

Index

A

- ACA. *See* Anticentromere antibodies
- Acetylcholine (ACh)
increased degradation, cholinesterase, 316
neuronal release, 316
parasympathetic nervous system, 316
signal, secretion, 322
- ACh. *See* Acetylcholine
- ACTH. *See* Adrenocorticotrophin hormone
- AD. *See* Autonomic dysfunction
- Adie's syndrome, 315
- Adrenocorticotrophin hormone (ACTH)
CRH, 332–333
HPA axis, 333
IL-6 stimulation, 333
- AECG. *See* American-European consensus group
- AIHA. *See* Autoimmune hemolytic anemia
- Alefacept
biological therapy, 619
treatment, chronic plaque psoriasis, 618
- American-European consensus criteria (AECC), 4, 6
- American-European consensus group (AECG)
defined, 432
excessive stringency, 439–440
scientific community, 440
tool, patient classification, 439
- ANA. *See* Antinuclear antibodies
- ANCA. *See* Anti-neutrophil cytoplasmic antibodies
- Angular stomatitis, 97
- ANS. *See* Autonomic nervous system
- Anticentromere antibodies (ACA), 419
- Anti-neutrophil cytoplasmic antibodies (ANCA), 177, 420
- Antinuclear antibodies (ANA)
diagnosis, 414–415
and RF, 334
SjS, 179
- Antiphospholipid antibodies (aPL)
coexistence, 418–419
description, 418
detection, classification, 418
- Antiphospholipid syndrome (APS), 296, 489
- APS. *See* Antiphospholipid syndrome
- ARTs. *See* Autonomic reflex tests
- ASP. *See* Autonomic symptom profile
- Atrioventricular (AV) block
CHB, 346
first-degree, 348–349, 351
second-degree, 351
- Autoimmune diseases, pSjS
APS, 489
cryoglobulinemia, systemic vasculitis, 489
MCTD, 488
organ-specific
celiac disease, 492
liver disease, 491
thyroiditis, 490–491
sarcoidosis, 489–490
- Autoimmune hemolytic anemia (AIHA)
causes, primary SjS, 364
description, 364
etiopathogenesis, 365
low prevalence, hemolytic assays, 363, 365
PRCA, 365
- Autoimmune hepatitis (AIH), 248
- Autoimmune pancreatitis (AIP)
radiologic abnormalities, 259
serum IgG4, 259
type 1 and 2, 260

- Autonomic dysfunction (AD)
 - description, 313
 - pathogenesis, pSS, 315–318
 - pSS patient diagnostic algorithm, 325–326
- Autonomic nervous system (ANS)
 - ARTs, activity, 321
 - cardiovascular tests, 321
- Autonomic neuropathy
 - AD pathogenesis, pSS
 - factors, M3R reduced stimulation, 316–318
 - immunological mechanisms, 315
 - secretory process and immune system interaction, 316, 317
 - serological factor, IgG fraction, 318
 - anti-M3R antibodies, 314–315
 - diagnostic tests
 - ANS, 318, 321
 - ASP, 321–322
 - finger skin blood flow, 320
 - HRV and ARTs, 321
 - orthostatic blood pressure, 318, 319
 - parasympathetic and sympathetic disorders
 - AD, 322
 - gastrointestinal, 324
 - orthostatic intolerance, 324–325
 - pupillomotor, 324
 - secretomotor, 322–323
 - urinary, 323
 - vasomotor, 325
 - patient autonomic nervous function, pSS, 313, 314
 - pSS patient diagnostic algorithm, AD, 325–326
 - treatment, 326–327
- Autonomic reflex tests (ARTs)
 - advantages, 321, 326
 - ANS activity measurement, 321
 - cardiovascular, 322
 - uses, 313, 318
- Autonomic symptom profile (ASP)
 - AD, 315
 - pSS, 321
- AV block. *See* Atrioventricular block

- B**
- B and T cell receptors
 - abatacept, 634
 - epratuzumab, 634
 - rituximab
 - proliferation, 633
 - retreatment, 634
- Baroreceptor sensitivity (BRS)
 - baroreflex sensitivity measurement, 321
 - and HRV, 321, 326
- B-cell hyperactivity, increase in BAFF
 - B-lymphocyte stimulator (BLyS), 604
 - Serum BAFF in SS, 604–605
- B cell lymphoma (Bcl)
 - chemotherapeutic agents, 585
 - DLBCL, 582–584
 - hematopathology, 572
 - marginal zone (MZ) lymphomas
 - B-cell maturation, peripheral lymph nodes., 572, 574
 - extranodal, MALT type, 573–578
 - histology, differential diagnosis, 581
 - NMZL management, 581–582
 - splenic lymphoid follicle, 572, 573
 - therapeutic approaches, 578–581
 - non-Hodgkin's, 168, 169
 - non-Hodgkin's lymphoma (NHL), 571
 - parotid, 169
 - types, SS patients, 169
- B cells, primary SjS
 - chemokines and antibody production
 - antimuscarinic receptor antibody, 75
 - organ-specific inflammation, 74
 - cytokines
 - BAFF and APRIL, 70
 - ELISA, 70–71
 - Th-17 lymphocyte subpopulation, 71
 - IgA autoantibodies and cytokines
 - BAFF, 75
 - sialic acid, 76
 - intrinsic abnormalities, 76
 - monoclonal expansion, 74
 - ontogeny, lymphocytes
 - activation-induced cytidine deaminase (AID), 72
 - BAFF, 72
 - bone marrow (BM), 71
 - subpopulations
 - Bm homeostasis, 72
 - relative expression, CD 38 and IgD, 73
- B-cell-targeted therapies in Sjögren's syndrome
 - BAFF-targeted therapy
 - A Proliferation-Inducing Ligand (APRIL), 608
 - tolerance, belimumab, 609
 - treatment strategies, 608, 609
 - epratuzumab, 608
 - hyperactivity
 - autoimmunity, 605

