Index

ABCC6 (multi-drug resistance associated protein 6), 175
abscesses
  aspergillosis, 14
  HHT/PAVM, 129–130, 132
  infective endocarditis, 68
ACE gene polymorphism, 626
N-acetylaspartate (NAA), 161–162
acroparesthesias, 159
activated protein C (APC)
  deficiency, 338–339
  in DIC/sepsis, 324
  factor V resistance to, 339, 450–455
acute febrile neutrophilic dermatosis, *see* Sweet syndrome
acute posterior multifocal placoid pigment epitheliopathy (APMPPE), 275–286
  association with other disorders, 275, 282–283
  clinical features, 283–284
  CNS disease, 276–281, 283, 284–286
acute renal failure, 469
acyclovir, 72–73
Adams–Oliver syndrome, 120
ADAMTS-13, 337
  DIC, 320
  TTP, 348, 350, 351
adenocarcinomas, 439
ADPKD (autosomal dominant polycystic kidney disease), 215
agalsidase alpha/beta (enzyme replacement therapy), 164, 165, 166
aging, premature, *see* progeroid syndromes
AIDS, 52
  see also HIV (human immunodeficiency virus)
air embolisms, 120, 130
albendazole, 24
albuminuria, 472
alcohol, 431, 432–433
ALK1/activin receptor-like kinase 1, 120, 128
alloimmunization, 154
alopecia, 140, 141, 187
α1-antitrypsin, 515
Alzheimer disease, 538
amaurosis fugax, 89, 518
American trypanosomiasis, *see* Chagas disease
ε-aminocaproic acid, 324, 332
amiodarone, 48–49
amnesia, 584
amphetamines, 429–430
amyloid-β (Aβ) peptide, 534–535, 538, 540
amyloid-β-related angiitis, 78, 541
  see also cerebral amyloid angiopathy
ANCA-associated vasculitis, *see* eosinophilic granulomatosis with polyangiitis; microscopic polyangiitis
andexanet alfa, 330
anemia, 437
aneurysms
  ADPKD, 215
  arterial dissection, 530
  Behçet disease, 107, 394
  cardiac, 47, 567
  Ehlers–Danlos syndrome, 182–183, 217
  familial, 219–223
    differences from sporadic aneurysms, 221
    epidemiology, 215, 219–221
    genetics, 216, 222
    non-genetic risk factors, 219, 222–223
    screening for those at risk, 215, 217, 221–222, 223
  FMD, 577
  HIV-associated, 56–57
  Kawasaki disease (coronary artery), 38, 39–41
  Loeys–Dietz syndrome, 217
  Marfan syndrome, 172, 217
  mycotic, 14, 24, 66, 68
  neurofibromatosis type 1, 218, 246
  pathogenesis, 218–219
  polyarteritis nodosa, 367, 397
  PXE, 178
  Takayasu disease, 99–100
  VHL disease, 211
  VZV vasculopathy, 72
angiitis, *see* vasculitis
angiogenesis in moyamoya disease, 549–550
angiography
  APMPPE (retinal), 275
  arterial dissections, 514, 521–522
    carotid artery redundancy, 513
  arteriovenous malformations
    in HHT, 130, 131
    pulmonary, 123–124
    retinal, 255
  CADASIL, 136
  dilatative arteriopathy, 161, 560
  Divry–van Bogaert syndrome, 259
  developmental venous anomalies, 238
  Eales disease (retinal), 273
  Ehlers–Danlos syndrome
    aneurysm, 182, 183
    carotid-cavernous fistula, 183
  Fabry disease, 161
  FMD, 514, 576, 578
  HERNS (retinal), 299
  HIV-associated vasculopathy, 58
  Menkes disease, 252
  moyamoya disease, 552
  mycotic aneurysms, 68
  neurofibromatosis type 1, 246
  PACNS, 81, 601
  polyarteritis nodosa (visceral), 367
  in pregnancy, 615
  PXE (retinal), 176
  RCVS, 600–601, 611
    with TTP, 351
  Sneddon syndrome, 485–486
  Sturge–Weber syndrome, 203
  Susac syndrome (retinal), 293–294
  syphilis, 2–3
  Takayasu disease, 98
  tuberculous meningitis, 9–11
  VHL disease, 210
angioid streaks, 176
angiokeratomas, 159
angiokeratoma corporis diffusum, *see* Fabry disease
angioma, intracranial, *see* Sturge–Weber syndrome
anhidrosis, 159
ankylosing spondylitis, 402
anterior ischemic optic neuropathy, *see* optic neuropathy
anti-beta-2-glycoprotein 1 antibodies, 305, 308, 311
antibiotics
  Marfan syndrome, 173
  mycotic aneurysms, 67
  syphilis, 4–5
  tuberculosis, 11
anti-cardiolipin antibodies, 305, 306, 308, 310, 456
antidepressants, 542, 603
anti-endothelial cell antibodies, 290–291
antiepileptic drugs, 236, 475, 593, 594
antifibrinolytic agents, 324, 332
antineutrophilic cytoplasmic antibodies (ANCAs)
  eosinophilic granulomatosis with polyangiitis, 378
  microscopic polyangiitis, 370, 373
antiphosphatidyl serine antibodies, 308
antiphospholipid syndrome (APS), 305–313, 389–391
  and cancer, 437
  catastrophic APS, 310, 323, 390, 391
  in children, 391
  CNS disease
    ICH, 390
    ischemic stroke, 306–309, 389–390
    venous thrombosis, 309, 390
    white matter disease, 310
  diagnosis, 305–306, 389, 390
  and IBD, 456
  and infections, 54, 312
  pathology, 310–312, 390

(APS) (cont.)
in pregnancy, 305, 389
and SLE, 305–306, 309, 384
and Sneddon syndrome, 309–310, 402, 483, 484
thrombosis risk, 306, 307–308
treatment, 312–313, 391, 486
antiprothrombin antibodies, 484
antithrombin (AT), 337
deficiency, 338, 456
in DIC, 318, 323
antithrombotic therapy
amyloid angiopathy, 541–542
anticoagulant-associated ICH
antiplatelet agents, 330–331
heparin, 330
oral agents, 328–330, 541
reversal, 329–330, 331, 332
thrombolytic agents/t-PA, 331–332
antiphospholipid syndrome, 312–313, 391, 486
arterial dissections, 528
Behçet disease, 105–106, 108, 394
CADASIL, 138
cancer patients, 442
cavernous malformations, 235
Chagas disease, 49
cryptogenic strokes/PFOs, 569–570
CVTs, 593–594, 614
DIC, 323
Fabry disease, 164–165
giant cell arteritis, 396
HIT, 323, 342–343
IBD, 449, 456
infective endocarditis, 65
Kawasaki disease, 41
nephrotic syndrome, 474–475
progeria, 191
sickle cell disease, 151
SLE, 386, 392
Sneddon syndrome, 486
Sturge–Weber syndrome, 205
Susac syndrome, 296
Takayasu disease, 99
TTP, 352
anti-TNF-α agents
antiphospholipid syndrome, 296
Behçet disease, 108
sarcoidosis, 117
Takayasu disease, 99
antiviral agents, 72–73, 369, 395
aortic arch syndrome
definition, 96
Takayasu disease, 96–100, 402–403
classification, 97
clinical features, 98, 402
diagnosis, 98–99, 403
epidemiology, 96
pathology, 96–97
strokes, 98, 99
treatment, 99–100, 403
aortic dilatations (in Marfan syndrome), 171, 172–173
aortic dissections, 581–585
clinical features, 582–583
diagnosis, 584–585
epidemiology, 582
location, 581–582
neurological disease, 581–582, 583–584, 585
treatment, 585
see also cervicocephalic arterial dissections
apathy, 135
apheresis, *see* plasmapheresis
apixaban, 313, 329, 442
reversal of effect, 330
APMPPE, *see* acute posterior multifocal placoid pigment epitheliopathy
apolipoprotein E (APOE), 534–535, 540
apoptosis, 420–421
APS, *see* antiphospholipid syndrome
L-arginine, 197–198
arterial dissections, *see* aortic dissections; cervicocephalic arterial dissections
arteriovenous malformations (AVMs)
intracranial
Behçet disease, 107
HHT, 128, 130–131, 132
Wyburn–Mason syndrome, 257
maxillofacial, 255, 256–258
pulmonary, *see* pulmonary arteriovenous malformations
retinal/orbital, 255–256, 258
spinal, 130, 131, 479–480
Wyburn–Mason syndrome, 255–258
arthritis
ankylosing spondylitis, 402
in IBD, 448
rheumatoid, 399–400
Aspergillus infections, 13–15
aspirin
giant cell arteritis, 396
ICHs related to use, 330–331
reversal of effect, 331
infective endocarditis, 65
Kawasaki disease, 41
nephrotic syndrome, 474
SLE, 386
Sturge–Weber syndrome, 205
Takayasu disease, 99
TTP, 352
asthma, 378
atherosclerosis
in HIV patients, 59
and hyperviscosity, 412–413
nephrotic syndrome, 472
and radiotherapy, 502
and SLE, 385, 386
ATP7A/ATP7A (Cu transporter), 251, 252
atrial fibrillation
in CAA patients, 541
IBD, 456
sarcoidosis, 114
scleroderma, 496
atrial septal aneurysms, 567
autoimmune diseases
antiphospholipid syndrome, *see* antiphospholipid syndrome
association with APMPPE, 282
Behçet disease, 102
Cogan syndrome, 302
GPA, 395–396
idiopathic thrombocytopenic purpura, 334
microscopic polyangiitis, 369–374
paraneoplastic syndromes, 437–438
scleroderma, 401–402, 495–496
Sjögren syndrome, 392–393
Susac syndrome, 290–291
systemic lupus erythematosus, *see* systemic lupus erythematosus
TTP, 348
autosomal dominant polycystic kidney disease (ADPKD), 215
AVMs, *see* arteriovenous malformations

