
  - long-term prognosis, 225
  - maintenance considerations, 224, 232–3
  - management general principles, 213
  - management goals, 224
  - monitoring, 212–13, 223, 233
  - neurologic compromise, 215
  - outpatient considerations, 223, 236
  - pacemaker and ICD management, 232
  - perioperative analgesia, 215–16
  - perioperative medication management, 211, 222, 231
  - postoperative care, 236
  - postoperative complications, 216
  - postoperative disposition, 225
  - premedication, 211
  - preoperative assessment, 210–11, 222, 223, 230, 231–2
  - prone positioning, 213–14
  - pulmonary artery pressures management, 225
  - regional anesthesia feasibility, 224–5
  - ventilatory management, 212, 224, 234
- atriopulmonary, 227–8
- candidates for, 227
- circulation determinants, 209–10
- circulation physiology, 209–10, 219, 223, 226
- for Ebstein anomaly, 77, 78–9
- extracardiac, 199, 209, 219–20, 227, 228
- failure, 226, 236
  - anesthetic options, 232
  - atrial tachyarrhythmias, 229, 231–2, 235
  - clinical status assessment, 230, 231
  - diagnosis, 232
  - heart transplantation, 236
  - hemodynamic management, 234, 235, 236
  - onset, 228
  - protein-losing enteropathy, 221, 229
  - symptoms and clinical manifestations, 228–30
- fenestration, 220–1, 227, 230
- for HLHS, 191, 198–9
- lateral tunnel, 199, 209, 219–20, 227, 228
- long-term complications, 221
- oxygen saturations, 220–1, 229, 230
- pulmonary vascular resistance, 221
- scoliosis surgery and, 210
- survival, 209–10
- for TGA with hypoplastic left ventricle, 217–18, 219–21
- total cavopulmonary anastomosis, 219, 227–8
- for tricuspid atresia, 209
- foreign bodies, esophageal, 73
- furosemide, perioperative issues for patients on, 12
- glycosaminoglycans, 367
- graft dysfunction. *See* primary graft dysfunction
- great arteries, 2
  - VSD relationships to, 14, 15, 16
- Haller index, 387–8
- HCM. *See* hypertrophic cardiomyopathy
- heart failure (HF). *See also* congestive heart failure
  - CCTGA, 162–4
  - Fontan procedure, onset, 228
  - ketamine effects, 249, 250
  - mechanical circulatory support for, 270–1, 276
  - post-heart transplantation, 294
  - truncus arteriosus, 180, 181
- heart transplantation, 290
  - ABO compatibility, 292–3
  - anesthetic implications
    - airway management, 302
    - anesthetic risk counselling, 295
    - bacterial endocarditis prophylaxis, 295
    - cardiac dysrhythmias, 295, 301
    - cardiac medication effectiveness, 295
    - ECG findings, 295
    - fasting considerations, 295
    - immunosuppression considerations, 295–7
    - induction and maintenance methods, 297, 301–2
    - intraoperative complications, 302–3
    - monitoring, 297, 301
    - postoperative concerns, 298
    - postoperative recovery, 303
    - preoperative evaluation, 294, 300–1
    - preoperative preparation, 301
    - same-day surgery considerations, 297–8
    - special drug and equipment considerations, 297
    - stress dose steroids, 296
- anesthetic risk, 295, 301
- complications, 291, 292, 299–300
- epidemiology, 299
- for Fontan failure, 236
- heart failure, 294, 299
  - anesthetic management, 300–3
  - anesthetic risks, 301
  - ECMO, 301
- immunosuppression, 295–6
  - anesthetic agent interactions, 296–7
- indications, 290, 299
- outcomes, 291, 299
- physiology, 293
- surgical techniques, 290–1, 292, 293
- HeartMate 3, 268
- HeartMate II, 268, 277
- HeartWare, 268, 277, 278
- hematocrit
  - BDG shunt patients, 202
  - Fontan procedure patients, 214
  - TOF, 42–3
- hemi-atrial switch procedure, 146
- hemi-Fontan procedure
  - Fontan procedure after, 227
  - for HLHS, 191, 198, 199, 200
  - for tricuspid atresia, 208–9
- hepatorenal dysfunction, Fontan failure, 229
- heterotaxy syndrome, 146, 169
- anesthetic implications
  - induction and maintenance methods, 174–5
  - intraoperative concerns, 175–6
  - malrotation management, 173
  - monitoring, 174
  - postoperative management, 176
  - preoperative evaluation, 174
  - risk discussion, 173–4
  - ventilation strategy, 175
- conduction anomalies, 169, 174
- postoperative sequelae, 171
- TAPVR with, 169–70, 171
  - classification, 172
  - infection-related precautions, 173
  - organ involvement, 172
  - perioperative outcomes, 171
  - visceral situs in, 2
- HF. *See* heart failure
- HLHS. *See* hypoplastic left heart syndrome
- HOCM. *See* hypertrophic obstructive cardiomyopathy
- Hurler syndrome, 254, 367
- anesthetic implications
  - aerodigestive evaluation considerations, 372
  - airway management, 369–70, 371, 372–3
  - extubation and postoperative management, 373
  - induction and maintenance methods, 372
  - intraoperative neuromonitoring, 371–2
  - laryngospasm management, 373

- Hurler syndrome (cont.)  
 mitral regurgitation, 371  
 preoperative assessment, 369  
 procedure location, 372  
 associated cardiac defects, 369  
 characteristic features, 368  
 classification, 367, 368  
 glycosaminoglycans, 367  
 pathogenesis, 367–8  
 treatment, 369  
 variants, 368
- hybrid stage I palliation, 190, 191. *See also* stage I palliation
- hypercyanotic spells. *See* tet spells
- hypertension  
 coarctation of the aorta, 122, 124, 125, 127  
 HCM, 120  
 hypoxemia with, 195  
 Marfan syndrome, 389, 391
- hypertrophic cardiomyopathy (HCM), 113, 114, 246–7, 252–3, 258  
 anatomic manifestations, 253–4  
 anesthetic implications  
 atrial fibrillation management, 120  
 emergence, 118  
 hypertension management, 120  
 induction technique, 117–18, 119  
 maintenance technique, 118  
 management goals, 117, 119, 257  
 monitoring, 117, 119  
 perioperative management, 117, 118, 119  
 post-induction hypotension, 119  
 postoperative care, 118  
 postoperative hemorrhage  
 management, 118, 119, 120  
 potential complications, 116  
 preoperative evaluation, 116–17  
 procedure venue, 117  
 ventilation strategy, 120  
 anesthetic management, 117–18  
 characteristics, 113  
 clinical presentation, 254  
 diagnostic imaging, 115  
 genetics, 113, 114  
 incidence, 253  
 medical therapy, 116, 254  
 myocardium effects, 114  
 obstructive cardiomyopathy, 115–16  
 primary, 253  
 secondary, 253  
 surgical approaches, 116, 254  
 symptoms, 114, 115
- hypertrophic obstructive cardiomyopathy (HOCM), hypertrophic cardiomyopathy versus, 115–16
- hypoalbuminemia, Fontan failure, 229
- hypoplastic left heart syndrome (HLHS), 187, 198. *See also* stage I palliation; stage II palliation  
 anatomy, 187, 198  
 anesthetic risk, 192, 193  
 blood flow, 187–8  
 stage I palliation, 190  
 stage II palliation, 199–201  
 hemodynamic goals, 191, 192, 193–4  
 incidence, 187  
 Q<sub>p</sub>:Q<sub>s</sub> ratio, 191, 192, 193–5  
 surgical approaches  
 Fontan procedure, 191, 198, 199  
 hybrid procedure, 190, 191  
 reason for staging, 198–9  
 stage I (Norwood) palliation, 188, 189–90, 191, 192, 198  
 stage II palliation, 191, 198, 199–201
- hypotension  
 AVSD patients, 30, 31  
 cardiomyopathy, 115–16, 119, 247, 250, 257  
 coarctation of the aorta, 125  
 Fontan failure, 234, 235  
 heart transplantation failure, 302–3  
 HLHS patients, 192, 193–4  
 intraoperative, AVSD patients, 30, 31
- ICD. *See* implantable cardioverter-defibrillator
- idiopathic pulmonary arterial hypertension (IPAH), 306  
 anesthetic management goals, 309  
 preoperative considerations, 309
- immunosuppression therapy  
 heart transplantation, 295–7  
 lung transplantation, 285–6, 287
- implantable cardioverter-defibrillator (ICD)  
 after atrial switch procedure, 147, 148  
 complications, 245  
 defibrillation or cardioversion  
 considerations, 384  
 defibrillator code, 381  
 dependency determination and precautions, 164  
 Duchenne muscular dystrophy, 242–3, 245  