- efficacy lack, TNF inhibition, 605
 - evidence, 604
 - increase, BAFF, 604–605
- rituximab
 - open and controlled studies, 605–606
 - safety, 607
 - therapy, BAFF increase, 607–608
 - visual analogic scale (VAS), 607
- unequivocal evidence, 609
- Bcl. *See* B cell lymphoma
- Biological therapies, pSS
 - B cell hyperactivity, 620
 - cytokine targeted therapies
 - emerging anticytokine, 617
 - etanercept, 616–617
 - infliximab, 614–615
 - interferon alpha, 617
 - lymphocytic dysfunction, 619
 - systemic lupus erythematosus (SLE), 614
 - T cell targeted
 - abatacept, 619
 - alefacept, 618–619
 - efalizumab, 618
 - initiation, autoimmune process, 618
- Bronchial hyperresponsiveness (BHR), 202
- C**
- Candidiasis, 97–98
- Carbonic anhydrase (CA), 278
- Cardiovascular involvement
 - atrioventricular block, 189
 - autonomic cardiovascular dysfunction, 191
 - complications, pSS, 187–188
 - diastolic dysfunction, 189
 - myocarditis, 188
 - PAH, 190
 - pericarditis, 188
 - subclinical atherosclerosis, 189–190
 - symptomatic, 193
 - therapeutic management
 - cardiac complications, pSS, 192
 - pSS treatment algorithm, PAH, 192, 193
 - valvular abnormalities, 189
- Central nervous system (CNS)
 - cerebral lesions
 - focal neurological deficits, 294
 - MRI, 292–293
 - MS and SLE, 294
 - patient white matter damage, pSS, 293
 - cranial nerve involvement, 297–298
 - cutaneous involvement, 180
 - differential diagnosis
 - cognitive and affective disorders, 295
 - MS, 295–296
 - NMO, 296
 - PML, 297
 - “T1 black holes”, 296
 - and peripheral, 182–183
 - prevalence and classification
 - pSS, 291–292
 - theories, 292
 - SS patient diagnostic algorithm
 - aseptic meningitis/myelitis, 298
 - MRI, 298–299
 - viruses, 298
- Cerebrospinal fluid (CSF)
 - MS, 296
 - PCR, 299
- CHB. *See* Congenital heart block
- Chronicity and activity measurement,
 - primary SjS
 - British approach, 454
 - clinical and serological peculiarities
 - B cell hyperactivity and proliferation, 446
 - focal lymphocytic infiltration, 446
 - heterogeneity, 447
 - defined, activity, 446
 - development, disease status, 448
 - disease activity/damage assessment,
 - autoimmune disease, 447
 - disease severity, 446
 - EULAR initiative, 454–457
 - immunologic and inflammatory process, 445
 - Italian approach
 - ESSDAI, 450–453
 - multivariate models, 448
 - SSDDI and SSDAI, 448, 449
- Classification criteria, primary SjS
 - AECG, 432
 - American-European
 - analysis, ROC curve, 437
 - classification, 438, 439
 - defined, 439
 - drawbacks, 439
 - prevalence, 440
 - salivary biopsy, heavy reliance, 440
 - disease heterogeneity, 431–432
 - hallmark, xerostomia and KCS, 431
 - historical overview
 - awareness, 434
 - Copenhagen and San Francisco criteria, 432