B cells
lymphomas, 630–632
lymphomatoid granulomatosis, 506–507
baldness, premature, 140, 141, 187
Bartleson syndrome, 625
basilar artery
dilatative arteriopathy, 560
Fabry disease, 160–161, 162
Bean syndrome (blue rubber bleb nevus syndrome), 268–270
Behçet disease, 102–108, 393–394
causes, 102–103
in children, 394
clinical features
cerebrovascular disease, 104–107, 394
meningoencephalitis, 103–104
diagnosis, 393, 465
pathology, 103
treatment, 105–106, 107–108, 394
benznidazole, 47–48
Bernard–Soulier syndrome, 333
beta-blockers, 172, 173
bevacizumab, 131, 440
Binswanger disease, 412
biopsy
CAD, 525–526
CADASIL (skin), 137
Divry–van Bogaert syndrome (skin), 259–260, 264
giant cell arteritis, 91–92
microscopic polyangiitis, 373
neurosarcoidosis, 113
PACNS, 81–83, 398
polyarteritis nodosa, 366–367
scleroderma, 496
Sneddon syndrome (skin), 484, 486
Susac syndrome, 291
blastomycosis, 17
bleeding disorders, *see* coagulopathies
blood–brain barrier, 26
blood pressure, *see* hypertension; hypotension
blood transfusions, 151, 152, 153–155
blood viscosity, 408–409
blue rubber bleb nevus syndrome (BRBNS), 268–270
bone disorders, 489–493
Camurati–Engelmann disease, 491–492
CARASIL, 141, 144
craniosynostosis, 491
epidermal nevus syndrome, 480
fibrocartilaginous embolism, 490
low bone mineral density, 489–490
Marfan syndrome, 170
osteochondromas, 492
osteopetrosis, 490–491
osteoporosis, 421
osteoprotegerin, 490
Paget disease, 491
periodontal disease, 492–493
progeria, 187–188
Bonnet–Dechaume–Blanc syndrome (Wyburn–Mason syndrome), 255–258
borreliosis, *see* Lyme disease
brain network disruption, 539
brainstem pathology
basilar artery occlusion, 519
Behçet disease, 103–104, 107
CARASIL, 141
cysticercosis, 21
dilatative arteriopathy, 561
BRBNS, *see* blue rubber bleb nevus syndrome

C-reactive protein (CRP), 91
CAA, *see* cerebral amyloid angiopathy
CAD, *see* cervicocephalic arterial dissections
CADASIL, 134–138

clinical features, 134–135
diagnosis, 135–136, 137, 138
epidemiology, 134
genetics, 134–135, 137–138
migraines, 134, 138, 625–626
pathology, 136–137
treatment, 138
calcifications
BRBNS, 268
calciphylaxis in renal disease, 423–424
embolism from heart valves, 423–424
homocysteinemia, 340
PXE, 175, 177
Sturge–Weber syndrome, 203, 204
calcium, 418
association with stroke, 418–422, 423–424
dietary, 424
calcium-channel antagonists, 296, 418–419, 422–423, 578
calf vein DVT, 568
Call–Fleming syndrome, *see* reversible cerebral vasoconstriction syndrome
Camurati–Engelmann disease, 491–492
cancer
causes of strokes, 437
autoimmunity, 437–438
direct effects, 439–440
hypercoagulability, 439
iatrogenic, 440, 502–503
clinical features of strokes, 440–441
and DIC, 321–322
hyperviscosity in leukemia, 411, 413
and ICHs, 332–333, 440–441
investigations of stroke, 441–442
lymphomas, 78, 630–632
paraneoplastic syndromes, 437
prevention of strokes, 442
Sweet syndrome, 462
Candida infections, 13, 15–16
cannabis, 431–432
CARASIL, 140–144
clinical features, 140–141
diagnosis, 141
genetics, 142–144
pathology, 142
treatment, 144
cardioembolic strokes
antiphospholipid syndrome, 312, 390
Behçet disease, 106
candidiasis, 16
Chagas disease, 46–47
Fabry disease, 163
HIV-associated, 54
infective endocarditis, 64–65
Marfan syndrome, 171–172
neoplastic, 440
sarcoidosis, 117
SLE, 383–384, 392
valvular calcifications, 423–424
cardiovascular disease
and alcohol, 432
Chagas disease, 45–46, 47, 48–49
cocaine, 430
dyslipidemia, 472
Fabry disease, 163, 165
in HIV patients, 54, 59, 60
hypereosinophilia, 498–499
hyperviscosity, 412–413
IBD, 456
infective endocarditis, *see* infective endocarditis
Kawasaki disease, 38, 39–41
Marfan syndrome, 171, 172–173
peripartum cardiomyopathy, 614–615
progeria, 188
PXE, 176–177
radiotherapy, 502
rheumatoid arthritis, 399, 400
sarcoidosis, 114–117
scleroderma, 496
SLE
atherosclerosis, 385, 386
endocarditis, 383, 386
Sneddon syndrome, 483, 484–485, 486
carotid artery
dissections (ICAD)
clinical features, 509, 516–518, 519
imaging, 513, 521, 525
incidence, 510
intracranial, 519
prognosis, 528–529
redundancy, 513
traumatic, 510–511, 529
Ehlers–Danlos syndrome
aneurysms, 182–183
carotid-cavernous fistulae, 183–184
Fabry disease, 163
PXE, 177
see also fibromuscular dysplasia; moyamoya disease
catastrophic antiphospholipid antibody syndrome, 310, 323, 390, 391
cathinone, 430
cavernous malformations (CMs), 227–237
clinical features, 231–233
diagnosis, 228–230
and DVA, 227, 238
epidemiology, 230–231
hemorrhage risk, 233–234
during pregnancy, 235–236
pathogenesis, 227–228
prognosis, 237
radiotherapy-induced, 503
treatment, 234–237
CBF, *see* cerebral blood flow
CCM1/CCM2/CCM3, 227–228, 235
cell death, 420–421
cerebellum
DVAs, 237, 238, 239
hemangioblastomas, 210–211
Menkes disease, 253
cerebral amyloid angiopathy (CAA), 534–542
amyloid-β-related angiitis, 78, 541
clinical features, 540–541
diagnosis (Boston criteria), 539–540
epidemiology, 535, 540
imaging, 535–539
intracerebral hemorrhages, 535, 540
management of acute ICHs, 541
prevention, 541–542
microbleeds, 538, 540–541, 542
pathology, 534–535
treatment, 541–542
cerebral atrophy
amyloid angiopathy, 539
sickle cell disease, 151
Sturge–Weber syndrome, 204
cerebral blood flow (CBF)
amyloid angiopathy, 539
and aneurysm development, 218
Behçet disease, 104
Chagas disease, 47
craniosynostosis, 491
cysticercosis, 24
dilatative arteriopathy, 560
migraine, 622
RCVS, 601
sickle cell disease, 147, 148, 151–152
Sneddon syndrome, 486
Sturge–Weber syndrome, 202–203
Takayasu disease, 97
cerebral venous (sinus) thrombosis (CVT/CVST), 589–594
anatomy of venous system, 589
antiphospholipid syndrome, 309, 390
Behçet disease, 104–106, 108, 394
BRBNS, 268
causes/risk factors, 472, 590
clinical features, 591
diagnosis, 591–592, 614
DIC, 322
epidemiology, 309, 589, 614
IBD, 449
nephrotic syndrome, 474
paraneoplastic, 440
pathology, 337, 589
in pregnancy/postpartum, 594, 607, 613–614
prognosis, 592
sarcoidosis, 114
Sjögren syndrome, 393
SLE, 385, 392, 474
Sturge–Weber syndrome, 202
treatment, 108, 394, 592–594, 614
tuberculosis, 7–8
VZV vasculopathy, 72
cerebromeningeal angiomatosis, 259
cerebroretinal vasculopathy (CRV), 298, 301
cerebrospinal fluid (CSF)
amyloid angiopathy, 540
APMPPE, 283, 285
Behçet disease, 103, 104, 107
CADASIL, 136
HaNDL, 625
Kawasaki disease, 39
Lyme disease, 29
PACNS, 79, 398
pneumococcal meningitis, 27
Susac syndrome, 294
Sweet syndrome, 465
syphilis, 1–2, 3, 4, 5
VZV vasculopathy, 72
cervical manipulative therapy, 511
cervical spinal cord, 3, 519
cervicocephalic arterial dissections (CADs), 509–530
clinical features, 509, 516–519
connective tissue diseases, 514–515, 525–526
Ehlers–Danlos syndrome, 184
Marfan syndrome, 171
PXE, 178
diagnosis, 509, 519–526
dilatative arteriopathy, 561
epidemiology, 510, 515–516
in FMD, 577
intracranial, 519, 521–523, 528, 529
pathology, 509, 510–516
prognosis, 528–530
spontaneous, 509–510, 512–516, 525–526
strokes, 509, 517–518, 519, 526–528, 529–530
traumatic, 509–511, 529
treatment, 526–529
see also aortic dissection
Chagas, Carlos, 44
Chagas disease, 44–49
cardiovascular disease, 45–46, 47, 48–49
clinical features, 45–46
CNS disease, 46–47, 49
diagnosis, 46
epidemiology, 44–45