 electromagnetic interference, 164–5, 232, 382  
 Fontan failure, 232  
 HCM, 116, 117, 118, 119  
 indications, 46–7, 380  
 long QT syndrome therapy, 377  
 magnet use, 165, 166–7, 383–4  
 monitoring, 383  
 perioperative management, 117, 118, 119, 164–5, 166–7, 232, 381  
 preoperative evaluation, 164, 165, 381, 382  
 rate adaptive functions, 383  
 reprogramming, 166–7, 382–3
- infective endocarditis. *See* bacterial endocarditis prophylaxis
- inhaled nitric oxide (iNO)  
 BDG shunt patients, 204  
 pulmonary hypertension, 11, 320, 321, 325  
 pulmonary hypertensive crisis, 310  
 VSD patients, 11  
 inlet ventricular septal defect, 9, 10, 25, 26  
 iNO. *See* inhaled nitric oxide
- insufflation. *See* abdominal insufflation
- intact ventricular septum, TGA with, 138, 139, 141
- intraabdominal pressure, omphalocele repair, 67, 68–9
- intraatrial baffles  
 complications, 147, 148, 152  
 echocardiography evaluation, 153–6
- intravenous agents, pulmonary hypertension effects, 312
- IPAH. *See* idiopathic pulmonary arterial hypertension
- Jervell syndrome, 376. *See also* long QT syndrome
- Kawasaki disease, 352  
 anesthetic implications  
 CMRI considerations, 356  
 induction and maintenance methods, 357  
 management plan, 355  
 monitoring, 356–7  
 perioperative medication  
 management, 355–6  
 premedication, 356  
 preprocedural assessment, 355  
 preprocedural preparation, 357  
 anesthetic risk, 357–8  
 CMRI, 354–5  
 definition, 352  
 diagnosis, 352, 353  
 echocardiography, 354  
 epidemiology, 352  
 pathophysiology, 352, 353  
 phases, 353  
 prognosis, 354  
 relevance, 353  
 selection of imaging technique, 354  
 sequelae, 353  
 treatment recommendations, 353–4
- ketamine  
 Duchenne muscular dystrophy patients, 243, 244  
 heart failure patients, 249, 250  
 pulmonary hypertension effects, 312, 321  
 PVR and cerebral perfusion effects, 336  
 TOF patients, 43  
 Williams syndrome patients, 110
- Konno operation, 93, 101
- Lange–Nielson syndrome, 376. *See also* long QT syndrome
- laparoscopic surgery  
 AVSD considerations, 29–30  
 Fontan procedure patients, 225

- HLHS, stage I palliation, 195–6  
 open approach versus, 166  
 pulmonary hypertension, 328–9  
 TAPVR, 175–6  
 VSD considerations, 12–13  
 laryngospasm, 205, 373  
 LCSD. *See* left cardiac sympathetic denervation  
 left atrial isomerism, 172  
 left cardiac sympathetic denervation (LCSD), 377  
 left dominance, unbalanced AVSD, 28  
 left ventricle (LV). *See also* hypoplastic left heart syndrome  
   Ebstein anomaly, 76  
   hypoplastic, TGA with, 217–18, 219–21  
   pulmonary hypertension effects, 325  
 left ventricular assist device (LVAD), 268–9, 270, 271. *See also* Berlin Heart EXCOR  
   right ventricular failure, 278, 279  
   right ventricular function, 277  
 left ventricular (LV) diverticulum, pentalogy of Cantrell, 65, 66  
 left ventricular (LV) hypertrophy, aortic stenosis, 83–4, 91–2, 99–100  
 left ventricular noncompaction cardiomyopathy (NCM), 252–3  
   definition, 255  
   other cardiomyopathy phenotypes versus, 246–7  
 left ventricular outflow tract (LVOT) AVSDs affecting, 20, 22, 23  
   cardiomyopathy effects, 256–7  
 left ventricular outflow tract obstruction (LVOTO)  
   aortic stenosis, 85, 86  
     subvalvular, 97–8, 99–100, 101, 102  
     valvular, 90, 91, 93  
   AVSD, 22, 23  
   CCTGA, 161  
   HCM, 113, 114, 115–16, 117, 118, 119, 120, 253–4  
   Shone complex, 129, 130, 131  
   TGA, 138, 139, 140–1  
 left-sided obstructive lesions, anesthetic risk, 6–7  
 left-to-right (L-to-R) shunting AVSDs, 25–6  
   VSDs, 9, 10, 12–13  
 levo (L)-transposition of the great arteries (L-TGA), 137–8, 146, 158–9. *See also* congenitally corrected transposition of the great arteries  
 long QT syndrome (LQTS), 374  
   acquired, 376  
   anesthetic implications  
     antiemetics, 379  
     induction and maintenance agents, 378  
     medication interactions, 377–8  
     monitoring, 378  
   nondepolarizing muscle relaxants, 378–9  
   pain management, 379  
   postoperative considerations, 379  
   premedication, 378  
   preoperative evaluation, 377  
   torsades de pointes management, 379  
 CIED therapy, 377  
 clinical presentation, 374, 375  
 congenital, 374, 375–6  
   Anderson–Tawil syndrome, 376  
   Jervell syndrome, 376  
   Lange–Nielsen syndrome, 376  
   Timothy syndrome, 376  
 definition, 374  
 genetics, 375–6  
 medical management, 376  
 mortality, 376  
 pacemaker therapy, 377  
 types  
   long QT1, 375, 376  
   long QT2, 375, 376  
   long QT3, 375, 376  
 L-TGA. *See* levo (L)-transposition of the great arteries  
 L-to-R shunting. *See* left-to-right shunting  
 lung development, TOF/PA/MAPCAs, 58  
 lung transplantation, 283  
   anesthetic implications  
     airway management, 287  
     bronchoscopic procedure complications, 288  
     extubation, 288  
     fluid balance, 288  
     induction and maintenance methods, 287, 288  
     monitoring, 287  
     outpatient considerations, 288  
     premedication, 287, 288  
     preoperative evaluation, 286, 287  
     ventilation strategy, 288  
   complications, 284  
     bronchiolitis obliterans, 283, 284–5  
     chronic allograft dysfunction, 284–5  
     primary graft dysfunction, 284–5  
     rejection, 284  
     restrictive allograft syndrome, 284–5  
   immunosuppression, 285–6, 287  
   indications, 283  
   medical comorbidities, 285–6  
   physiologic changes, 285  
   survival, 283, 284  
 LV. *See* left ventricle  
 LVAD. *See* left ventricular assist device  
 LVOT. *See* left ventricular outflow tract  
 LVOTO. *See* left ventricular outflow tract obstruction  
 magnetic resonance imaging (MRI). *See also* cardiac magnetic resonance imaging  
   anesthetic issues, 335, 336  
 Marfan syndrome, 388  
 TOF/APVS, 54  
 malignant hyperthermia, 242  
 MAPCAs. *See* multiple aortopulmonary collaterals  
 Marfan syndrome (MFS), 386  
   airway issues, 389  
   anesthetic implications  
     airway concerns, 389  
     arrhythmia onset, 390, 391  
     bacterial endocarditis prophylaxis, 390  
     emergence and extubation, 391  
     hemodynamic goals, 389  
     history of mitral valve repair, 390  
     induction methods, 390  
     maintenance concerns, 390  
     monitoring, 389  
     patient positioning, 390  
     perioperative medication continuation, 388  
     postoperative recovery, 391  
     premedication, 389  
     preoperative diagnostic studies, 388  
     preoperative evaluation, 388  
     regional or epidural anesthesia concerns, 389  
   cardiovascular abnormalities, 386–7  
   definition, 386  
   medical therapy, 387, 388, 391  
   pectus excavatum correction, 387–8  
   z-score, 387  
 mBTS. *See* modified Blalock–Taussig shunt  
 mechanical circulatory support (MCS). *See also* extracorporeal membrane oxygenation; ventricular assist device  
   indications, 270–1, 276  
   types, 276  
 membranous subvalvular aortic stenosis, 97–8  
 MFS. *See* Marfan syndrome  
 midazolam, 49, 73  
 pulmonary hypertension effects, 312  
 milrinone  
   Fontan procedure patients, 214–15  
   pulmonary hypertensive crisis, 311  
   pulmonic stenosis, 35  
 mitral regurgitation (MR), 371  
 mitral valve. *See also* mitral regurgitation; Shone complex  
   anterior leaflet cleft, 20, 21  
   parachute, 129, 130, 131  
   systolic anterior motion, 114  
 mitral valve repair, Marfan syndrome, 390  
 modified Blalock–Taussig shunt (mBTS) DORV, 17, 18–19  
   Ebstein anomaly, 77–8  
   HLHS, 189, 190, 191, 192, 194, 196, 198–9  
   monitoring, 218  
   pulmonary stenosis, 38  
   TAPVR and heterotaxy syndrome, 173–5

- modified Blalock-Taussig shunt (cont.)  