- Classification criteria, primary SjS (*cont.*)
 diagnostic tests and ranges, 432–433
 importance, focal sialoadenitis, 434
 “probable” and “definite”, 432
 sets, 432
 similarities and dissimilarities, 433
 tests, diagnosis, 433–434
 preliminary European
 characteristics, 436–437
 classification, 436
 disorders and controls, 435
 EEC-COMAC, 434–435
 methodology and statistics, RA, 435
 monovariate, multivariate analyses and
 stepwise multiple regression, 435
- Congenital heart block (CHB)
 NLS epidemiology and clinical features,
 345–347
 pathogenesis, 350–351
- Conjunctival-associated lymphoid tissue
 (CALT), 110
- Connective tissue disorders (CTDs)
 ILD, 202
 UIP, 204
- Corticotropin releasing hormone (CRH)
 ACTH, 332–333
 cytokines, 332
 IL-6 stimulation, 333
- C-reactive protein (CRP)
 ESR, 358
 testing levels, 358
- CRH. *See* Corticotropin releasing hormone
- CRISP-3 gene. *See* Cysteine-rich secretory
 protein-3 gene
- CRP. *See* C-reactive protein
- Cryptogenic organizing pneumonia (COP),
 206
- Cysteine-rich secretory protein-3 (CRISP-3)
 gene
 activation, 334
 expression, 335
- Cytokines
 anti-cytokine therapies, 632–633
 IL-6
 acute phase reactions, 630
 chronic inflammation, 630–632
 systemic autoimmunity, 632
 IL-7, 632
 IL-17, 632
 IL-12, IL-18, and IL-23, 632
 TNF- α , 630
- D**
- Dehydroepiandrosterone sulphate (DHEAS)
 and DHEA, 335, 337
 serum, 338
- Dental caries, 94–95
- Denture-induced stomatitis, 96–97
- Diagnostic procedures II, primary SjS
 MRI, 401–404
 salivary gland biopsy, 404–409
 salivary gland involvement, 395
 salivary scintigraphy
 abnormalities, 398
 description, 396, 398
 percentage, sensitivity and specificity,
 396
 qualitative and non-invasive measures,
 396
 system, Shall’s grading, 397
 sialography, 398, 399
 tomography, CT scans, 401
 ultrasound
 diagnosis and longitudinal follow-up,
 398
 Frankfurt horizontal plane, 399
 parenchymal inhomogeneity, 400
 sensitivity, hypoechoic zones and echo-
 genic lines, 399–400
 substantial drawbacks, 398–399
 ultrasonographic findings, 399, 400
 ultrasonographic grading, 400
 ultrasonographic images, 401
- Diagnostic procedures I, primary SjS
 definitions, 381
 eye and mouth dryness, conditions, 380
 ocular (*see* Ocular tests, primary SjS)
 oral (*see* Oral tests, primary SjS)
 prevalence, xerostomia, 380
 questionnaires, 381
 symptoms evaluation, 381
- Diffuse large B-cell lymphomas (DLBCL)
 histology
 lymph node biopsy, 583, 584
 nodal and extranodal dissemination,
 583
 MALT, 503
 MZBCL, 502–503
 SS diagnosis and lymphoma, 508
 treatment, 584
- Distal RTA (dRTA)
 and IN, 275
 latent, 274

- pathogenesis, pSS, 278
 - SS patients, 278
 - DLBCL. *See* Diffuse large B-cell lymphomas
 - Dry mouth, SjS
 - acidic beverages, 545
 - anticholinergic side effects, 542
 - “burning mouth” syndrome, 542
 - cevimeline effects, 555–556
 - chemoprophylaxis and polyol sweeteners, 550
 - cholinergic agonists, 556
 - dry skin treatment, 559–560
 - evaluation and treatment, rheumatologist, 551
 - fluoride, 550
 - functional circuit, 542
 - home products, safety, 551
 - lip balms, 543
 - measures, 544
 - oral
 - candidiasis, 557–559
 - hygiene, 551–553
 - pH maintenance, 545
 - pilocarpine benefit, 554
 - radiation therapists, 545
 - recommendations, 543–544
 - saliva content and stimulation, 546–547
 - salivary glands, 542
 - terminology, oral medicine, 547–549
 - time of surgery, 562–563
 - TNF-alpha blockers
 - anti-CD20 antibody, 557
 - B-cells, 557
 - treatments, 556–557
 - vaginal dryness
 - and dyspareunia, 553
 - estrogen replacement, 561–562
 - sexual considerations, 561
 - yeast infection, 561
 - Dysphagia
 - esophageal candidiasis, 234, 235
 - peristaltic velocity, 234
- E**
- Ear, nose and throat manifestations
 - audiometric evaluation, 124
 - hearing loss, 123–124
 - laryngopharyngeal and tracheal
 - bamboo node, 127
 - crusting, 128
 - dry mouth and xerostomia, 126
 - granulomatous and nongranulomatous nodules, 127
 - hoarseness and dysphonia, 127
 - vocal cord lesions, 128
 - sensorineural hearing loss, 124
 - sinus and nasal
 - epistaxis, 126
 - hypofunctioning, 125
 - nasal dryness, 126
 - Technetium 99m scan (^{99m}Tc), 125
 - UPSIT test, 126
 - Endocrine involvement
 - axis
 - HPA, 333
 - HPG, 333–336
 - HPT, 338
 - description, 332
 - hormonal treatment, SS, 338–339
 - immune-neuroendocrine system, SS
 - impact, stress, 333
 - proinflammatory cytokines, 332
 - prolactin and SS
 - hyperprolactinemia, 336–337
 - role, 337
 - Epidermal nerve fiber biopsy (EFNB), 143
 - Erythrocyte sedimentation rates (ESRs)
 - circulating gammaglobulins, 358
 - CRP, 358
 - primary SjS, 358
 - ESSDAI. *See* EULAR SS disease activity index
 - Etanercept
 - effect, 616
 - interferon (IFN)-alpha activity, 617
 - T cell adhesion molecules, 616
 - EULAR SS disease activity index (ESSDAI), 450–453, 455, 456
 - European league against rheumatism (EULAR) initiative, pSjS
 - indices, 455
 - multiple regression model, 456
 - poles, SS spectrum, 454
 - relationship, curves and plots, 456–457
 - symptoms, patients, 455–456
 - Experimental therapies, Sjögren’s syndrome
 - gene therapy, 634–635
 - lacrima and salivary glands, 623
 - molecular targets, therapeutic interventions
 - B and T cell receptors, 633–634
 - B cell activating factors, 633