chaperone molecules, 165
chemotherapy
  as a cause of stroke, 440
  lymphomas, 631–632
  lymphomatoid granulomatosis, 507
chickenpox (varicella), 72
  see also varicella zoster virus (VZV) vasculopathy
children
  antiphospholipid syndrome, 391
  arterial dissections, 516
  Behçet disease, 394
  cerebral aneurysmal arteriopathy in AIDS, 56
  craniosynostosis, 491
  diffuse neonatal hemangiomatosis, 270
  dilatative arteriopathy, 561–562
  FMD, 575–576
  hemolytic uremic syndrome, 350
  Henoch–Schönlein purpura, 356–357, 400–401
  Kawasaki disease, 38–41, 403
  moyamoya disease, 545, 550, 551, 553
  nephrotic syndrome, 469, 470, 473, 474
  neurofibromatosis type 1, 246
  PACNS, 398–399
  PAVM screening, 125
  polyarteritis nodosa, 397
  progeroid syndromes, 187–191
  radiotherapy, 503
  sickle cell disease
    silent cerebral infarctions, 148, 149, 150–151
    stroke risk, 147, 148
    treatment/prevention of strokes, 151–155, 413
  SLE, 392
  Sturge–Weber syndrome, 200–205
  Sweet syndrome, 462, 466
  thrombophilia, 343
  varicella, 72
chiropractic, 511
chronic kidney disease, 469
chronic mucocutaneous candidiasis, 16
ciaraparantag, 330
cigarette smoking, 219, 222–223, 412, 433
cilostazol, 144
clubbing, 121
CMs, *see* cavernous malformations
coagulation
  laboratory tests
    DIC, 317–318
    Henoch–Schönlein purpura, 400
    INR, 329
    for thrombophilia, 344
  physiology, 320, 336–337, 347–348, 419–420
  and warfarin, 329
coagulopathies, 328–334
  and antiplatelet agents, 330–331
  characteristics of ICHs, 328, 329, 331
  hemophilia, 333
  and heparin, 330
  and leukemia, 332–333
  and oral anticoagulants, 328–330
  platelet disorders, 333–334
  treatment
    inherited disorders, 333
    leukemias, 332
    reversal of antithrombotic therapies, 329–330, 331, 332
  von Willebrand disease, 333
  see also thrombophilia
cocaine, 398, 430–431
coccidioidomycosis, 17
Cogan syndrome, 302–304
  clinical features, 302–303
  diagnosis, 303
  pathology, 302
  treatment, 303–304
cognitive impairment
  amyloid deposition, 538
  CADASIL, 135, 138
  CARASIL, 140–141
  Chagas disease, 49
  epidermal nevus syndrome, 479
  Fabry disease, 160
  PACNS, 78
  radiotherapy, 503
  Sneddon syndrome, 309–310, 483
  Sturge–Weber syndrome, 202
  Susac syndrome, 292, 293
COL3A1/type III collagen, 181, 217
coma
  aortic dissections, 583
  CVTs, 591, 593
computed tomography, *see* CT scans
congenital contractural arachnodactyly, 172
copper in Menkes disease, 251–252, 253
corneal whirls (cornea verticillata), 159
coronary artery disease
  Fabry disease, 163
  Kawasaki disease, 38, 39–41
  PXE, 177
corpus callosum, 293
cortical spreading depressions, 622
cortical superficial siderosis, 538, 540
corticosteroids
  APMPPE, 286
  Cogan syndrome, 304
  cysticercosis, 24
  Eales disease, 274
  eosinophilic granulomatosis with polyangiitis, 379
  giant cell arteritis, 89, 92–93, 396
  Henoch–Schönlein purpura, 401
  HERNS, 301
  Kawasaki disease, 41
  nephrotic syndrome, 474
  neurosarcoidosis, 117
  PACNS, 83–84, 399
  polyarteritis nodosa, 363, 369, 397
  Susac syndrome, 295
  Sweet syndrome, 462, 464–465, 466
  Takayasu disease, 99, 403
  TTP, 351
  tuberculous meningitis, 11
  VZV vasculopathy, 72–73
cranial nerve disease
  carotid artery dissection, 517, 518
  dilatative arteriopathy, 561
  Fabry disease, 163–164
  giant cell arteritis, 89, 92, 396
  orbital AVMs, 256
  polyarteritis nodosa, 362
  syphilis, 2
craniocervical dislocation in rheumatoid arthritis, 400
craniosynostosis, 491
Crohn disease, *see* inflammatory bowel disease
CRP (C-reactive protein), 91
cryoglobulinemia, 394–395
cryoprecipitate, 332
cryosupernatant plasma, 351
cryptococcal infections, 13, 16–17
cryptogenic strokes, 566
  and DVTs, 568–569
  and PFOs, 308–309, 566, 567–568
  treatment, 569–571
  see also paradoxical embolisms
CSF, *see* cerebrospinal fluid
CT scans (computed tomography)
  arterial dissection, 524–525
  Behçet disease, 105
  CARASIL, 141
  CVTs, 591
  Fabry disease, 160
  HIV patients, 58, 59
  PAVM, 123
  in pregnancy, 615
  sickle cell disease, 150
  Sturge–Weber syndrome, 203
  subarachnoid hemorrhages, 220
  tuberculous meningitis, 9
  VHL disease, 208
Cunninghamella bertholletiae, 15
cutaneous lesions, *see* skin lesions
CVT/CVST, *see* cerebral venous (sinus) thrombosis
cyclophosphamide, 84, 373, 379
cysticercosis, *see* neurocysticercosis
cytokines
  aneurysm development, 218
  Behçet disease, 102–103, 108
  moyamoya disease, 550
  polyarteritis nodosa, 368
  sarcoidosis, 112
  tuberculosis, 8

D-dimer
  cancer, 441, 442
  CVTs, 592
dabigatran, 313, 329, 442, 542
  reversal of effect, 330
deep venous thromboses (DVTs)
  IBD, 449
  and PFOs/paradoxical embolisms, 384, 439, 568–569
  SLE, 384
Degos disease (malignant atrophic papulosis), 445–447
dementia
  amyloid deposition, 538
  CADASIL, 135, 138
  CARASIL, 140–141
  Fabry disease, 160
  radiotherapy, 503
  Sneddon syndrome, 309–310, 483
  Susac syndrome, 293
  see also cognitive impairment
dendritic cells, 87–88
designer drugs, 430, 431
developmental venous anomalies (DVAs), 237–240
  and cavernous malformations, 227, 238
  clinical features, 238–239
  diagnosis, 237–238
  epidemiology, 238
  hemorrhage risk, 240
  treatment, 240
dexamethasone, 11, 24
diabetes mellitus, 412, 472
dilatative arteriopathy (dolichoectasia), 560–563
  Fabry disease, 162, 163–164
disseminated intravascular coagulation (DIC), 317–324
  associated conditions, 317
  clinical features, 321–322
  diagnosis, 317–318
    differentials, 322
  epidemiology, 318–320
  pathogenesis, 320–321
  treatment, 323–324

distal motor neuropathy, ATP7A-related, 251
Divry–van Bogaert syndrome, 259–267
  clinical features, 259, 261
  diagnosis, 259–264
DNA repair abnormalities, 190
dolichoectasia, 560–563
  Fabry disease, 162, 163–164
donepezil, 138
Doppler ultrasonography, *see* ultrasonography
drug abuse, *see* substance abuse
DVAs, *see* developmental venous anomalies
DVT, *see* deep venous thrombosis
dyslipidemia, 472
dysmorphic features
  Ehlers–Danlos syndrome, 217
  Fabry disease, 158–159
  Menkes disease, 250
  progeria, 187

Eales disease, 273–274, 283
ear/hearing loss
  APMPPE, 283
  Cogan syndrome, 302–303, 304
  pulsatile tinnitus, 517
  Susac syndrome, 291, 292, 294
echocardiography
  Chagas disease, 47
  PAVM, 124
  PFOs, 566–567
eclampsia, 607, 609, 610, 613
  see also pre-eclampsia
ecstasy (MDMA), 430
ectopia lentis, 170
eculizumab, 341, 446
Ehlers–Danlos syndrome
  subtypes, 181
  type IV (vascular), 181–184, 215–217
    arterial dissection, 184
    carotid-cavernous fistula, 183–184
    intracranial aneurysms, 182–183, 217
  type IX (occipital horn syndrome), 251
embolization, therapeutic
  AVMs, 124–125, 258
  carotid-cavernous fistulae, 183–184
  hemangioblastoma, 211
  infective endocarditis, 67
  moyamoya disease, 555
encephalo-duro-arterio-synangiosis (EDAS), 153, 554
encephalofacial angiomatosis, *see* Sturge–Weber syndrome
encephalopathy
  CVTs, 591
  eosinophilic granulomatosis with polyangiitis, 379
  Henoch–Schönlein purpura, 357
  MELAS, 197
  paraneoplastic strokes, 440
  PRES, 612–613
  radiation damage, 503
  Susac syndrome, 292, 293
  see also CADASIL; CARASIL
endocarditis
  hypereosinophilia, 498–499
  infective, *see* infective endocarditis
  Libman–Sacks antiphospholipid syndrome, 312, 390
  cancer, 439
  DIC, 322
  SLE, 383, 386
endothelial progenitor cells, 549–550
endothelium
  Fabry disease, 163
  physiology, 337
  polyarteritis nodosa, 368
  radiation damage, 502
endovascular procedures
  arterial dissections, 527–529, 530
  AVMs, 124–125, 258
  carotid-cavernous fistulae, 183–184
  CVTs, 593
  infective endocarditis, 67
  Marfan syndrome, 173
  moyamoya disease, 555
  in pregnancy, 616
ENG/endoglin, 120, 128
enterocolitis, 16
eosinophilic granulomatosis with polyangiitis (Churg–Strauss syndrome), 378–380, 403, 499
  CNS disease, 379, 499
  diagnosis, 378–379, 403
  treatment, 379
eosinophils, hypereosinophilic syndromes, 498–500
ephedra, 430
ephedrine, 430
epidermal nevus syndrome, 478–480
  CNS disease, 478–480
  diagnosis, 480
  genetics, 480
  other systemic features, 478, 480
epilepsy, *see* seizures
epistaxis, 121, 129
Epstein–Barr virus, 506
ergotamine and derivatives, 626
erythrocyte sedimentation rate (ESR), 91, 98
erythrocytes, *see* red blood cells
erythropoietin, 411
essential thrombocythemia, 341–342, 411, 413
ethanol, 431, 432–433
everolimus, 205
Exserohilum rostratum, 17
eye/loss of vision
  angioid streaks in PXE, 176
  APMPPE, 275, 283–284
  carotid artery dissection, 518
  Cogan syndrome, 302
  CVTs, 591, 594
  diabetic retinopathy, 472
  Eales disease, 273–274, 283
  epidermal nevus syndrome, 480
  Fabry disease, 159
  giant cell arteritis, 89, 92, 396
  HERNS, 298
  Marfan syndrome, 170
  microscopic polyangiitis, 369
  PACNS, 79
  PRES/eclampsia, 612
  Sneddon syndrome, 483
  Sturge–Weber syndrome, 201, 202, 205
  Susac syndrome, 292, 293–294
  Takayasu retinopathy, 96
  Wyburn–Mason syndrome, 255–256, 258