 TGA with hypoplastic left ventricle, 217–18  
 thrombosis, 194, 196  
 tricuspid atresia, 208–9  
 moyamoya disease, 331  
 anesthetic management, 334–7  
 associated conditions, 331  
 cerebral autoregulation, 332, 333  
 coarctation of the aorta with, 332–3  
 definition, 331  
 diagnosis, 331–2  
 treatment, 332  
 MPS. *See* mucopolysaccharidoses  
 MR. *See* mitral regurgitation  
 MRI. *See* magnetic resonance imaging  
 mucopolysaccharidoses (MPS). *See also* Hurler syndrome  
 airway management, 369–70, 371, 372–3  
 anesthetic concerns, 367  
 classification, 367, 368  
 glycosaminoglycans, 367  
 preoperative assessment, 369  
 multiple aortopulmonary collaterals (MAPCAs). *See also* tetralogy of Fallot with pulmonary atresia and multiple aortopulmonary collaterals  
 acquired or persistent, 61  
 direct, 58  
 indirect, 58  
 TOF with, 41, 57, 58, 60  
 unifocalization of, 58–60  
 muscular ventricular septal defect, 9, 10  
 Mustard procedure, 140, 146, 147, 161–2  
 myectomy, HCM, 116  
 National Surgical Quality Improvement Project-Pediatrics (NSQIP-P), CHD severity classification, 166  
 NCM. *See* left ventricular noncompaction cardiomyopathy  
 neostigmine, post-heart transplantation, 297  
 neuromuscular blocking drugs, Duchenne muscular dystrophy, 242, 244  
 nicardipine, 127  
 Nikaidoh procedure, 141  
 nitric oxide. *See* inhaled nitric oxide  
 noncommitted ventricular septal defect, 15–16  
 noncompaction cardiomyopathy. *See* left ventricular noncompaction cardiomyopathy  
 nonrestrictive shunts. *See* dependent shunts  
 nonrestrictive ventricular septal defect, 10, 25–6  
 Noonan syndrome, 34–5  
 Norwood procedure, 189–90, 198. *See also* stage I palliation  
 NSQIP-P. *See* National Surgical Quality Improvement Project-Pediatrics  
 Nuss procedure, 387–8  
 complications, 390, 391  
 CPR during or after, 391  
 postoperative pain strategies, 388–9  
 obliterative bronchiolitis, 283, 284–5  
 obstructions  
 dynamic, 3, 40  
 fixed, 3, 40  
 obstructive airway disease  
 APVS and, 52, 53–4, 55, 56  
 TOF/PA/MAPCAs and, 60–1  
 oculocardiac reflex, 18  
 omphalocele  
 abdominal compartment syndrome, 68–9  
 pentalogy of Cantrell, 64–5, 66, 67  
 one-and-a-half-ventricle palliation, Ebstein anomaly, 72, 77  
 opioids  
 pulmonary hypertension effects, 312  
 PVR and cerebral perfusion effects, 336  
 orthotopic heart transplantation. *See* heart transplantation  
 overriding aorta, TOF with, 39–40  
 PA. *See* pulmonary artery  
 pacemaker. *See also* cardiovascular implantable electronic devices; implantable cardioverter-defibrillator  
 after atrial switch procedure, 147, 148  
 complications, 245  
 defibrillation or cardioversion considerations, 384  
 dependence, 381  
 dependency determination and precautions, 164  
 dual chamber, 380  
 Duchenne muscular dystrophy, 242–3, 245  
 electromagnetic interference, 164–5, 232, 382  
 Fontan failure, 229, 232  
 indications, 380  
 interrogation, 381–2  
 long QT syndrome therapy, 377  
 magnet use, 165, 166–7, 383–4  
 monitoring, 383  
 multisite pacing, 380  
 pacemaker code, 380  
 perioperative management, 164–5, 166–7, 232, 381  
 preoperative evaluation, 164, 165, 381, 382  
 rate adaptive functions, 383  
 rate modulation, 380  
 reprogramming, 166–7, 382–3  
 single chamber, 380  
 PAH. *See* pulmonary arterial hypertension  
 palpitations, HCM, 115  
 Panama classification, 333  
 parachute mitral valve, 129, 130, 131  
 parallel circulation, 3, 4, 137–8, 174–5, 188, 217  
 partial atrioventricular septal defect, 20–2  
 patent ductus arteriosus (PDA)  
 aortic stenosis, 83–4, 90–1  
 coarctation of the aorta, 122, 123  
 Ebstein anomaly, 75–6, 77  
 HLHS, 187–8  
 parallel circulations, 3, 4  
 pulmonic stenosis, 35  
 stent placement, 366  
 TGA, 137, 138, 139  
 TGA with hypoplastic left ventricle, 217–18  
 TOF/APVS, 53  
 TOF/PA/MAPCAs, 58, 60  
 TOF-type DORV, 15–16  
 tricuspid atresia, 208  
 unbalanced AVSD, 28, 29  
 VACTERL association, 360–1, 365–6  
 patent foramen ovale (PFO)  
 pulmonary hypertension and, 332, 333  
 TGA, 137, 138, 139  
 TOF, 41  
 PBF. *See* pulmonary blood flow  
 PCE. *See* pericardial effusion  
 PDA. *See* patent ductus arteriosus  
 pectus excavatum  
 CPR strategy, 391  
 Nuss correction, 387–9, 390, 391  
 Pediatric Difficult Intubation Registry, airway management, 370, 371  
 pentalogy of Cantrell (POC), 64  
 abdominal wall defects, 64–5, 66  
 anatomy, 64  
 anesthetic implications  
 abdominal compartment syndrome, 68–9  
 induction and intubation considerations, 68  
 intraoperative concerns for omphalocele repair, 67  
 intraoperative concerns for sternal malformation repair, 67–8  
 postoperative concerns, 69  
 preoperative evaluation, 67  
 prognosis, 69  
 vascular access and monitoring, 68  
 ventilatory strategies, 68  
 associated anomalies, 64  
 cardiac defects, 65, 66  
 classification, 64  
 ectopia cordis, 65, 66, 67  
 LV diverticulum, 65, 66  
 prevalence, epidemiology, and embryology, 64  
 pulmonary hypertension, 65–6, 68  
 sternal malformations, 65, 67  
 surgical options and strategies, 66–7  
 pentalogy of Fallot, 41  
 pericardial effusion (PCE), 345

- anesthesia implications  
fasting considerations, 347  
hemodynamic goals, 349  
induction method, 348–9  
management plan, 348  
monitoring, 348  
past medical history, 347  
physical examination, 347  
sedation, 349  
separation anxiety, 347–8  
cardiac tamponade and, 345–6  
cardiopulmonary interactions, 347  
definition and causes, 345  
ECG findings, 346  
hemodynamic goals, 349  
hemodynamic monitoring findings, 346  
pericardiocentesis, 349–50  
complications, 350–1  
pericardiocentesis, 349–50  
complications, 350–1  
perimembranous ventricular septal defect, 9  
peripherally inserted central catheter (PICC), placement, 311  
PFO. *See* patent foramen ovale  
PGD. *See* primary graft dysfunction  
PGE<sub>1</sub>. *See* prostaglandin E<sub>1</sub>  
PH. *See* pulmonary hypertension  
phenylephrine  
HCM, 119  
intraoperative cyanosis, DORV patients with, 18–19  
tet spell management, 43–4, 361  
VSD patients, 12  
phosphodiesterase inhibitors, 307–8, 320, 325, 334  
physiologic approach, 2–3  
physiologic shunting, 2, 3  
PI. *See* pulmonary insufficiency  
PICC. *See* peripherally inserted central catheter  
Pierre Robin sequence, 182  
pink tet, 42, 360–1  
plastic bronchitis, 221, 229  
PLE. *See* protein-losing enteropathy  
pneumothorax  