- Experimental therapies, Sjögren's syndrome (*cont.*)
- B cell depletion, 627, 629
 - cytokines, 630–633
 - development, cytokine-directed drugs, 627, 628
 - interferons, 627–630
 - rheumatoid arthritis (RA), 627
 - phase I and II trials, 636
 - progression and disease activity
 - labial/parotid tissue, 627
 - saliva, 624–626
 - serum, 626–627
 - stem cell therapy, 635–636
- Eye-associated lymphoid tissue (EALT), 110
- F**
- Fatigue, primary Sjögren's syndrome
- assessment
 - FACIT, 132
 - FSS, 132, 135
 - helplessness, 135
 - MCID, 132
 - Profile of Fatigue and Discomfort (PROFAD-SSI), 132
 - questionnaires, measurement, 133–134
 - cognitive symptoms and depression
 - affective disorder, 137
 - "brain fog", 138
 - brain imaging studies, 138–139
 - cerebral perfusion abnormalities, 138
 - epidemiology
 - "neurasthenia", 131
 - persistent and abnormal, 132
 - fibromyalgia (FM)
 - dysregulation, stress response system, 141
 - EFNB, 143
 - features, 140
 - nociceptive and neuropathic pain, 142
 - pain mechanisms, 141
 - polysymptomatic distress, 141
 - small-fiber neuropathy, 142
 - management, 143
 - prevalence and impact, primary SS
 - hypothesized individual risk factors, 136
 - oral sicca severity, 137
 - seropositive and seronegative patients, 135
 - SF-36, 136–137
 - sleep quality, and neuroendocrine function
 - adrenal axis hypofunction, 139
 - animal models, 140
 - cardiovascular reflexes, 139–140
 - IL-6, 140
 - polysomnography and actigraphy, 139
- Fibromyalgia (FM)
- dysregulation, stress response system, 141
 - EFNB, 143
 - features, 140
 - nociceptive and neuropathic pain, 142
 - pain mechanisms, 141
 - polysymptomatic distress, 141
 - small-fiber neuropathy, 142
- FM. *See* Fibromyalgia
- G**
- Gammaglobulin, primary SjS
- hypogammaglobulinemia and immunoglobulin deficiency, 360
 - monoclonal immunoglobulins
 - clinical significance, 361
 - detection, monoclonal B-cell population, 360
 - IgMk and IgGk, 361, 362
 - peripheral monoclonal expression, 361–362
 - prevalence, 361
 - polyclonal hypergammaglobulinemia
 - classification and parameters, prevalence, 359, 360
 - description, 359
 - immunological marker, 359
- Gastroesophageal reflux (GER), 234–235
- Gastrointestinal involvement, primary Sjögren's syndrome
- celiac disease, 237
 - chronic gastritis
 - anti-PCA antibodies, 235, 236
 - prevalence, 235
 - diseases, 234
 - dysphagia
 - esophageal candidiasis, 234, 235
 - peristaltic velocity, 234
 - GER (*see* Gastroesophageal reflux)
 - Helicobacter pylori* infection
 - gastric lymphomagenesis, 237
 - seroprevalence, dyspeptic patients, 236
 - inflammatory mucosal response, 238
 - intestinal vasculitis
 - biopsy-proven vasculitis, 238
 - cryoglobulins, 238
- Gene therapy
- exocrine component, SjS, 635
 - restore normal function, 634
- GER. *See* Gastroesophageal reflux