Fabry disease, 158–166
  clinical features, 158–159, 160
  CNS disease, 158, 159–165
  epidemiology, 158, 159–160
  genetics, 158
  prognosis, 164
  treatment, 164–166
facial features
  Ehlers–Danlos syndrome, 217
  Fabry disease, 158–159
  Menkes disease, 250
  progeria, 187
factor V Leiden and other mutations, 163, 339, 450–455
factor VIIa, recombinant, 329
factor VIII
  deficiency (hemophilia A), 333
  elevation (in IBD), 456
factor IX deficiency (hemophilia B), 333
factor Xa inhibitors (rivaroxaban and apixaban), 313, 329, 442
  reversal of effect, 330
factor XI deficiency (hemophilia C), 333
farnesyltransferase inhibitors, 191
fasudil hydrochloride, 419, 422–423
FBN1/fibrillin-1, 170, 173
fenfluramine, 430
fibrin degradation products (FDPs), 318
fibrinogen
  in DIC, 318
  in IBD, 450
  and plasma viscosity, 410
  and stroke risk, 339, 409
fibrinoid necrosis (degeneration), 7
fibrinolysis, 337
  abnormalities, 112, 339, 456
fibrinopeptide A, 450
fibrocartilaginous embolism, 490
fibromuscular dysplasia (FMD), 514, 575–578
  CNS disease, 577–578
  diagnosis, 578
  epidemiology, 575–576
  pathology, 575, 576
  treatment, 578
Five Factor Score, 369
florbetapir, 538
fluorescein retinal angiography
  APMPPE, 275
  AVMs, 255
  Eales disease, 273
  HERNS, 299
  PXE, 176
  Susac syndrome, 293–294
FMD, *see* fibromuscular dysplasia
fresh frozen plasma (FFP), 323, 329, 351
fundoscopy
  angioid streaks, 176
  APMPPE, 275
  Eales disease, 273
  giant cell arteritis, 89
  PACNS, 79
  Susac syndrome, 292
  Wyburn–Mason syndrome, 256, 257
fungal infections, 13
  aspergillosis, 13–15
  candidiasis, 13, 15–16
  cryptococcosis, 13, 16–17
  mucormycoses, 15
  other, 17

α-galactosidase A deficiency, *see* Fabry disease
α-galactosidase A replacement therapy, 164, 165, 166
gastrointestinal tract
  BRBNS, 268
  candidiasis, 16
  Degos disease, 446
  HHT, 129
  IBD, *see* inflammatory bowel disease
  PXE, 177
GCA, *see* giant cell arteritis
gene therapy, Fabry disease, 165–166
genetics
  ADPKD, 215
  amyloid-β, 534–535
  aneurysms (familial), 216, 222
  arterial dissections, 525
  Behçet disease, 102
  BRBNS, 270
  CAD, 512–513
  CADASIL, 134–135, 137–138

genetics (cont.)
Camurati–Engelmann disease, 491
CARASIL, 142–144
cavernous malformations, 227–228, 235
Ehlers–Danlos syndrome, 181, 217
epidermal nevus syndrome, 480
Fabry disease, 158
HERNS, 298, 299
HHT, 119, 120, 128–129
Marfan syndrome, 170
Menkes disease, 252
migraines, 625–626
mitochondrial disorders, 194, 195
moyamoya disease, 549
neurofibromatosis type 1, 245
progeroid syndromes
LMNA, 189–190
WRN, 190
PXE, 175
Sturge–Weber syndrome, 200–201
Takayasu disease, 97
thrombophilia, 338, 339, 340, 341
TTP, 348
VHL disease, 208, 210
giant cell arteritis (GCA), 87–93, 396
and cancer, 438
clinical features, 88–90, 396
diagnosis, 90–92, 396
epidemiology, 87
large vessel disease, 90, 91
pathology, 87–88, 90
VZV, 73–74
prognosis, 93
treatment, 89, 92–93, 396
Glanzmann thrombasthenia, 333
glaucomas, 201, 202, 205
glomerulonephritis, 470, 474
glue sniffing, 432
glutamate, 421
GNAQ/Gαq (G protein), 200–201
granulocyte colony-stimulating factor (G-CSF), 462, 464
granulomas (sarcoidosis), 111–112
granulomatosis with polyangiitis, 395–396
granulomatous angiitis of the CNS, *see* primary angiitis of the central nervous system
Grönblad–Strandberg disease, *see* pseudoxanthoma elasticum
Haemophilus influenza meningitis, 26
hair
loss
CARASIL, 140, 141
progeria, 187
in Menkes disease, 250–251
"halo sign", 91
Harada disease, 275
headache
arterial dissection, 516, 517, 518, 623
CADASIL, 134, 138, 625–626
and calcium, 423
CVTs, 591, 592, 594
DVAs, 239
giant cell arteritis, 88
HaNDL (Bartleson syndrome), 625
migraines, 621–627
moyamoya disease, 551
PACNS, 78
PAVM, 122, 124
RCVS, 599, 601–602, 625
Susac syndrome, 291–292
hearing loss
APMPPE, 283
Cogan syndrome, 302–303, 304
pulsatile tinnitus, 517
Susac syndrome, 291, 292, 294
HELLP syndrome, 323, 607–608
hemangioblastomas, 208–212
hematocrit, 409, 411
hemimegalencephaly, 479
hemispherectomy, 205
hemodynamics, *see* cerebral blood flow
hemoglobinopathies, 147
see also sickle cell disease
hemolytic uremic syndrome, 350
hemophilia, 333
hemoptysis, 121
hemostasis, physiological, 320, 336–337, 347–348
Henoch–Schönlein purpura (HSP), 356–357, 400–401
neurological features, 357, 400
treatment, 357, 401
heparin
Behçet disease, 105–106
cancer patients, 442
CVTs, 593, 614
DIC, 323
ICHs related to use, 330
infective endocarditis, 65
reversal of effect, 330
Takayasu disease, 99
heparin-induced thrombocytopenia (HIT), 323, 342–343
hepatic AVMs, 129
hepatitis B, 359, 361, 368, 369
hepatitis C, 395
hereditary endotheliopathy with retinopathy, nephropathy, and stroke (HERNS), 298–301
clinical features, 298
diagnosis, 298–299
pathology, 299–301
treatment, 301
hereditary hemorrhagic telangiectasia (HHT), 128–132
clinical features, 121, 122, 129–131
diagnosis, 119, 131
epidemiology, 120, 128
genomics, 119, 120, 128–129
pathology, 128
and PAVM, 120, 129–130
screening, 122–123, 125, 129, 132
pregnancy, 122, 132
treatment, 131–132
hereditary vascular retinopathy, 298
HERNS, *see* hereditary endotheliopathy with retinopathy, nephropathy, and stroke
heroin, 429
herpes simplex virus encephalitis, 72
heteroplasmy, 195
Heubner's endarteritis, 2, 16
HHT, *see* hereditary hemorrhagic telangiectasia
histoplasmosis, 17
HIT (heparin-induced thrombocytopenia), 323, 342–343
HIV (human immunodeficiency virus), 52–60
cardiovascular disease, 54, 60
atherosclerosis, 59
and other infections, 54
syphilis, 3
tuberculosis, 8, 9
VZV, 57–59
prothrombotic states, 54–55
in South Africa, 52
and stroke risk, 52–53
and stroke treatment, 59–60
substance abuse, 55
vasculopathy, 55
cerebral aneurysms in children, 56
large- and medium-vessel, 56–59
small-vessel, 56
homocyst(e)inemia, 339–340, 456
homocystinuria, 172, 340
Horner syndrome, 517, 529
Horton, Bayard, 87
HSV-1 encephalitis, 72
HTRA1 (serine protease), 142–144
human leukocyte antigens (HLA), 97, 102
Hutchinson, Jonathan, 87, 187
Hutchinson–Gilford progeria syndrome (HGPS), 187–191, 495
hydroxychloroquine, 313, 392
hydroxyurea, 152–153
hyperbaric oxygen therapy, 296
hypercalcemia, 418–422
hypercoagulability, *see* thrombophilia
hypereosinophilic syndrome (HES), 498–500
see also eosinophilic granulomatosis with polyangiitis
hyperparathyroidism, 421
hyperperfusion syndrome, 555
hypertension
aneurysms/SAHs, 218, 223
arterial dissections, 515
microalbuminuria, 472
in pregnancy, *see* pre-eclampsia
Sneddon syndrome, 483
Takayasu disease, 99
hypertrophic cardiomyopathy, 163
hyperviscosity syndromes, 408–413
association with strokes, 409–410, 412–413
covert syndromes, 412
increased cellularity, 341–342, 411, 413
plasma abnormalities, 410, 412, 413
reduced red cell deformability, 408, 411–412, 413
treatment, 413
viscosity of blood, 408–409
hypomagnesemia, 424–425
hypotension
in aortic dissections, 582, 583
orthostatic, in Fabry disease, 163