Marfan syndrome, 390  
pacemaker or ICD placement  
complications, 245  
POC. *See* pentalogy of Cantrell  
positive pressure ventilation (PPV)  
Fontan procedure patients, 224, 234  
HLHS  
BDG shunt patients, 204–5  
stage I palliation patients, 194  
TAPVR, 175  
TGA, 144  
Potts shunt, 308  
PPV. *See* positive pressure ventilation  
PR. *See* pulmonary regurgitation  
preexcitation, Ebstein anomaly, 71  
premature infants  
anemia, 11–12  
apnea, 11–12, 13  
bronchopulmonary dysplasia, 12, 323–4, 325  
chronic lung disease, 11–12  
hospital discharge, 13  
pulmonary hypertension, 323  
airway management, 325  
anesthetic implications, 326–9  
anesthetic risk, 327  
bronchopulmonary dysplasia and, 323–4, 325  
cardiac function changes, 325  
definition, 323  
diagnosis, 324–5  
iNO effects, 325  
medical management, 325  
risk factors, 324  
screening, 324  
shunt effects, 325–6  
sildenafil effects, 325  
VSDs, 9, 10, 11–12, 13  
preoperative analysis, 2  
basic concepts and terminology, 2–3, 4  
high-risk patient populations, 6–7, 79, 107, 111  
imaging, 3–6  
pressure gradients  
cardiac catheterization data, 4, 5  
echocardiography data, 6  
preterm infants. *See* premature infants  
primary graft dysfunction (PGD), lung transplantation, 284–5  
propofol  
BDG shunt patients, 204  
Duchenne muscular dystrophy patients, 243, 244  
pulmonary hypertension effects, 312, 321  
PVR and cerebral perfusion effects, 336  
Williams syndrome patients, 110  
prostacyclin analogs  
pulmonary arterial hypertension, 307–8, 334  
pulmonary hypertension, 320, 321  
prostaglandin E<sub>1</sub> (PGE<sub>1</sub>)  
adverse effects, 35, 188  
aortic stenosis, 84, 90–1  
AVSD, 28, 29  
coarctation of the aorta, 123  
DORV, 15–16  
Ebstein anomaly, 77  
inability to wean from, 77–8  
HLHS, 187–8  
pulmonic stenosis, 35, 38  
TGA, 138, 139  
TGA with hypoplastic left ventricle, 217–18  
tricuspid atresia, 208  
VACTERL association, 361, 365–6  
protein-losing enteropathy (PLE), 221, 229  
PS. *See* pulmonary (pulmonic) stenosis  
pulmonary arterial hypertension (PAH), 305  
acute vasoreactivity testing, 307  
anesthetic management goals, 309  
anesthetic risk, 308–9  
classification, 306, 308  
definition, 305  
diagnosis, 306–7  
idiopathic, 306, 309  
pathophysiology, 305–6  
preoperative planning, 309  
treatment  
goals, 307  
initial options, 307  
procedural options, 308  
targeted therapies, 307–8, 309  
in trisomy 21 patients, 28–9  
pulmonary artery (PA)  
banding  
AVSDs, 22  
CCTGA, 163  
hybrid stage I palliation of HLHS, 191  
hypoplasia  
DORV, 14–15  
pentalogy of Cantrell, 65–6  
supravalvular stenosis, 105–6  
truncus arteriosus, 178–9  
pulmonary atresia. *See also* tetralogy of Fallot with pulmonary atresia and multiple aortopulmonary collaterals  
Ebstein anomaly, 71, 72, 75–6, 77–8  
hypoplastic left ventricle with, 217–18  
TOF, 41, 57, 58  
VSD/MAPCAs, 58, 59  
pulmonary blood flow (PBF, Q<sub>p</sub>). *See also* Q<sub>p</sub>:Q<sub>s</sub> ratio  
cardiac catheterization data, 4, 5  
Ebstein anomaly, 75–6, 77, 81  
after BDG shunt, 79, 80  
effective, 2  
Fontan procedure patients, 209–10, 219, 223, 226  
mBT shunt effects, 218  
TOF/PA/MAPCAs, 57, 58, 60  
total, 3  
tricuspid atresia, 208  
pulmonary function studies  
Duchenne muscular dystrophy, 241  
Marfan syndrome, 388  
pulmonary hypertension (PH), 305. *See also* pulmonary arterial hypertension  
acute vasoreactivity testing, 307  
anesthetic implications  
airway management, 312, 325, 328  
anesthetic medication effects, 312, 321  
emergence precautions, 313  
fluid management, 313  
induction and maintenance methods, 311–12, 328, 336–7  
laparoscopic approach, 328–9  
management goals, 309, 335–6  
monitoring, 311, 321, 327–8, 337  
MRI issues, 335, 336

- pulmonary hypertension (PH) (cont.)  
 perioperative medical management, 320  
 PICC line placement, 311  
 postoperative care, 313, 322, 329, 337  
 premedication, 311  
 preoperative evaluation, 308–9, 316–17, 318–20, 326–9, 334–5  
 preoperative medication preparation, 311, 320, 321  
 preoperative planning, 309  
 ventilation strategy, 312–13, 328  
 anesthetic risk, 6–7, 308–9, 316, 318–19  
 assessment  
   cardiac catheterization, 306–7, 308–9, 317, 318, 319, 324–5, 334  
   echocardiography, 306–7, 309, 316–17, 324  
   preoperative, 308–9, 316–17, 318–20  
   severity, 318–19  
 AVSDs with, 22, 27, 28–9  
 classification, 306  
   Panama, 333  
   WHO, 306, 308, 333  
 definition, 305  
 diagnosis, 306–7  
 diaphragmatic hernia with, 261  
 Down syndrome and, 314, 315–16, 319–20  
 medical therapy  
   anticoagulation, 307, 334  
   calcium channel blockers, 307, 320  
   endothelin receptor antagonists, 307–8, 334  
   goals, 307  
   initial options, 307  
   iNO, 11, 320, 321, 325  
   mechanisms of action and potential side effects, 334  
   perioperative management, 320  
   phosphodiesterase inhibitors, 307–8, 320, 325, 334  
   premature infants, 325  
   preoperative considerations, 309  
   prostacyclin, 320, 321  
   prostacyclin analogs, 307–8, 334  
   selection, 308  
 moyamoya disease, 331  
   anesthetic management, 334–7  
   associated conditions, 331  
   cerebral autoregulation, 332, 333  
   coarctation of the aorta with, 332–3  
   definition, 331  
   diagnosis, 331–2  
   treatment, 332  
 PAH versus, 305  
 pathophysiology, 305–6  
 pentalogy of Cantrell, 65–6, 68  
 prematurity, 323  
   airway management, 325  
   anesthetic implications, 326–9  
   anesthetic risk, 327  
   bronchopulmonary dysplasia and, 323–4, 325  
   definition, 323  
   diagnosis, 324–5  
   iNO effects, 325  
   medical management, 325  
   risk factors, 324  
   screening, 324  
   shunt effects, 325–6  
   sildenafil effects, 325  
 pulmonary arterial hypertension, 28–9  
 pulmonary hypertensive crisis, 310  
   definition, 329  
   presentation, 310, 321, 329  
   treatment, 310–11, 321–2, 329  
 PVR measurement, 5  
 risk factors, 315  
   bronchopulmonary dysplasia, 323–4  
   Down syndrome, 314, 315–16  
   Shone complex, 132  
   trisomy 21 and risk for, 28–9  
   VAD complications, 271  
 pulmonary hypoperfusion, DORV with, 15–16, 17  
 pulmonary insufficiency (PI), after TOF repair, 46, 47  
 pulmonary overcirculation  
   AVSDs, 20–3  
   DORV, 15–16, 17  
   truncus arteriosus, 180, 181  
   ventilation and, 12  
   VSDs, 11, 12  
 pulmonary regurgitation (PR), peak velocity, 316–17  
 pulmonary (pulmonic) stenosis (PS), 33.  