- Glomerulonephritis (GN), pSS
 clinical features, 279–280
 description, 279
 differential diagnosis, 282
 histology, 280–281
 pathogenesis, 281–280
 treatment, 282–283
- Granulocyte colony-stimulating factor (G-CSF), 369
- Gynecological and reproductive complications, pSS
 description, 343
- NLS
 epidemiology and clinical features and CHB, 345–347
 maternal and fetal outcomes, 347–348
 pathogenesis, CHB, 350–351
 pregnancy management, anti-Ro/La autoantibodies, 351–352
 risk factors, 349–350
- SS, 344–345
- H**
- Health-related quality of life (HRQoL), pSJS
 measures, “disease-specific”
 correlations, 466
 domains, EQ–5D, 464
 domains, OHIP, 465
 importance, 465
 NHP and SIP, 465
 rheumatoid arthritis, 464
 predictors, SF–36, 464–466
 predictors, WHOQoL
 measures, global and objective, 467
 stepwise regression analysis, 469
 symptomatic measures, 468
 total damage score, 467
 variables, correlation analysis, 468
- SF–36 questionnaire
 difference and comparison, 463–464
 domains, 463
 therapeutic interventions
 androgen deficiency, 470
 cyclosporine ophthalmic emulsion, 469
 efficacy, rituximab, 469
 non-pharmacologic approach, 470
- Heart rate variability (HRV)
 and BRS, 321, 326
 cardiovascular autonomic function, 321
- Hepatitis C virus (HCV)
 infection, 361–362
 and parvovirus B19, 368
- Highly active antiretroviral treatments (HAART), 60–61
- High-resolution computed tomography (HRCT)
 features, 209
 ILD pattern, 208
 lung determinations, 201
 parenchymal lung abnormalities
 detection, 206
 pSS patients, 208
- HPA axis. *See* Hypothalamus-pituitary-adrenal axis
- HPG axis. *See* Hypothalamus-pituitary-gonadal axis
- HPT axis. *See* Hypothalamus-pituitary-thyroid
- Human leukocyte antigen (HLA)
 HLA-DR3, 362, 369
 light chain, 362
- Human T-lymphotropic virus type I (HTLV-I), 61
- Hyposalivation
 chronic complications
 angular stomatitis, 97
 bacterial sialadenitis, 98
 candidiasis, 97–98
 dental caries, 94–95
 denture-induced stomatitis, 96–97
 oral functional impairments, 95
 oral infections, 95–96
 oral ulceration, 98–99
 periodontal health, 95
 clinical features, 91–92
 defined, 86
 examination, 92
 management, 94
 oral complaints, 91
 quality of life, effect, 93–94
 reduced salivary output, 92
 signs, clinical
 dry mouth, food residues, 92, 93
 lobulated tongue, 93
- Hypothalamus-pituitary-adrenal (HPA) axis
 basal ACTH and cortisol levels
 creation, 333
 hormonal alterations, SS, 338
- Hypothalamus-pituitary-gonadal (HPG) axis
 androgens and estrogens role,
 333–334
 DHEA and DHEAS, 335
 dihydrotestosterone, estrogen and
 testosterone, 334
 interaction, CRISP–3 gene, 335, 336
- Hypothalamus-pituitary-thyroid (HPT) axis, 338

I

- Immunological tests, primary SjS
 - ACA, 419
 - ANA, 414–415
 - ANCA, 420
 - antibodies, anti-SSA/Ro and-SSB/La, 415
 - anti-citrullinated antibodies
 - and anti-keratin, 421
 - defined, CCP, 420
 - anti-DNA antibodies, 415–416
 - anti-RNP antibodies, 417–418
 - anti-Scl70 antibodies, 419
 - anti-Sm antibodies, 417
 - aPL, 418–419
 - B-lymphocyte hyperactivation, 413
 - clinical significance, 414
 - complement, 421–423
 - evaluation, 414
 - nonnuclear antigens
 - antimitochondrial and anti-parietal cell antibodies, 415
 - prevalence and clinical significance, 416
 - rheumatoid factor and cryoglobulins, 421
- Immunosuppressive agents, Sjögren's syndrome
 - interferon- α
 - cladribine, 600–601
 - diquafosol, 600
 - fingolimod, 601
 - mizoribine, 599
 - mycophenolic acid (MPA), 598
 - neurological symptoms, 598
 - refamipide, 599–600
 - restore salivary gland function, 597
 - leflunomide (LEF), 595
 - oral agents, disease modifying activity, 595, 596
- Immunosuppressive and immunomodulatory drugs
 - antimalarials, 590
 - azathioprine, 591
 - cyclophosphamide, 591–592
 - cyclosporine, 592
 - effect, 589–590
 - glucocorticoids, 591
 - methotrexate, 592
 - NSAID, 591
 - SS management, 592
 - symptoms, Sjögren's syndrome (SS), 589
- Inclusion body myositis (IBM)
 - and SS, 154
 - vacuolar myopathic degeneration, 155

Infliximab

- therapeutic approaches, 614, 615
- TNF-alpha blockers, 615

Interferons

- immunomodulating properties, 627
- intracellular pathogens, 630, 631
- visual analogue scale (VAS), 629

Interstitial lung disease (ILD)

- clinical features, 206
- COP, 206
- CTD, 202
- follicular bronchiolitis, 205
- imaging studies
 - parenchymal bullae and cysts, 208
 - respiratory involvement HRCT patterns frequency, pSS, 206, 207
- LIP, 205–206
- NSIP, 203–204
- pathology, 203
- pulmonary function tests, pSS-associated, 208
- UIP, 204–205