IBD, *see* inflammatory bowel disease
ICAD, *see* carotid artery, dissection
ICHs, *see* intracranial hemorrhages
idarucizumab, 330
idiopathic thrombocytopenic purpura, 334
IE, *see* infective endocarditis
IgA vasculitis (Henoch–Schönlein purpura), 356–357, 400–401
neurological features, 357, 400
treatment, 357, 401
imaging
amyloid angiopathy, 535–539
angiokeratomas, 506–507
antiphospholipid syndrome, 310

aortic dissections, 584–585
APMPPE, 275, 276–281
arterial dissections, 509, 514, 520–525
  carotid artery redundancy, 513
Behçet disease, 103–104, 105
CADASIL, 135–136
cancer patients, 441, 442
CARASIL, 141
cavernous malformations, 228–230, 239
cerebral venous anatomy, 589
cerebral venous thrombosis, 474, 590, 591–592, 614
Chagas disease (heart), 47
Cogan syndrome, 303
cysticercosis, 23–24
dilatative arteriopathy, 561, 562
Divry–van Bogaert syndrome, 259
DVAs, 237–238, 239
Ehlers–Danlos syndrome, 182, 183
Fabry disease, 160–162
FMD, 514, 576, 578
giant cell arteritis, 91
Henoch–Schönlein purpura, 401
HERNS, 299
HHT, 123, 130, 131
HIV patients, 59
  vasculopathy, 58
homocysteinemia
  calcifications, 340
hypereosinophilic syndromes, 499
infective endocarditis, 66, 67–68
lymphomas, intravascular, 631
MELAS, 197
Menkes disease, 251
microscopic polyangiitis, 367
migraines, 623, 626–627
moyamoya disease, 550, 551–553
neurofibromatosis type 1, 246
PACNS, 79–81, 600, 601
PAVM, 122–124
polyarteritis nodosa, 367, 368
  in pregnancy, 615
PRES, 612–613
progeria, 189
RCVS, 597, 599–601, 611
sarcoidosis, 114
sickle cell disease, 148, 149, 150–151
SLE, 382–383
Sneddon syndrome, 485–486
Sturge–Weber syndrome, 203
subarachnoid hemorrhages, 220
Susac syndrome, 293–294, 295
Sweet syndrome, 465
syphilis, 2–3
  Takayasu disease, 98
  thrombolysis-related ICHs, 331
  TTP, 350–351
  tuberculous meningitis, 9–11
  VHL disease, 208–210
  VZV vasculopathy, 72
immune reconstitution inflammatory syndrome, 54
immunoglobulin, intravenous (IVIg), 41, 296
  risk factor for strokes, 410
immuno-osseous dysplasia, 491
immunosuppressants/immunomodulators
  Behçet disease, 108, 394
  eosinophilic granulomatosis with polyangiitis, 379
  giant cell arteritis, 93, 396
  GPA, 396
  microscopic polyangiitis, 373–374
  PACNS, 83–84, 398, 399
  polyarteritis nodosa, 397
  sarcoidosis, 117
  SLE, 392
  Susac syndrome, 296
  Takayasu disease, 99
  TTP, 351–352
  see also corticosteroids
immunothrombosis, 320–321
infective endocarditis (IE), 63–69
  epidemiology, 63
  imaging, 66, 67–68
  mycotic aneurysms, 66, 68
  strokes, 16, 64–65, 429
  valve surgery, 65–66
  vegetations, 63–64
inflammatory bowel disease (IBD), 448–457
  clinical features, 448
  epidemiology, 448
  extra-intestinal complications, 448–449
  hypercoagulability, 449–456
  pathogenesis, 448
  strokes, 449–456
  and Sweet syndrome, 462
  treatment, 449, 456
infliximab, 117, 296
interleukins (IL), 8, 112, 218, 368
international normalized ratio (INR), 329
interstitial keratitis, 302
intracranial hemorrhages (ICHs)
  amyloid angiopathy, 535, 540, 541
  antiphospholipid syndrome, 390
  aspergillosis, 14
  Behçet disease, 107, 394
  brain damage caused by, 328, 329, 331
  cancer, 332–333, 440–441
  candidiasis, 16
  cavernous malformations, 231–233, 237
    hemorrhage risk, 233–234, 235–236
  coagulation factor deficiencies, 333
  cysticercosis, 21–22
  DIC, 322–323
  drug abuse, 429, 430
  DVAs, 238–239, 240
    hemorrhage risk, 240
  eosinophilic granulomatosis with polyangiitis, 379
  Fabry disease, 164
  Henoch–Schönlein purpura, 357
  HHT, 122, 130–131
  infective endocarditis, 65
  migraine, 621, 623
  moyamoya disease, 550, 554–555
  PACNS, 398, 399
  paraneoplastic strokes, 440–441
  platelet disorders, 332, 333–334
  polyarteritis nodosa, 363, 397
  in pregnancy, 235–236, 606–607, 609
  RCVS, 599
  sarcoidosis, 114
  sickle cell disease, 149, 151
  Sjögren syndrome, 393
  SLE, 385, 392
  Sturge–Weber syndrome, 202
  Takayasu disease, 98, 99
  treatment-related
    antiplatelet agents, 330–331
    heparin, 330
    oral anticoagulants, 328–330, 541
    reversal, 329–330, 331, 332
    thrombolytic agents/t-PA, 331–332
  VHL disease, 211–212
intracranial pressure, raised, 104–106, 591, 592, 593, 594
intrathecally produced antibodies (ITAbs), 29, 72
intravenous gamma globulin (IVIg), 41, 296
  risk factor for strokes, 410
iron overload, 150, 153–154
ISTH scoring system for DIC, 318

JAAM scoring system for DIC, 318
jaw claudication, 88

Kawasaki disease (KD), 38–41, 403
  clinical features, 39
  CNS disease, 39–41
  diagnosis, 39, 403
  epidemiology, 38
  pathology, 38–39
  treatment, 41
keratitis, 302
kidney, *see* renal disease
Klippel–Trenaunay–Weber syndrome, 201, 270

lactate dehydrogenase, 350
left atrial appendage occlusion, 542
leukemia
  and DIC, 322
  hyperviscosity syndrome, 411, 413
  and ICHs, 332–333, 440–441
leukoaraiosis (white matter hyperintensities)
  amyloid angiopathy, 538–539
  antiphospholipid syndrome, 310
  dilatative arteriopathy, 563
  Fabry disease, 161–162
  migraines, 626–627
  pre-eclampsia, 617
leukoencephalopathy
  CADASIL, 134–138
  CARASIL, 140–144
  HERNS, 299
  PRES, 612–613
  radiation damage, 503
leukotriene antagonists, 378
Libman–Sacks endocarditis
  antiphospholipid syndrome, 312, 390
  cancer, 439
  DIC, 322
  SLE, 383, 386
lipoproteins
  and blood viscosity, 412
  LDL apheresis, 166, 475
  lipoprotein (a), 339, 472
  nephrotic syndrome, 472, 475
livedo racemosa/livedo reticularis, 259–260, 482, 484, 486
LMNA/lamin A, 189–190
locked-in syndrome, 519
Loeys–Dietz syndrome, 217
lonafarnib, 191
LSD (D-lysergic acid diethylamide), 431
lung
  lymphomatoid granulomatosis, 506
  microscopic polyangiitis, 369
  pulmonary embolisms, 449, 569
  see also pulmonary arteriovenous malformations
lupus anticoagulant, 305, 306, 307–308
Lyme disease, 27–28
  diagnosis, 28–29
  stroke, 27, 29–36
lymphoma
  and angiitis, 78

lymphoma (cont.)
intravascular, 630–632
lymphomatoid granulomatosis, 506–507

Maffucci syndrome, 270
magnesium, 418, 424–426
magnesium sulfate, 425–426, 609, 613
magnetic resonance imaging, *see* MRI scans
malignant atrophic papulosis (Degos disease), 445–447
mandibuloacral dysplasia, 190
Marfan syndrome, 170–173
aneurysms, 172, 217
aortic dilatations, 171, 172–173
clinical features, 170–172, 582
CNS disease, 171–172, 173
diagnosis, 171, 172
genetics, 170
treatment, 172–173
marijuana, 431–432
matrix metalloproteinases, 218, 563
maxillofacial AVMs, 255, 256–257
MDMA (ecstasy), 430
MELAS (mitochondrial encephalomyopathy, lactic acidosis and stroke-like episodes), 194–198
clinical features, 195, 626
diagnosis, 195
genetics, 194, 195
pathology, 194–195, 196
treatment, 197–198
membranous glomerulonephropathy, 470, 474
meningitis, 26–27
APMPPE, 283, 285
aspergillosis, 14–15
coccidioidal, 17
cryptococcosis, 13, 16–17
cysticercosis, 21
Kawasaki disease, 39
Lyme disease, 27–36
pneumococcal, 26, 27
vaccination, 26, 282
syphilitic, 1–2
see also meningovascular syphilis
tuberculous (TBM), 7–11, 17
viral, 26–27
meningoencephalitis
APMPPE, 283
Behçet's disease, 103–104
meningovascular syphilis, 2–3, 27
Menkes disease, 250–253
methamphetamine, 429–430
methotrexate
eosinophilic granulomatosis with polyangiitis, 379
giant cell arteritis, 93, 396
microscopic polyangiitis, 373
microalbuminuria, 472
microangiopathic hemolytic anemia, 437
microangiopathy of the retina, inner ear, and brain, *see* Susac syndrome
microbleeds
amyloid angiopathy, 538, 540–541, 542
CADASIL, 135
differential diagnosis, 230
moyamoya disease, 550–551
microembolic signals
Behçet disease, 106
SLE, 383
Sneddon syndrome, 486
microinfarcts, *see* silent cerebral infarctions
microscopic polyangiitis (MPA), 369–374
clinical features, 361, 369
neurological, 367, 370
diagnosis, 370–373
pathology, 373
treatment, 373–374
migraines, 621–627
arterial dissections, 516, 623
CADASIL, 134, 138, 625–626
and calcium, 423
diagnosis of migrainous strokes, 621–622
genetic associations, 625–626
HaNDL(Bartleson syndrome), 625
pathology, 622–623
PAVM, 122, 124
PFO, 623
prevention of strokes, 627
after radiotherapy (SMART syndrome), 503, 624–625
RCVS, 625
risk factor for strokes, 621
anti-migraine medications, 626
thrombophilia, 623–624
white matter hyperintensities, 626–627
migratory thrombophlebitis, 439
mitochondrial disorders, 194–198
clinical features, 195, 626
diagnosis, 195
genetics, 194, 195
overlap syndromes, 197
pathology, 194–195, 196
treatment, 197–198
mitral valve
Fabry disease, 163
Marfan syndrome, 171, 173
PXE, 177
Sneddon syndrome, 483, 484–485, 486
see also infective endocarditis
mononeuritis multiplex, 362
mood disorders, 135
moyamoya disease, 545–555
clinical features, 550–551
diagnosis, 551–553
epidemiology, 545–548, 551
moyamoya syndrome in other conditions, 549
NF1, 246
radiotherapy, 503
sickle cell disease, 148, 153
SLE, 392
pathology, 548–550
prognosis, 553–554
treatment, 153, 554–555
MPA, *see* microscopic polyangiitis
MPO-ANCA antibodies, 370, 373
MRI scans (magnetic resonance imaging)
amyloid angiopathy, 535–539
angiokeratoma, 506–507
antiphospholipid syndrome, 310
arterial dissections, 514, 520, 522–524
carotid artery redundancy, 513
Behçet disease, 103–104, 105
CADASIL, 135–136
cancer patients, 441, 442
CARASIL, 141
cavernous malformations, 228–230, 239
cerebral venous anatomy, 589
cerebral venous thrombosis, 590, 592, 614
Chagas disease (heart), 47
Cogan syndrome, 303
cysticercosis, 23
dilatative arteriopathy, 561, 562
DVAs, 237, 239
Ehlers–Danlos syndrome, 182, 183
Fabry disease, 160–162
Henoch–Schönlein purpura, 401
HERNS, 299
HHT, 130
hypereosinophilic syndromes, 499
infective endocarditis, 66, 67–68
lymphoma, intravascular, 631
MELAS, 197
microscopic polyangiitis, 367
migraines, 623, 626–627
moyamoya disease, 550, 552–553
PACNS, 79–81, 600
PAVM, 123
polyarteritis nodosa, 366, 368
PRES, 612–613
progeria, 189
RCVS, 597, 599–600, 611
sarcoidosis, 114
sickle cell disease, 150–151
SLE, 382–383
Sneddon syndrome, 485
Sturge–Weber syndrome, 203
Susac's syndrome, 293, 295
Sweet syndrome, 465
syphilis, 2–3
thrombolysis-related ICHs, 331
TTP, 350–351
tuberculous meningitis, 9
VHL disease, 208
VZV vasculopathy, 72
MTHFR (methylenetetrahydrofolate reductase), 456, 626
mTOR inhibitors, 205
mucormycoses, 15
multiple sclerosis, 295, 340
mycophenolate mofetil, 84
mycotic aneurysms, 14, 24, 66, 68
myelitis, syphilitic, 3
myelomas, 410
myeloproliferative diseases, 341–342, 411