   *See also* tetralogy of Fallot with pulmonary stenosis  
   anesthetic implications  
     balloon valvuloplasty evaluation, 38  
     extubation, 37–8  
     fluid administration, 37  
     hemodynamic goals, 36–7  
     induction plan, 37  
     maintenance plan, 37  
     postprocedural management, 38  
     potential intraoperative complications, 37  
     preoperative cardiac evaluation and workup, 35–6  
     surgical procedures for unsuccessful catheterization, 38  
     ventilation strategy, 37  
   associated lesions and syndromes, 34–5  
   balloon valvuloplasty procedure, 35, 36  
   critical, 33, 34–5  
   definitions and classifications, 33  
   DORV with, 15–16, 17–18  
     subvalvular, 17  
     supravalvular, 17  
     valvular, 17  
   oxygen saturation expectations, 35  
   pathophysiology, 33, 34  
   PGE<sub>1</sub> infusion, 35  
   TGA with, 142, 143–4  
   TOF with, 39–40, 41, 42  
   tricuspid atresia with, 207–8  
 pulmonary valve (PV). *See also* absent pulmonary valve syndrome;  
   pulmonary (pulmonic) stenosis  
   replacement of, after TOF repair, 47  
   TOF with absent, 41, 52, 53  
 pulmonary vascular occlusive disease (PVOD)  
   AVSDs leading to, 20–2  
   trisomy 21, 22, 28–9  
   VSDs leading to, 10  
 pulmonary vascular resistance (PVR)  
   anesthetic effects, 12, 30, 31, 336  
   AVSDs, 25–6, 30, 31  
   BDG shunt, 204, 219  
   cardiac catheterization data, 4, 5  
   Ebstein anomaly, 75–6, 77, 81  
   Fontan procedure patients, 221  
   intraoperative reduction, 320  
   pulmonary hypertension, 305, 314, 323–4, 325–6  
   pulmonary hypertensive crisis, 310  
     definition, 329  
     presentation, 310, 321, 329  
     treatment, 310–11, 321–2, 329  
   pulmonic stenosis, 36–7  
   shunts and, 3, 325–6  
   TAPVR, 175, 176  
   truncus arteriosus, 180, 181  
   VAD considerations, 281  
   VSDs, 9, 10–11, 12–13  
 pulmonary veins. *See also* total anomalous pulmonary venous return  
   stenosis, 323–4  
 pulmonary venous obstruction (PVO), TAPVR, 169–70, 171, 174, 175–6  
 pulmonary venous stenosis, recurrent, TAPVR and heterotaxy syndrome, 171  
 PV. *See* pulmonary valve  
 PVO. *See* pulmonary venous obstruction  
 PVOD. *See* pulmonary vascular occlusive disease  
 PVR. *See* pulmonary vascular resistance  
 pyridostigmine, post-heart transplantation, 297  
 Q<sub>p</sub>. *See* pulmonary blood flow  
 Q<sub>p</sub>:Q<sub>s</sub> ratio, 3  
   balanced, 218  
   calculation, 10–11  
   cardiac catheterization data, 4, 5  
   HLHS, 191  
     anesthetic implications, 193–5  
     stage I palliation, 191, 192  
   TOF/PA/MAPCAs, 60, 62–3  
   truncus arteriosus, 180, 181  
   VSDs, 10–11  
 Q<sub>s</sub>. *See* systemic blood flow

- QT prolongation. *See also* long QT syndrome  
Williams syndrome, 106
- RA. *See* right atrium
- RAA. *See* right aortic arch
- RAS. *See* restrictive allograft syndrome
- Rastelli classification, 26
- Rastelli procedure, 141
- RCM. *See* restrictive cardiomyopathy
- rejection  
heart transplantation, 292  
lung transplantation, 284
- Remodulin®. *See* treprostinil
- renal dysfunction, Fontan failure, 229
- réparation à l'étage ventriculaire (REV), 141
- resistance. *See also* pulmonary vascular resistance; systemic vascular resistance  
cardiac catheterization data, 5
- restrictive allograft syndrome (RAS), 284–5
- restrictive ASD, 25–6
- restrictive cardiomyopathy (RCM), 252–3, 254  
clinical presentation, 254  
incidence, 254  
intraoperative anesthetic goals, 257  
other cardiomyopathy phenotypes versus, 246–7  
pathophysiology, 254–5, 258  
prognosis and management options, 255
- restrictive ventricular septal defect, 10, 25–6
- REV. *See* réparation à l'étage ventriculaire
- right aortic arch (RAA), 339, 340  
TOF, 41  
VACTERL association, 360, 362
- right atrial isomerism, 172
- right atrium (RA), TAPVR, 169–70
- right dominance, unbalanced AVSD, 28
- right ventricle (RV). *See also* Ebstein anomaly  
atrial switch procedure, 148–9, 150–2  
CCTGA, 160–1, 163–4  
congenital defects, 70, 71, 73, 75–6  
hypoplastic, tricuspid atresia with, 207–9  
LVAD use and, 277, 278  
pulmonary hypertension assessment, 317  
pulmonary hypertension effects, 325
- right ventricle to pulmonary artery (RV–PA) conduit, truncus arteriosus repair, 181  
stenosis, 182, 183–4
- right ventricular assist device (RVAD), 276, 278, 279  
decreased blood flow considerations, 281  
indications, 279  
respiratory support, 281–2
- right ventricular hypertrophy (RVH)  
TGA, 143  
TOF, 39–40
- right ventricular outflow tract (RVOT), stenting, 361, 365–6
- right ventricular outflow tract obstruction (RVOTO)  
DORV, 14–16  
dynamic, 40  
Ebstein anomaly, 75–6  
fixed, 40  
Tet spells and, 41  
after TGA repair, 141  
TOF, 3, 39–40, 43  
truncus arteriosus, 181, 183–4  
VACTERL association, 360–1
- right-to-left (R-to-L) shunting, Fontan failure, 229
- Ross classification for pediatric heart failure, 163, 164
- Ross procedure, valvular aortic stenosis, 93, 94  
surgical outcomes, 94–5
- Ross–Konno procedure  
subvalvular aortic stenosis, 101  
valvular aortic stenosis, 93–4, 95  
surgical outcomes, 94
- R-to-L shunting. *See* right-to-left shunting
- RV. *See* right ventricle
- RVAD. *See* right ventricular assist device
- RVH. *See* right ventricular hypertrophy
- RVOT. *See* right ventricular outflow tract
- RVOTO. *See* right ventricular outflow tract obstruction
- RV–PA conduit. *See* right ventricle to pulmonary artery conduit
- SAM. *See* systolic anterior motion
- Sano shunt, stage I palliation, 189, 190, 191, 192, 198–9
- saturation data, cardiac catheterization, 4
- SAV. *See* surgical aortic valvotomy
- SCD. *See* sudden cardiac death
- SCPA. *See* superior cavopulmonary anastomosis
- segmental approach, 2
- Senning procedure, 140, 146, 148  
CCTGA, 161–2
- Senning–Rastelli procedure, 162, 163
- series circulation, 3, 4, 137–8, 188, 226
- sevoflurane  
TOF patients, 43–4  
trisomy 21 patients, 29
- Shone complex, 129  
anesthetic implications  
afterload monitoring and management, 133–4  
anesthetic agent selection, 132  
contractility monitoring and management, 134  
intraoperative considerations, 133  
intravenous catheter placement, 132–3  
postoperative disposition, 134–5  
postoperative pain control, 134
- preload monitoring and management, 133
- preoperative evaluation, 132
- preoperative management, 132
- aortic coarctation correction, 131
- characteristic cardiac anomalies, 129–30, 131
- coarctation of the aorta, 121, 129, 130, 131
- correction surgery or techniques, 129–30, 131
- parachute mitral valve, 129, 130, 131
- partial, 129
- pulmonary hypertension, 132
- respiratory involvement, 132
- surgical approaches, 129–30, 131
- shortening fraction, 4, 6
- shunts, 2–3. *See also* specific shunts  
anatomic, 2, 3  
cardiac catheterization data, 4, 5  
complex, 3  
dependent (nonrestrictive), 3  
physiologic, 2, 3  
pulmonary hypertension with, 325–6  
simple, 3
- sildenafil, 325
- simple shunts, 3
- single ventricle palliation. *See also* Fontan procedure; stage I palliation; stage II palliation  
CCTGA, 162  
Ebstein anomaly, 72, 77, 78–9  
reason for staging, 198–9  
superior cavopulmonary anastomosis indications, 199
- single ventricle physiology, 187. *See also* hypoplastic left heart syndrome  
anesthetic risk, 6–7, 79  
cerebral perfusion pressure, 203–4  
parallel circulations, 3, 4  
TAPVR, 171, 173–4  
TGA with hypoplastic left ventricle, 217–18, 219–21  
tricuspid atresia, 207–9
- situs ambiguus, 2
- situs inversus, 2
- situs solitus, 2, 159–60
- stage I palliation, 198  
anesthetic implications  
anesthetic risk counselling, 192, 193  
ECMO capability considerations, 192, 196, 197  
extubation, 196–7  
failure to thrive, 192  
hypoxemia associated with hypertension, 195  
induction method and hemodynamic goals, 193–4  
intraoperative and postoperative analgesic options, 196  
laparoscopy considerations, 195–6  
open laparotomy considerations, 195–6

- stage I palliation (cont.)  