Interstitial nephritis (IN), pSS

- clinical and laboratory determinations, 275, 276
- description, 274
- differential diagnosis, 278
- and dRTA, 275
- histology, 277
- pathogenesis, 277–278
- patient plain abdominal film, 275, 276
- treatment, 278–279

K**KCS. See Keratoconjunctivitis sicca****Keratoconjunctivitis sicca (KCS)**

- Bijsterveld score, 384
- defined, 380, 381
- dry eyes, 525–526
- fluorescein staining, 385, 386
- rose bengal staining, 383
- schirmer test, 529
- treatment, 531
- validation, DEQ, 381

L**Laboratory abnormalities, primary**

- Sjögren's Syndrome
 - description, 358
 - hematological
 - AIHA, 364–365

- aplastic anemia, 365
 - eosinophilia, 370
 - leukopenia, 366–367
 - lymphopenia, 367–368
 - MDS, 366
 - neutropenia, 368–370
 - normocytic anemia, 363–364
 - pernicious anemia, 366
 - PRCA, 365–366
 - thrombocytopenia, 370
 - serum proteins
 - acute phase reactants, 358
 - b₂-microglobulin, 362–363
 - gammaglobulins, 359–362
 - Laboratory abnormalities, pSS
 - cytopenias, 371
 - hematological
 - AIHA, 364–365
 - aplastic anemia, 365
 - eosinophilia, 370
 - leukopenia, 366–367
 - lymphopenia, 367–368
 - MDS, 366
 - neutropenia, 368–369
 - normocytic anemia, 363–364
 - pernicious anemia, 366
 - PRCA, 365–366
 - thrombocytopenia, 370
 - serum proteins
 - acute phase reactants, 358
 - gammaglobulins, 359–362
 - β₂-microglobulin, 362–363
 - SjS, 358
 - Lacrimal functional unit (LFU)
 - components, 108
 - dysfunction treatment
 - artificial tears, 114
 - consensus and evidence-based algorithms, 113
 - dry eye severity grading scheme, 114, 115
 - management recommendation, dry eye, 116
 - ocular surface prosthesis, 114
 - punctal occlusion, 114
 - LD. *See* Linkage disequilibrium
 - Leflunomide (LEF)
 - immunosuppressive agent, 595
 - liver function test abnormalities, 596
 - therapy, 596
 - Leukopenia, primary SjS
 - positive autoantibodies vs. cytopenias, 367
 - prevalence, 366–367
 - LFU. *See* Lacrimal functional unit
 - Linkage disequilibrium (LD)
 - description, 16
 - SNPs, human genome, 17
 - variable patterns, 16
 - Liver involvement, Sjögren's syndrome
 - AIH, 248
 - algorithm, diagnosis, 249–250
 - HCV Infection and sicca syndrome, 248–249
 - LFT, 244
 - PBC (*see* Primary biliary cirrhosis)
 - Lymphocytic interstitial pneumonia (LIP), 205–206
 - Lymphoma, SS
 - B-cell subsets, 509
 - British SS cohort, 510–511
 - chemokine expression, 504
 - disease activity, 509
 - MALT and DLBCL, 502–503
 - medication, 510
 - NHL development, 504
 - RA and SLE, 510
 - risk factors
 - B cell transformation, 505–508
 - CD4+ T-lymphocytopenia, 505
 - DLBCL and MALT, 508–509
 - dose effect, 504
 - lip biopsies, germinal centers, 506
 - risk levels
 - pSS population and cohort, 502
 - sicca syndrome, 501
 - SIR and NHL, 501–502
 - serum immunoglobulins, 504
 - Lymphopenia, primary SjS
 - prevalence, CD4+, 367
 - viral infections, 368
- M**
- Magnetic resonance cholangiography (MRCP), 255
 - Magnetic resonance imaging (MRI)
 - APS, 297
 - axial CT image, 402
 - cerebral changes, 292
 - classification, 404
 - CNS lesions, 299
 - comparison, findings, 402
 - defined, 401
 - modalities, 402
 - MS lesions, 296