NAA (*N*-acetylaspartate), 161–162
nails, 39
neonatal antiphospholipid syndrome, 391
neonatal progeria, 190
nephrotic syndrome, 469–475
CNS disease, 472, 473–474
glomerulonephritis, 470, 474
pathology, 469–472
treatment, 474–475
neuroborreliosis, *see* Lyme disease
neurocysticercosis (NCC), 20–24
clinical features, 21–22
diagnosis, 22–24
pathology, 20–21
treatment, 24
neurofibromatosis type 1 (NF1), 218, 245–248
neurofibromatosis type 2 (NF2), 245
neurosarcoidosis, 111–117
clinical features, 111, 112–114
diagnosis, 111
epidemiology, 111
pathology
case reports, 112–114
immunology, 111–112
prognosis, 117
treatment, 117
neurosyphilis, 1–5
diagnosis, 2–4
epidemiology, 1, 2, 27
and HIV, 3
pathology, 1–2
spinal involvement, 3
strokes, 2
treatment, 4–5
Treponema pallidum, 1
neutrophils
microscopic polyangiitis, 373

neutrophil extracellular traps in DIC, 320
Sweet syndrome, 462–463
VZV vasculopathy, 75
nevus sebaceous, 479, 480
nicardipine, 418, 422
nifurtimox, 47–48
nimodipine, 296, 418–419, 422, 423
nitric oxide, 196
nitric oxide synthase, 218, 421
NMDA antagonists, 421
NOACs (new oral anticoagulants), 329, 442, 542, 594
antiphospholipid syndrome, 313
reversal of effect, 330
non-bacterial thrombotic endocarditis, *see* Libman–Sacks endocarditis
nose bleeds (epistaxis), 121, 129
NOTCH3, 134–135, 137–138

occipital horn syndrome, 251
ophthalmology, *see* eye/loss of vision
opiates, 429
optic neuropathy
carotid artery dissections, 518
giant cell arteritis, 89, 92, 396
orbital AVMs, 256
oral anticoagulants, *see* NOACs; warfarin
oral contraceptives, 590, 594
oral maxillofacial AVMs, 255, 256–257
orthostatic hypotension, 163
Osler, William, 63, 356, 381
Osler–Weber–Rendu disease, *see* hereditary hemorrhagic telangiectasia
osteochondroma, 492
osteopetrosis, 490–491
osteoporosis, 421
osteoprotegerin, 490
overgrowth syndromes, 480
oxidative phosphorylation, 194

PACNS, *see* primary angiitis of the central nervous system
Paget disease of bone, 491
pain
arterial dissection, 516–517, 518, 582
Fabry syndrome, 159
see also headache
PAN, *see* polyarteritis nodosa
Paracoccidioides brasiliensis, 17
paradoxical embolisms, 565–571
cryptogenic strokes, 566
diagnosis, 565
and DVTs, 439, 568–569
and PAVMs, 120, 129–130
and PFOs, 439, 565, 566–568, 569
and pulmonary embolisms, 569
RoPE score, 567–568
SLE, 384
treatment, 569–570
paraneoplastic strokes, 437–442
autoimmune causes, 437–439
classification, 437
clinical features, 440–441
hypercoagulability, 439
investigations, 441–442
treatment, 442
paroxysmal nocturnal hemoglobinuria, 340–341
Parry–Romberg syndrome, 495
partial thromboplastin time (PTT), 318
patent foramen ovales (PFOs)
closure, 570, 623
and migraines, 623
paradoxical embolisms, 439, 565, 566–568, 569
PAVM, *see* pulmonary arteriovenous malformations
pediatrics, *see* children
pelvic vein DVT, 568–569
penicillin, 4–5
pentoxifylline, 117
periodontal disease, 492–493
peripartum cardiomyopathy, 614–615
peripheral neuropathies
aortic dissections, 584
ATP7A-related distal motor neuropathy, 251
eosinophilic granulomatosis with polyangiitis, 379
polyarteritis nodosa, 362
perivascular spaces, enlarged, 539
PET (positron emission tomography)
amyloid angiopathy, 538
Fabry disease, 162
giant cell arteritis, 91
Sturge–Weber syndrome, 203
PFOs, *see* patent foramen ovales
PHACTR1/PHACTR1, 512–513
phencyclidine, 431
phenylpropanolamine, 430
pheochromocytomas, 211
phlebotomy, 413
PiB imaging of amyloid, 538
plasma viscosity, 409–410
hyperviscosity syndromes, 410, 412, 413
plasmapheresis
Fabry disease, 166
hyperviscosity syndromes, 413
microscopic polyangiitis, 374
nephrotic syndrome, 475
TTP, 351, 352
plasmin, 337
plasminogen, 337
plasminogen activator inhibitor-1 (PAI-1), 337
platelets
in DIC, 318, 320, 323–324
disorders associated with ICHs, 332, 333–334
HIT, 323, 342–343
paraneoplastic thrombocytopenia, 437
physiological role, 336–337
thrombocythemia
essential, 341–342, 411, 413
in IBD, 450
transfusion, 331, 332
pneumococcal meningitis, 26, 27
vaccination, 26, 282
polyarteritis nodosa (PAN), 359–369, 397
causes, 359
in children, 397
clinical features, 38, 359–361
neurological, 362–363
diagnosis, 359, 363–367, 397
epidemiology, 359
pathology, 366–367, 368, 397
treatment, 369, 397
polyclonal gammopathies, 410
polycythemia
primary (polycythemia vera), 341–342, 411, 413
secondary, 210, 411
polymyalgia rheumatica, 88–89
port-wine nevus, 201, 205
positron emission tomography, *see* PET
posterior reversible (leuko) encephalopathy syndrome (PRES), 612–613
postpartum period
pre-eclampsia, 608
RCVS, 603, 610–612
thrombosis risk, 606
see also pregnancy
praziquantel, 24
pre-eclampsia
CNS disease, 609, 610–615
diagnosis, 607–608
epidemiology, 608
HELLP syndrome, 323, 607–608
pathology, 609–610
risk of future vascular disease, 617
treatment, 609, 616
pregnancy, 606–617
antiphospholipid syndrome, 305, 389
cardiomyopathy, 614–615
causes of stroke, 607
cavernous malformations, 235–236
CVTs, 594, 607, 613–614
eclampsia, 607, 609, 610, 613
epidemiology of strokes, 606–607, 609, 614
HHT and PAVMs, 122, 132
investigations into strokes, 615
outcomes of strokes, 607
pre-eclampsia, *see* pre-eclampsia
PRES, 612–613
RCVS, 603, 610–612
Sweet syndrome, 461
thrombophilia, 343
treatment of strokes, 615–616
TTP, 347, 349
PRES (posterior reversible (leuko)encephalopathy syndrome), 612–613
primary angiitis of the central nervous system (PACNS), 77–84, 397–399
in children, 398–399
clinical features, 78, 283, 398–399
diagnosis, 78–83, 398
compared with RCVS, 78, 598, 600, 601, 602
pathology, 77–78, 398, 399
treatment, 83–84, 398, 399
progeroid syndromes, 187–191
CNS disease, 188–189
genetics
LMNA, 189–190
WRN, 190
mandibuloacral dysplasia, 190
neonatal progeria (WRS), 190
progeria (HGPS), 187–191, 495
treatment, 190–191
Werner syndrome, 190
atypical, 190
progressive facial hemiatrophy, 495
protamine sulfate, 330
protein C
deficiency, 338–339
in DIC/sepsis, 324
factor V resistance to, 339, 450–455
protein S deficiency, 54–55, 312, 338–339, 456
Proteus syndrome, 480
prothrombin complex concentrate (PCC), 324, 329–330
prothrombin G20210A mutation, 339, 456
prothrombin time (PT), 318, 329
prothrombotic state, *see* thrombophilia
pseudoephedrine, 430
pseudoxanthoma elasticum (PXE), 175–178
clinical features, 175–177
CNS disease, 177–178
genetics, 175
treatment, 178