 postoperative disposition, 196  
 preoperative assessment, 192  
 refractory hypoxemia, 196  
 systemic and pulmonary blood flow management, 193–5  
 systemic-to-pulmonary shunt considerations, 193–4  
 vascular access and monitoring, 193  
 ventilation strategy, 194  
 blood flow pattern after, 190  
 hybrid approach, 190, 191  
 repair timing, 188  
 surgical procedure and goals, 189–90  
 surgical stages after, 191
- stage II palliation, 191, 198. *See also*  
 bidirectional Glenn shunt  
 BDG shunt procedure, 198, 199, 200  
 blood flow pattern after, 199–201  
 hemi-Fontan procedure, 191, 198, 199, 200  
 indications for SCPA, 199  
 oxygen saturation expectations, 201  
 physiologic goals, 199  
 reason for staging, 198–9  
 tricuspid atresia, 208–9
- Starnes procedure, 72, 77, 78–9
- sternal malformations, pentalogy of  
 Cantrell, 65, 67–8
- stress dose steroids, 296
- stress perfusion CMRI, 354–5
- subaortic stenosis  
 AVSDs, 22, 23  
 DORV, 14–15  
 Shone complex, 129, 130
- subaortic ventricular septal defect, 15–16, 17
- subarterial/subpulmonic ventricular septal defect, 9, 10, 15–16
- subclavian artery, aberrant origin of, TOF, 41
- subvalvular aortic stenosis. *See* aortic stenosis
- succinylcholine  
 Duchenne muscular dystrophy, 242, 244  
 long QT syndrome, 378–9
- sudden cardiac death (SCD)  
 HCM, 113  
 long QT syndrome, 374, 375, 376  
 after TOF repair, 46–7  
 Williams syndrome, 107  
 risk factors, 107, 111
- suicide right ventricle, 38
- superior cavopulmonary anastomosis (SCPA), 28, 173. *See also*  
 bidirectional Glenn shunt  
 central venous access considerations, 203  
 indications, 199  
 stage II palliation, 191, 198, 199, 200  
 BDG versus hemi-Fontan, 199, 200  
 cerebral perfusion pressure effects, 203–4  
 for tricuspid atresia, 208–9
- TGA with hypoplastic left ventricle, 219
- supravalvular aortic stenosis (SVAS), 90, 91, 105. *See also* Williams syndrome  
 surgical repair indications, 107  
 without WS, 106
- supravalvular pulmonary artery stenosis (SVPS), Williams syndrome, 105–6
- supraventricular reentrant tachycardia, 147
- surgical aortic valvotomy (SAV), aortic stenosis, 84, 85, 86, 93
- SVAS. *See* supravalvular aortic stenosis
- SVPS. *See* supravalvular pulmonary artery stenosis
- SVR. *See* systemic vascular resistance
- syncope, HCM, 114
- systemic blood flow ( $Q_s$ ). *See also*  $Q_p:Q_s$  ratio  
 cardiac catheterization data, 4, 5  
 effective, 2  
 total, 3
- systemic circulation, series versus parallel, 3, 4
- systemic vascular resistance (SVR)  
 anesthesia effects, 12, 30, 31  
 AVSDs, 25–6, 30, 31  
 cardiac catheterization data, 4, 5  
 pulmonary hypertensive crisis treatment, 310–11  
 Shone complex, 133–4  
 shunts and, 3  
 truncus arteriosus, 180, 181  
 VAD considerations, 274  
 VSDs, 10–11, 12
- systemic-to-pulmonary artery shunt. *See also* modified Blalock-Taussig shunt  
 TAPVR and heterotaxy syndrome, 173–4  
 TOF, 42
- systolic anterior motion (SAM), mitral valve, 114
- TA. *See* tricuspid atresia; truncus arteriosus
- tachycardia  
 cardiomyopathy, 115, 250, 257  
 post-heart transplantation, 294  
 Shone complex, 134
- tacrolimus, 296–7
- TAPSE. *See* tricuspid annular plane systolic excursion
- TAPVR. *See* total anomalous pulmonary venous return
- Taussig-Bing anomaly, 15–16
- TCPA. *See* total cavopulmonary anastomosis
- TEF. *See* tracheoesophageal fistula
- tet spells, 41, 52, 360. *See also* tetralogy of Fallot  
 management, 43–4, 361  
 pink tets, 42, 360–1  
 preoperative assessment, 43  
 variability in, 2
- tetralogy of Fallot (TOF)  
 absent pulmonary valve and, 41, 52, 53  
 anatomy, 39–40, 45, 360  
 associated cardiac lesions, 41  
 associated genetic syndromes, 47–8  
 diagnosis, 360  
 DiGeorge syndrome, 48  
 genetic testing in, 47–8  
 MAPCAs and, 41, 57, 58, 60  
 obstruction and shunting, 3  
 pathophysiologic and anatomic variations, 2, 41, 46  
 pulmonary atresia and, 41, 57, 58  
 pulmonic stenosis and, 39–40, 41, 42  
 surgical approaches, 45–6  
 postoperative sequelae, 46, 47  
 VACTERL association, 360–1, 362, 365–6
- tetralogy of Fallot (TOF), repaired, 45  
 anesthetic implications  
 anesthetic risks, 49  
 bacterial endocarditis prophylaxis, 49  
 extubation, 51  
 induction options, 50  
 intraoperative arrhythmias, 50–1  
 intraoperative fluid management, 50  
 intraoperative monitoring, 50  
 maintenance goals, 50  
 peripheral intravenous catheter placement, 49–50  
 postoperative monitoring, 51  
 premedication, 49  
 preoperative assessment, 48–9  
 preoperative testing, 49  
 cardiac anomalies of TOF, 45  
 chronic postoperative sequelae, 46, 47  
 DiGeorge syndrome relevance, 48  
 extracardiac manifestations of genetic syndromes associated with TOF, 48  
 genetic testing, 47–8  
 ICD placement consideration, 46–7  
 pulmonary valve replacement consideration, 47  
 TOF variation impacts on repair, 46  
 transannular patch implications, 46  
 typical surgical approaches, 45–6  
 ventriculotomy implications, 46
- tetralogy of Fallot with absent pulmonary valve syndrome (TOF/APVS), 52  
 anatomic features, 52, 53  
 anesthetic implications  
 induction options, 55  
 lower airway obstruction, 55  
 postoperative issues, 56  
 preoperative evaluation, 54–5  
 ventilation strategies, 53–4, 55–6  
 chest imaging findings, 54  
 PDA role, 53  
 respiratory pathology, 41, 52, 53–4  
 surgical repair options and considerations, 54  
 symptomatology, 53–4