- Magnetic resonance imaging (MRI) (*cont.*)
 T2-weighted, 293, 294
 ultrasonography, 403
- MALT. *See* Mucosa-associated lymphoid tissue
- Marginal zone B-cell lymphoma (MZBCL), 502–503
- MCTD. *See* Mixed connective tissue disease
- MDS. *See* Myelodysplasia
- Meibomian glands and eyelids, 110
- β_2 -Microglobulin, primary SjS
 dTRA, 363
 and extraglandular involvement, 362
 levels, 362
 transmembrane glycoproteins, 362
- Mixed connective tissue disease (MCTD), 488
- Mucosa-associated lymphoid tissue (MALT) lymphomas
 extranodal marginal zone lymphomas
 hashimoto thyroiditis, 573
 international prognostic index, 578
 lactate dehydrogenase (LDH), 575
 lymphoepithelial lesion, 573, 574
 neoplastic marginal zone cells, 576
 neoplastic process, 575, 576
 staging, 575, 577
 histological transformation, 583
 low-grade lymphomas, 509
 pSS, 502
 salivary glands, 503
 therapeutic approaches
 alkylating agents, 579
 anti-CD20 treatments, 579
 chemotherapy, 578–579
 management, SS-associated MZ lymphomas, 580
- Multiple sclerosis (MS)
 CNS damage, 294
 diagnosis, 295
 disability cause, 295
 and PML, 297
 and SLE, 294, 297
- Musculoskeletal involvement
 arthralgias and arthritis, 152
 description, 151
 differential diagnosis
 patients diagnostic algorithm, arthritis and sicca symptoms, 153
 sarcoidosis, 154
 expression patterns, arthritis, 152
 myalgias and myositis
 classification, pSS, 155
 SS and IBM, 154, 155
- Myelodysplasia (MDS), 366
- Myoepithelial sialadenitis (MESA), 103
- MZBCL. *See* Marginal zone B-cell lymphoma
- N**
- Neonatal lupus syndrome (NLS)
 CHB pathogenesis, 350–351
 diagnosis, 348–349
 epidemiology and clinical features and CHB
 classic periorbital rash consistent, 345, 346
 defined, 345
 dilated cardiomyopathy, 346
 heart involvement, 346, 347
 skin disease, 345
- maternal and fetal outcomes
 autoimmune disease, 347–348
 children prognosis, 348
 evolution, 347, 348
- pregnancy management, anti-Ro/La autoantibodies
 glucocorticoids, 351
 intrauterine and postnatal, CHB, 351, 352
 IVIG administration, 352
- risk factors
 anti-Ro/SSA and anti-La/SSB, 349, 350
 CHB, 349, 350
 idiotypic-anti-idiotypic network, 350
- Nephro-urological involvement
 IN
 clinical features, 275–276
 differential diagnosis, 278
 histology, 277
 pathogenesis, 277–278
 treatment, 278–279
- GN
 clinical features, 279–280
 differential diagnosis, 282
 histology, 280–281
 pathogenesis, 281–282
 treatment, 282–283
 PBS/IC, 283–285
- Neuromyelitis optica (NMO)
 MRI, 294
 MS, 296, 298
- Neutropenia
 cytopenias, 368
 description, 368
 high frequency, immunological markers, 368

- immune granulocytopenia/
 - agranulocytosis, 369
 - primary SJS, 368
 - significant predictive factor, 369
 - treatment, agranulocytosis, 368
- NHL. *See* Non-Hodgkin lymphoma
- NLS. *See* Neonatal Lupus Syndrome
- Nodal MZ Lymphomas (NMZL)
 - histology, differential diagnosis, 581
 - management
 - bone marrow transplantation, 582
 - therapeutic dilemma, 581
- Non-Hodgkin lymphoma (NHL)
 - CD4+ T-lymphocytopenia, 505
 - development, primary SS, 504, 508
 - sicca syndrome, 501
 - SS cohorts, 502
- Non-specific interstitial
 - pneumonia (NSIP)
 - characterization, 203
 - UIP and LIP, 204
- Nonsteroidal anti-inflammatory drugs (NSAIDs), 591
- Non-vasculitic cutaneous involvement
 - description, 159
 - epidemiology, 162–163
 - primary SJS, skin changes
 - annular erythema, 164–166
 - Bcl, 168–170
 - eyelid dermatitis, 166–167
 - panniculitis, 167
 - PNCA, 167–168
 - pruritus, 164
 - xerosis, 163
 - skin changes, secondary SJS
 - lupus erythematosus, 170–171
 - RA, SSC and idiopathic inflammatory myopathies, 171
 - skin change types classification, SJS
 - patients, 160–161
- Normocytic anemia, primary SJS
 - cause, patients, 364
 - cell membrane expression, 364
 - defined, 363
 - and extraglandular manifestations, 363–364
- O**
- Ocular involvement
 - conjunctiva
 - conjunctival-associated lymphoid tissue (CALT), 110
 - Goblet cells, 109
 - topographic zones, 109
 - cornea, 110
 - dysfunction mechanism
 - lacrimal gland, 111
 - ocular surface, 111–112
 - lacrimal functional unit
 - MUC5AC, 108
 - tear film instability and corneal epithelial disease, 108
 - lacrimal gland
 - mucosal-associated lymphoid tissue (MALT), 109
 - sympathetic nerves, 109
 - meibomian glands and eyelids, 110
 - neural innervation, 110–111
 - Sjögren's syndrome
 - corneal epithelial filaments, 113
 - diffuse punctate staining, 112–113
 - fluorescein tear breakup time, 112
 - lissamine green and rose bengal dyes, 113, 114
 - Schirmer test, 112
- Ocular tests, primary SJS
 - cytology, 388
 - destruction, lacrimal gland, 382
 - ferning test, 387
 - fluorescein, 385, 386
 - lissamine green, 385, 386
 - methods, dry eye evaluation, 382
 - rose bengal, 384–385
 - Schirmer, 383–384
 - tear break-up time, 385, 387
 - tear meniscus, 387
 - tear osmolarity, 387
 - tear proteins, 387
 - vital dyes, 384
- Oral involvement
 - hyposalivation
 - chronic complications, 94–99
 - clinical features, 91–92
 - examination, 92
 - management, 94
 - quality of life, effect, 93–94
 - signs, clinical, 92–93
 - orofacial manifestations
 - neurological involvement, 89
 - salivary involvement, 87, 89
 - salivary flow
 - Saxon test, 86
 - USFR, 86
 - salivary gland enlargement
 - causes, 100