pulmonary arteriovenous malformations (PAVMs), 119–125
  clinical features, 120–122, 129–130
  diagnosis, 122–124, 129
  epidemiology, 120
  pathology, 119–120
  pediatric screening, 125
  treatment, 119, 124–125, 132
pulmonary embolisms, 449, 569
pulvinar sign, 162
purpura fulminans, 321
PXE, *see* pseudoxanthoma elasticum

radiography
  aortic dissections, 584
  PAVMs, 123
radionuclide perfusion lung scanning, 124
radiosurgery, 236
radiotherapy
  for hemangioblastomas, 212
  for neurosarcoidosis, 117
  vascular injury caused by, 440, 502–503
    necrosis, 301
    SMART, 503, 624–625
RCVS, *see* reversible cerebral vasoconstriction syndrome
red blood cells
  polycythemia vera, 341–342, 411, 413
  secondary polycythemia, 210, 411
  in TTP, 350
  and viscosity of blood, 408, 409, 411–412, 413
  see also sickle cell disease
renal artery, 575, 577
renal disease
  acute renal failure, 469
  ADPKD, 215
  calciphylaxis, 423–424
  chronic kidney disease, 469
  Fabry disease, 165
  glomerulonephritis, 470, 474
  hemolytic uremic syndrome, 350
  Henoch–Schönlein purpura, 356
  HERNS, 298
  microscopic polyangiitis, 369
  nephrotic syndrome, 469–475
renal transplantation, 165
retina
  angioid streaks in PXE, 176
  APMPPE, 275, 283–284
  diabetic retinopathy, 472
  Eales disease, 273–274, 283
  HERNS, 298
  PACNS, 79
  retinal vasculopathy with cerebral leukodystrophy, 298
  Susac syndrome, 292, 293–294
  Takayasu retinopathy, 96
  Wyburn–Mason syndrome, 255–256, 258
reversible cerebral vasoconstriction syndrome (RCVS), 597–603, 625
  clinical features, 599
  diagnosis, 599–602, 611–612
    compared with PACNS, 78, 598, 600, 601, 602
  postpartum, 603, 610–612
  risk factors and triggers, 431, 598–599, 612
  treatment, 602–603, 612
  and TTP, 351
rheumatoid arthritis, 399–400
rhinocerebral mucormycosis, 15
Rho kinase, 228, 236–237
Rhupus syndrome, 403
Rich focus, 7
rifampicin, 11
rituximab
  microscopic polyangiitis, 374
  sarcoidosis, 117
  TTP, 351
rivaroxaban, 313, 329, 442
  reversal of effect, 330
RNF213 (ring finger protein 213), 549
RPR test (for syphilis), 3–4

SAHs, *see* subarachnoid hemorrhages
sarcoidosis, *see* neurosarcoidosis
scalp
  alopecia, 140, 141, 187
  ischemia after arterial harvest, 555
  necrosis, 90
  tenderness, 88
SCD, *see* sickle cell disease
schistocytes, 350
Schulman–Upshaw syndrome, 347
schwannomatosis, 245
scleroderma, 401–402, 495–496
screening
  for familial aneurysms, 215, 217, 221–222, 223
  for Lyme disease, 28, 36
  for PAVM, 122–123, 125, 129, 132
  for stroke risk in sickle cell disease, 152
  for thrombophilia, 344
sedatives, 432
seizures
  cavernous malformations, 231, 236
  CVTs, 591, 592, 593, 594
  DVAs, 239, 240
  eclampsia, 607, 609, 610, 613
  epidermal nevus syndrome, 479
  Sneddon syndrome, 483
  Sturge–Weber syndrome, 201–202, 205
  tuberculous meningitis, 9
  and warfarin, 475
selective serotonin reuptake inhibitors (SSRIs), 542, 603
serology
  Chagas disease, 46
  Lyme disease, 28–29
  syphilis, 3–4
severe pre-eclampsia–eclampsia, *see* pre-eclampsia
sickle cell disease (SCD), 147–155
  clinical features, 149
  diagnosis, 149–151
  epidemiology, 147–148
  pathology, 147
    reduced red cell deformability, 408, 411–412
  risk factors for strokes, 148
  treatment, 151–155, 413
siderosis, cerebral, 538, 540
silent cerebral infarctions (SCIs) (microinfarcts)
  amyloid angiopathy, 539
  progeria, 188
  sickle cell disease, 148, 149, 150–151
sirolimus, 205
Sjögren syndrome, 392–393
skeletal disorders, *see* bone disorders
skin lesions
  angiokeratoma, 159
  BRBNS, 268
  CADASIL, 137
  Degos disease, 445, 446
  Divry–van Bogaert syndrome, 259–260, 264
  Ehlers–Danlos syndrome, 181, 217
  epidermal nevi, 478, 479
  Henoch–Schönlein purpura, 356
  lymphomatoid granulomatosis, 506
  microscopic polyangiitis, 369
  purpura fulminans in DIC, 321
  PXE, 175–176
  Sneddon syndrome, 482, 484, 486
  Sturge–Weber syndrome (port-wine nevus), 201, 205
  Sweet syndrome, 462
  telangiectasia, 121, 122, 128, 129
skull disorders
  Camurati–Engelmann disease, 491–492
  craniosynostosis, 491
  osteopetrosis, 490–491
  Paget disease, 491
SLE, *see* systemic lupus erythematosus
SMAD4, 120
Small-vessel disease
  CADASIL, 134–138
  CARASIL, 140–144
  eosinophilic granulomatosis with polyangiitis, 378–380, 403, 499
  Henoch–Schönlein purpura, 356–357, 400–401
  HIV, 56
  hyperviscosity syndromes, 412
  sarcoidosis, 112–113
  SLE, 382–383
  Susac syndrome, 283, 284, 290–296
  thrombotic microangiopathies, 323, 340
    TTP, 347–352
  see also cerebral amyloid angiopathy
SMART (stroke-like migraine attacks after radiation therapy), 503, 624–625
smoking, 219, 222–223, 412, 433
smooth muscle contraction, 418–419
smooth muscle progenitor cells, 549–550
Sneddon syndrome, 402, 482–486
  antiphospholipid antibodies, 309–310, 402, 483, 484
  causes, 484–485
  clinical features, 482–483
  CNS disease, 402, 482–483, 484, 486
  diagnosis, 260–267, 485–486
  epidemiology, 482
  pathology, 484
  skin lesions, 482, 484, 486
  treatment, 486
SOLAMEN syndrome, 480
soluble fibrin, 318
spice (synthetic cannabinoid), 431
spine/spinal cord
  ankylosing spondylitis, 402
  aortic dissections, 584
  arteriovenous malformations, 130, 131, 479–480
  CARASIL, 141, 144
  fibrocartilaginous embolisms, 490
  Marfan syndrome, 171, 172
  PACNS, 78
  subarachnoid hemorrhages, 107
  syphilis, 3
  vertebral artery dissections, 519
spondyloepiphyseal dysplasia, 491
spondylosis deformans, 141, 144

Staphylococcus aureus, 64
statins, 386, 456, 542
stents, 527–528, 530
steroids, *see* corticosteroids
storage pool disorders, 333
Streptococcus pneumonia
  meningitis, 26, 27
  vaccination, 26, 282
Sturge–Weber syndrome, 200–205
  clinical features, 201–202
  diagnosis, 203, 257
  GNAQ mutation, 200–201
  pathology, 203–205
    deterioration in function, 202–203
  treatment, 205
subarachnoid hemorrhage (SAH)
  aspergillosis, 14
  Behçet disease, 107, 394
  Ehlers–Danlos syndrome, 183
  familial aneurysms
    epidemiology, 219–221
    non-genetic risk factors, 222–223
    screening for those at risk, 215, 217, 221–222, 223
  imaging, 220
  and magnesium, 425, 426
  in pregnancy, 607
  in RCVS, 599
  Sjögren syndrome, 393
  SLE, 385, 392
  treatment with calcium-channel antagonists, 422–423
  vertebral artery dissections, 107, 518
subclavian steal syndrome, 98, 100
substance use/abuse
  alcohol, 431, 432–433
  amphetamines, 429–430
  cocaine, 398, 430–431
  and HIV, 55
  inhalants, 432
  LSD, 431
  marijuana, 431–432
  opiates, 429
  phencyclidine, 431
  sedatives, 432
  tobacco, 219, 222–223, 412, 433
surgery
  aortic dissections, 585
  cavernous malformations, 236
  hemangioblastomas (VHL disease), 211–212
  in infective endocarditis
    cardiac, 65–66
    mycotic aneurysms, 66
  intracranial hypertension, 593
  Marfan syndrome, 172–173
  moyamoya disease, 153, 554–555
  neurofibromatosis type 1, 247–248
  PAVM, 124
  sickle cell disease, 153
  Sturge–Weber syndrome, 205
  Takayasu disease, 99–100
Susac syndrome, 284, 290–296
  clinical features, 291–293
  diagnosis, 283, 293–295
  epidemiology, 290
  pathology, 290–291
  treatment, 295–296
Sweet syndrome, 461–467
  classification, 461–462
  clinical features, 462, 465
  CNS disease, 464–466
  diagnosis, 463–464, 465
  epidemiology, 461
  medication-associated, 462, 464, 466
  pathology, 462–463, 464, 465
  treatment, 462, 464–465, 466–467
syphilis, *see* neurosyphilis
systemic lupus erythematosus (SLE), 381–386, 391–392
  and antiphospholipid syndrome, 305–306, 309, 384
  in children, 392
  diagnosis, 381, 391
  and DIC, 323
  pathology of CNS disease, 381–385, 392, 474
  stroke risk, 381, 391
  treatment, 385–386, 392
systemic sclerosis (scleroderma), 401–402, 495–496