- tetralogy of Fallot with pulmonary atresia and multiple aortopulmonary collaterals (TOF/PA/MAPCAs), 57
- anatomic characteristics, 57, 58
- anesthetic implications
- associated genetic syndromes, 61
  - imaging, 61–2
  - induction and maintenance considerations, 62–3
  - postoperative disposition, 63
  - preoperative assessment priorities, 62
- blood flow path at birth, 58
- long-term complications, 60–1
- MAPCA impacts, 58, 60
- oxygen saturation,  $Q_p:Q_s$  ratio, and hemodynamics, 60, 62–3
- pulmonary blood flow, 57, 58, 60
- surgical palliative or repair approaches, 58–60
- tetralogy of Fallot with pulmonary stenosis (TOF/PS), 39
- anatomic variations, 41
- anesthetic implications
- induction options, 43
  - maintenance considerations, 44
  - peripheral intravenous line placement, 43
  - physical examination findings, 43
  - postoperative care, 44
  - preoperative assessment questions, 43
  - preoperative diagnostic tests, 42–3
  - tet spell management, 43–4
- associated lesions or abnormalities, 41
- defects present in classic TOF, 39–40
- overriding aorta, 39–40
- pink tet, 42
- repair timing and procedures, 42
- RVOTO, 39–40
- tet spell severity, 41
- VSD, 39–40
- TGA. *See* transposition of the great arteries
- TGA-IVS. *See* transposition of the great arteries with intact ventricular septum
- TGA-type DORV, 15–16, 17
- TGA-VSD. *See* transposition of the great arteries with ventricular septal defect
- thromboembolism, Fontan failure, 230
- Timothy syndrome, 376. *See also* long QT syndrome
- TOF. *See* tetralogy of Fallot
- TOF/APVS. *See* tetralogy of Fallot with absent pulmonary valve syndrome
- TOF/PA/MAPCAs. *See* tetralogy of Fallot with pulmonary atresia and multiple aortopulmonary collaterals
- TOF/PS. *See* tetralogy of Fallot with pulmonary stenosis
- TOF-type DORV, 15–16, 17
- torsades de pointes, 374
- management, 379
- total anomalous pulmonary venous return (TAPVR), 169
- anatomic characteristics, 169–70
- anatomic subtypes and classification
- cardiac, 170, 171
  - infracardiac, 170, 171
  - mixed, 170, 171
  - supracardiac, 170, 171
- anesthetic implications
- extubation, 176
  - induction and maintenance methods, 174–5
  - intraoperative concerns, 175–6
  - malrotation management, 173
  - monitoring, 174
  - postoperative management, 176
  - preoperative evaluation, 174
  - risk discussion, 173–4
  - ventilation strategy, 175
- anesthetic risk, 173–4
- associated cardiac defects, 171
- heterotaxy syndrome with, 169–70, 171
- classification, 172
  - infection-related precautions, 173
  - organ involvement, 172
  - perioperative outcomes, 171
- incidence, 169
- postoperative sequelae, 171
- pulmonary venous obstruction with, 169–70, 171, 174, 175–6
- surgical repair, 171
- Total Artificial Heart, 268
- total blood flow, 3
- total cavopulmonary anastomosis (TCPA), 219, 227–8. *See also* Fontan procedure
- TPG. *See* transpulmonary gradient
- TR. *See* tricuspid regurgitation
- tracheobronchomalacia, APVS, 41, 53–4, 55–6
- tracheoesophageal fistula (TEF), 359–60, 361. *See also* esophageal atresia
- tranexamic acid, 127
- transannular patch, TOF repair, 45–6
- transfusion
- Fontan procedure patients, 214–15
  - guidelines, 127
- transitional atrioventricular septal defect, 20–2
- transposition of the great arteries (TGA), 137, 145. *See also* arterial switch operation; atrial switch procedure; congenitally corrected transposition of the great arteries
- anatomy, 137, 138, 145
- anesthetic implications
- anesthetic risk counselling, 142–3
  - cardiac catheterization procedures, 152–3
  - general anesthetic considerations for pulmonary stenosis, 143
  - high-risk patients, 151–2
  - induction considerations, 143–4
  - monitoring and invasive access considerations, 143, 152, 153
  - outpatient considerations, 144
  - preoperative assessment, 142, 150–1
  - procedure location, 144, 156
  - pulmonary stenosis diagnosis, 142
  - ventilation strategy, 144
- anesthetic risks, 142–3, 151–2
- associated anomalies, 139
- atrioventricular discordance, 158, 159
- dextro-, 137–8, 145
- subtypes, 138–9
- DORV and, 14–15
- historical background, 146
- levo-, 137–8, 146, 158–9
- newborn physiology, 138–9
- parallel circulations in, 3, 4, 137–8
- physiologic shunting in, 2
- series circulation in, 137–8
- surgical approaches, 140
- arterial switch operation, 17, 140, 141, 145
  - atrial switch procedure, 140, 145, 146, 147, 148–50
  - follow-up, 142, 149–50
  - long-term complications, 141, 147, 148, 149, 150
  - Mustard procedure, 140, 146, 147
  - Nikaidoh procedure, 141
  - Rastelli procedure, 141
  - REV, 141
  - Senning procedure, 140, 146, 148
- ventriculoarterial discordance, 137–8, 158, 159, 217
- transposition of the great arteries with intact ventricular septum (TGA-IVS), 138, 139, 141
- transposition of the great arteries with left ventricular outflow tract obstruction (TGA-LVOTO), 138, 139, 140–1
- transposition of the great arteries with ventricular septal defect (TGA-VSD), 138, 139, 141
- transpulmonary gradient (TPG), 209–10, 219, 223, 226
- treprostinil (Remodulin®), 320
- tricuspid annular plane systolic excursion (TAPSE), 149
- tricuspid atresia (TA), 207
- anatomy, 207–8
- blood flow pathway, 208
- classifications, 208
- pulmonary blood flow sources, 208
- surgical approaches, 208–9
- Fontan procedure, 209
- tricuspid regurgitation (TR)
- atrial switch procedure, 148–9, 150–2
- CCTGA, 160–1, 163

- tricuspid regurgitation (TR) (cont.)  