T cells
  Behçet disease, 102
  giant cell arteritis, 88
  IBD, 448
  sarcoidosis, 112
Taenia solium infestation, *see* cysticercosis
Takayasu disease, 96–100, 402–403
  classification, 97
  clinical features, 98, 402
  diagnosis, 98–99, 403
  epidemiology, 96
  pathology, 96–97
  strokes, 98, 99
  after Sweet syndrome, 466
  treatment, 99–100, 403
tapeworms, *see* cysticercosis
tau peptide, 540
TEK, 270
telangiectasia, *see* hereditary hemorrhagic telangiectasia
temporal arteritis (giant cell arteritis), 87–93, 396
  and cancer, 438–439
  clinical features, 88–90, 396
  diagnosis, 90–92, 396
  epidemiology, 87
  large-vessel disease, 90, 91
  pathology, 87–88, 90
    VZV, 73–74
  prognosis, 93
  treatment, 89, 92–93, 396
TGF-β, *see* transforming growth factor beta
thalassemias, 147
thrombectomy, 593
thrombin, 320, 328, 336
thrombocythemia
  essential, 341–342, 411, 413
  in IBD, 450
thrombocytopenia
  heparin-induced, 323, 342–343
  and ICHs, 332, 334
  paraneoplastic, 437
thrombolysis
  aortic dissections, 585
  arterial dissections, 526–527
  in CVTs, 593
  in IBD, 456
  ICHs related to use, 331–332
  in infective endocarditis, 64–65
  in pregnancy, 616
  reversal, 332
  in SLE, 151, 385
thrombomodulin, 324, 337
thrombophilia, 336
  antithrombin deficiency, 338, 456
  in cancer patients, 439
    anticoagulation therapy, 442
  in children, 343
  cryptogenic stroke, 568
  Fabry disease, 163
  factor V Leiden/APC resistance, 163, 339, 450–455
  fibrinolytic abnormalities, 339
  HIT, 323, 342–343
  HIV patients, 54–55
  homocysteinemia, 339–340, 456
  hypereosinophilia, 499
  IBD, 449–456
  mechanisms, 337–338
  migraines, 623–624
  myeloproliferative diseases, 341–342
  nephrotic syndrome, 470–471
  paroxysmal nocturnal hemoglobinuria, 340–341
  in pregnancy/postpartum, 343
    TTP, 347, 349
  protein C/protein S deficiencies, 54–55, 312, 338–339, 456
  prothrombin G20210A mutation, 339, 456
  scleroderma, 495
  Sneddon syndrome, 484–485
  testing for, 344
  thrombotic microangiopathies, 323, 340, 347–352
thrombotic microangiopathies (TMAs), 323, 340
  see also thrombotic thrombocytopenic purpura
thrombotic thrombocytopenic purpura (TTP), 347–352
  clinical features, 348–350
  diagnosis, 350–351
  epidemiology, 347
  pathology, 347–348
  prognosis, 350, 352
  treatment, 350, 351–352
thunderclap headache
  CAD, 517
  RCVS, 599, 601–602, 625
TIAs, *see* transient ischemic attacks
tinnitus, pulsatile, 517
tissue factor (TF), 312, 320
tissue factor pathway inhibitor (TFPI), 320
tissue plasminogen activator (t-PA)
  in IBD, 456
  ICHs related to use, 331–332
  in infective endocarditis, 64–65
  in pregnancy, 616
  reversal, 332
  sickle cell disease, 151, 385
  see also thrombolysis
TNF-α, *see* tumor necrosis factor alpha
tobacco use, 219, 222–223, 412, 433
tocilizumab, 99, 108, 396
tranexamic acid, 324, 332
transcranial Doppler, *see* ultrasonography
transforming growth factor beta (TGF-β)
  aneurysm development, 218
  Camurati–Engelmann disease, 491, 492
  CARASIL, 144
  HHT, 120, 128
transient global amnesia, 584
transient ischemic attacks (TIAs)
  amyloid angiopathy, 541
  cysticercosis, 21
  moyamoya disease, 550
transient visual loss (amaurosis fugax), 89, 518
traumatic arterial dissections, 509–511, 529
Treponema pallidum, 1
  see also neurosyphilis
treprostinil, 446–447
TREX1 (3′–5′ exonuclease), 298, 299
triatomine bugs, 45

trichopoliodystrophy (Menkes disease), 250–253
triptan, 626
Trousseau syndrome, 439
trypanosomiasis, American, *see* Chagas disease
TTP, *see* thrombotic thrombocytopenic purpura
tuberculosis (TB), 7–11, 17
  clinical features, 8–9
  diagnosis, 9
  imaging, 9–11
  pathology, 7–8
  treatment, 11
tumor necrosis factor alpha (TNF-α)
  aneurysm development, 218
  Behçet disease, 102–103, 108
  sarcoidosis, 112, 117
  tuberculosis, 8

ulcerative colitis, *see* inflammatory bowel disease
ultrasonography
  arterial dissections, 525
  Behçet disease, 104
  Chagas disease, 47
  cysticercosis, 24
  giant cell arteritis, 91
  PAVM, 124
  in pregnancy, 615
  RCVS, 601
  sickle cell disease, 147, 148, 151–152
uveo-cerebral vasculitic syndromes
  APMPPE, 275–286
  Eales disease, 273–274, 283
  Susac syndrome, 283, 284, 290–296

VADs, *see* vertebral artery dissections
Valsalva maneuver, 565
varicella zoster virus (VZV) vasculopathy, 71–75
  epidemiology, 71
  giant cell arteritis, 73–74
  in HIV patients, 57–59
  intracerebral, 71–73, 74
  and PACNS, 78, 399
  pathogenesis, 74–75
vasculitis
  APMPPE, 282–283
  bacterial meningitis, 2, 26, 29
  Behçet disease, 103, 106
  causes, 79
  classification, 359
  cryoglobulinemia, 394–395
  cysticercosis, 20, 21, 22, 24
  drug abuse, 429–430
  eosinophilic granulomatosis with polyangiitis, 378–380, 403, 499
  giant cell (temporal) arteritis, 87–93, 396, 438–439
  granulomatosis with polyangiitis, 395–396
  Henoch–Schönlein purpura, 356–357, 400–401
  HIV-associated, 55
    aneurysmal arteriopathy in children, 56
    large- and medium-vessel vasculopathy, 56–59
    small-vessel vasculopathy, 56
  IBD, 450
  Kawasaki disease, 38–41, 403
  microscopic polyangiitis, 361, 369–374
  PACNS, 77–84, 397–399
  paraneoplastic, 437–439
  polyarteritis nodosa, 38, 359–369, 397
  retinal (Eales disease), 273–274, 283
  Sjögren syndrome, 393
  SLE, 384–385, 386
  Takayasu disease, 97
  tuberculosis, 7, 8
vasoconstriction
  and calcium, 418–419, 422–423
  drug abuse, 430–431
  and magnesium, 424
  migraines, 622, 623
  in pre-eclampsia, 609–610
  see also reversible cerebral vasoconstriction syndrome
vasogenic edema
  PRES, 612–613
  TTP, 351
VDRL test (for syphilis), 4
venous thromboembolism, *see* cerebral venous (sinus) thrombosis; deep venous thromboses
verapamil, 419
vertebral artery dissections (VADs)
  clinical features, 518–519
  imaging, 521
  intracranial, 519, 528
  prognosis, 529
  traumatic, 511
  and vertebral artery hypoplasia, 514
vertebrobasilar system
  dilatative arteriopathy, 560
  Fabry disease, 160–161, 162
verteporfin, 205
vestibular dysfunction
  Cogan syndrome, 302–303, 304
  Susac syndrome, 292
VHL disease, *see* von Hippel–Lindau disease
VHL/pVHL, 208, 210
viscoelastic hemostatic assays, 318
viscosity of blood, 408–409
vision, loss of, *see* eye/loss of vision
vitamin D, 236
vitamin K, 329
von Hippel–Lindau disease (VHL), 208–212
  clinical features, 210–211
  diagnosis, 208–210, 257
  genetics, 208, 210
  treatment, 211–212
von Recklinghausen disease (neurofibromatosis type 1), 218, 245–248
von Willebrand disease (vWD), 333
von Willebrand factor (vWF), 55, 320, 336, 347–348
VZV, *see* varicella zoster virus vasculopathy

Waldenström macroglobulinemia, 410
warfarin
  antiphospholipid syndrome, 312–313, 391, 486
  Chagas disease, 49
  ICHs related to use, 328–329, 541
    reversal of effect, 329–330
  infective endocarditis, 65
  Sneddon syndrome, 486
  Takayasu disease, 99
WATCHMAN device, 542
Werner syndrome, 190
  atypical, 190
white matter pathology
  amyloid angiopathy, 538–539
  antiphospholipid syndrome, 310
  CADASIL, 134–138
  CARASIL, 140–144
  dilatative arteriopathy, 563
  Fabry disease, 161–162
  HERNS, 299
  migraines, 626–627
  pre-eclampsia, 617
  PRES, 612–613
  radiation damage, 503
  Susac syndrome, 293
  TTP, 351
Wiedemann–Rautenstrauch syndrome (neonatal progeria), 190
WRN/DNA helicase, 190
Wyburn–Mason syndrome, 255–258

X-rays
  aortic dissections, 584
  PAVM, 123

Ziehl–Neelsen stain, 9
zoster, *see* varicella zoster virus vasculopathy (VZV)