 Ebstein anomaly, 70, 71, 72, 74, 75–6, 77–8  
 pulmonary hypertension, 306–7, 316
- tricuspid valve (TV). *See also* Ebstein anomaly  
 CCTGA abnormalities, 161  
 congenital defects, 70, 71, 75–6
- triple therapy, pulmonary arterial hypertension, 308
- trisomy 13, 14–15
- trisomy 18, 14–15
- trisomy 21. *See* Down syndrome
- truncus arteriosus (TA), 178  
 anatomic characteristics, 178, 179  
 anesthetic implications  
 anomalies associated with 22q11DS, 181, 182  
 endocarditis prophylaxis, 183  
 intraoperative anesthetic plan, 183–4  
 postoperative care, 184  
 preoperative evaluation, 182  
 RV–PA conduit stenosis  
 management, 183–4  
 associated anomalies, 179–80  
 classifications  
 Collett and Edwards, 178–9  
 modified Van Praagh, 178–9  
 coronary blood flow and myocardial perfusion, 180, 181  
 incidence, 178, 179–80  
 natural history, 180, 181  
 oxygen saturation, 180  
 pulmonary and systemic blood flow  
 distribution, 180  
 surgical repair, 181  
 postoperative complications and long-term sequelae, 181
- tunnel-type subvalvular aortic stenosis, 98, 99
- Turner syndrome, coarctation of the aorta, 121
- TV. *See* tricuspid valve
- 22q11.2 microdeletion syndrome (22q11DS), 14–15. *See also* DiGeorge syndrome  
 anomalies associated with, 181, 182  
 TOF with, 41, 47–8  
 TOF/PA/MAPCAs with, 61  
 truncus arteriosus with, 179–80, 181, 182
- two-ventricle repair, Ebstein anomaly, 72, 77
- unbalanced common atrioventricular septal defect, 25  
 anatomic characteristics of complete AVSD, 25, 26  
 anesthetic implications  
 arterial and central line monitoring, 29  
 intraoperative hypotension  
 considerations, 30, 31  
 laparoscopic surgery considerations, 29–30  
 medications for anesthesia induction and maintenance, 30  
 patients with trisomy 21, 28–9  
 postoperative considerations, 31  
 preoperative diagnostic tests, 29  
 pulmonary hypertension risk, 28–9  
 ventilation management, 30  
 cardiac surgical treatment options, 28  
 degree of unbalance in, 28  
 implications, 27  
 long-term survival expectations, 28  
 L-to-R shunting, 25–6  
 neonatal symptoms of complete AVSD, 27  
 physiologic issues of complete AVSD, 25  
 Rastelli classification, 26  
 right dominance versus left dominance, 28  
 trisomy 21 relationship, 27  
 ventricular dominance, 28
- unifocalization, aortopulmonary collaterals, 58–60
- VACTERL association, 48, 359  
 anesthetic implications  
 airway and ventilation management, 362–3, 364  
 cardiorespiratory compromise, 364  
 complications, 365  
 extubation, 365  
 hypoxemia management, 365–6  
 induction and intubation technique, 363  
 monitoring, 364  
 open TEF repair, 364–5  
 postoperative care, 365  
 preoperative bronchoscopy, 363  
 preoperative workup, 362  
 thoroscopic TEF repair, 364  
 cardiac anomalies, 359  
 definition, 359  
 esophageal atresia/tracheoesophageal fistula, 359–60, 361  
 TOF, 360–1, 362, 365–6
- VAD. *See* ventricular assist device
- valvular aortic stenosis. *See* aortic stenosis
- valvulopathies. *See also specific valves*  
 hemodynamic goals, 371
- vascular rings, 338  
 anesthetic implications  
 airway management, 342–3  
 emergence and extubation plan, 343  
 induction method, 342–3  
 management goals, 342  
 monitoring, 342–3  
 multiple procedures, 341–2  
 postoperative care, 343  
 preoperative evaluation, 341  
 respiratory infection considerations, 341  
 associated lesions, 340  
 clinical presentation, 339–40
- definition and anatomy, 338  
 diagnosis, 340  
 embryopathogenesis, 339  
 incidence, 338–9  
 outcomes, 343  
 surgical repair, 340  
 monitoring, 342–3  
 types, 339
- vasopressin, HCM, 119
- veno-arterial ECMO, 263, 264, 266–7
- veno-venous ECMO, 263, 264, 266–7, 281–2
- ventilation strategy  
 AVSD, 30  
 BDG shunt, 204–5  
 diaphragmatic hernia, 262  
 Ebstein anomaly, 73–4, 81  
 ECMO, 264–5  
 Fontan procedure patients, 212, 224, 234  
 HCM, 120  
 lung transplantation, 288  
 pentalogy of Cantrell, 68  
 pulmonary hypertension, 312–13, 328  
 pulmonary overcirculation and, 12  
 pulmonary stenosis, 37  
 stage I palliation, 194  
 TAPVR, 175  
 TGA, 144  
 TOF/APVS, 53–4, 55–6  
 VACTERL association, 362–3, 364
- ventricular assist device (VAD)  
 anesthetic implications, 273  
 anticoagulation, 271–2, 274, 279, 280  
 complications, 271, 273  
 decreased blood flow considerations, 281  
 device management, 273–4  
 expected surgical procedures, 272  
 hematologic considerations, 274, 280  
 hemodynamic considerations, 274, 281  
 infection control, 274  
 invasive line placement, 274  
 management plan, 272, 280  
 patient transport, 279–80  
 preanesthesia assessment, 272  
 respiratory support, 281–2
- biventricular, 279  
 classifications  
 axial pump, 268, 277  
 centrifugal pump, 268, 277  
 continuous flow pumps, 268, 277  
 extracorporeal, 277  
 intracorporeal, 277  
 long-term, 277  
 paracorporeal pump, 268  
 pulsatile pumps, 268, 277  
 short-term, 277  
 components, 268  
 definition, 268  
 devices  
 Berlin Heart EXCOR, 268–9, 270–2, 273–4, 275, 277

- CentriMag, 268, 278, 279
- HeartMate 3, 268
- HeartMate II, 268, 277
- HeartWare, 268, 277, 278
- Total Artificial Heart, 268
- ECMO versus, 264, 276
- indications, 270–1
- left, 268, 271
  - Berlin Heart EXCOR, 268–9, 270
  - right ventricular failure, 278, 279
  - right ventricular function, 277
- loss of device output, 273
- outcomes, 271
- pediatric design challenges, 270
- right, 276, 278, 279
  - decreased blood flow considerations, 281
  - indications, 279
  - respiratory support, 281–2
- selection, 268
- ventricular dysfunction
  - anesthetic risk, 6–7, 79
  - Fontan failure, 229
- ventricular inversion, 159, 160. *See also*
  - congenitally corrected
  - transposition of the great arteries
- ventricular septal defect (VSD), 9
  - anesthetic implications
  - anesthetic plan, 13
  - diuretic therapy issues, 12
  - postoperative monitoring, 13
  - preoperative considerations, 11–12
  - PVR and SVR effects, 12
- CCTGA, 161, 163
- characterization, 9, 10
- complete AVSD with, 25, 26
  - degree of shunting, 11
  - DORV compared with, 14
  - doubly committed, 15–16
  - great artery relationships to, 14, 15, 16
  - hemodynamic effects, 9
  - inlet, 9, 10, 25, 26
  - malalignment, 40
  - muscular, 9, 10
  - noncommitted, 15–16
  - nonrestrictive, 10, 25–6
  - parallel circulations, 4
  - perimembranous, 9
  - pulmonary atresia/MAPCAs and, 58, 59
  - pulmonary overcirculation, 11, 12
  - pulmonary stenosis with, 15–16, 17–18
  - $Q_p:Q_s$  ratio, 10–11
  - repair age and considerations, 11
  - restrictive, 10, 25–6
  - restrictive compared with
    - nonrestrictive, 10
  - subaortic, 15–16, 17
  - subarterial/subpulmonic, 9, 10, 15–16
  - TGA with, 138, 139, 141
  - TOF with, 3, 39–40
  - tricuspid atresia with, 207–8
- ventricular tachycardia, after TOF repair, 46–7, 50–1
- ventriculoarterial alignment, DORV
  - concordant, 15–16
  - discordant, 15–16
- ventriculoarterial discordance, 137–8, 158, 159, 217
- ventriculotomy, TOF repair, 46
- visceral situs, 2
- volatile agents
  - long QT syndrome, 378
  - pulmonary hypertension effects, 312
  - PVR and cerebral perfusion effects, 336
- VSD. *See* ventricular septal defect
- VSD-type DORV, 15–16, 17
- WHO. *See* World Health Organization
- Williams syndrome (WS), 34–5, 105
  - anesthetic considerations, 107–11
  - anesthetic risk, 6–7, 107, 111
  - cardiac abnormalities, 105–6
  - cardiac arrest, 107
  - cardiac surgical repair indications, 107
  - coronary artery abnormalities, 105–6, 107, 108, 111
  - elastin deficiency in, 105
  - hemodynamic management, 109
  - physical characteristics, 107–8
  - QT prolongation, 106
- Williams–Beuren syndrome. *See* Williams syndrome
- Wolff–Parkinson–White syndrome (WPW), 71, 74, 160–1
- Wood units (WU), 305
- World Health Organization (WHO),
  - pulmonary hypertension classification, 306, 308, 333
- WPW. *See* Wolff–Parkinson–White syndrome
- WS. *See* Williams syndrome
- WU. *See* Wood units
- z-score, 6, 387