- Oral involvement (*cont.*)
- facial palsy, 99, 102
 - swelling, 101
 - salivary swelling
 - forms, 103
 - MESA, 103
 - parotid glands, 102
 - sialochemical changes, SS
 - prostaglandin PGE-2, saliva, 90
 - proteins, minor salivary glands, 91
 - unstimulated whole saliva (UWS)
 - flow, 89
 - xerostomia (*see* Xerostomia)
- Oral tests, primary SjS
- determination, salivary gland function, 388
 - impression cytology, 390
 - reaction, iodine-starch, 390
 - Saxon, 389–390
 - sialometry, 388
 - wafer, 389
 - whole saliva flow collection, 389
- P**
- PAH. *See* Pulmonary arterial hypertension
- Painful bladder syndrome/interstitial cystitis (PBS/IC) and pSS
- clinical, cytoscopic and histologic features, 283
 - differential diagnosis, 284
 - ESSIC, 283
 - pathogenesis and association, SS, 284
 - treatment, 285
- PBC. *See* Primary biliary cirrhosis
- PCNA. *See* Primary cutaneous nodular amyloidosis
- Peripheral neuropathy
- axonally polyneuropathies, 305
 - electrophysiologic determination, SS, 305
 - multiple mononeuropathy/mononeuritis multiplex, 306
 - prevalence and classification
 - description, 303
 - patterns, SS, 304
 - sensory neuropathies subsets, 304
 - restless leg syndrome, 309
 - SAN, 307–308
 - sensorimotor demyelinating polyneuropathy, 305–306
 - small fiber painful sensory
 - description, 308
 - patterns, 309
- PNCA. *See* Primary nodular cutaneous amyloidosis
- Polymerase chain reaction (PCR), 299
- Polymorphonuclear leukocytes (PMN)
- activation, 369
 - serum counts, 369
- PRCA. *See* Pure red cell aplasia
- Primary biliary cirrhosis (PBC)
- animal models
 - NOD.c3c4 mouse, 246
 - Treg cells, 247
 - xenobiotics, 247
 - definition and diagnosis, 245
 - epithelium involvement, 246
 - histology and serology
 - anti-pyruvate dehydrogenase complex, 248
 - autoantibody profiles, 247
 - serum ANA, 247
 - similarities, differences and overlap, 245–246
- Primary cutaneous nodular amyloidosis (PCNA)
- immunoglobulin light chains deposition, 167
 - SjS-associated, 167–168
- Primary nodular cutaneous amyloidosis (PNCA)
- immunoglobulin, 167, 168
 - leg, 167, 168
- Primary Sjögren's syndrome (pSjS). *See also* Laboratory abnormalities, pSS
- IN, 274–279
 - AD pathogenesis, 315–318
 - arthritis, 154
 - arthropathy, 152
 - articular involvement, extra-glandular manifestation, 152
 - autoimmune epithelitis, 515
 - autoimmune exocrinopathy, 524
 - autoimmune lesions, characteristics
 - BAFF, 37
 - IgG concentrations, 37
 - incidence, inflammatory cells, 35
 - lymphocytic infiltrates, 34
 - mononuclear cell types, incidence, 36
 - T and B lymphocytes, 36
 - T cell receptor (TCR) repertoire, 36–37
 - autonomic dysfunction, 191
 - cardiac involvement, 187
 - comorbidities and occupational disability
 - adverse effects, 530
 - non adults, 530–531

- diagnosis
 - classification criteria, 528–529
 - immunological tests, 530
 - keratoconjunctivitis sicca, 529
 - laboratory features, 530
 - salivary gland biopsy, 529
 - xerostomia, 529
- diagnostic criteria
 - and classification, 4
 - variations, occurrence, 4
- ECG study, 189
- epithelial cells, autoimmune response
 - regulators
 - exosomes, 40
 - immunoactive molecules, 39, 40
 - molecules and cellular products, 38, 39
 - salivary glands, 38
 - TLR signaling, 40
- etiopathogenic factors, 34
- extraepithelial manifestations, 516
- extraglandular organ involvement, 34
- fatigue and myalgias, 154
- glucocorticoids, 516
- GN, 279–283
- hyperprolactinemia, 336–337
- incidence
 - adult populations, 4
 - age and sex distribution, Greek study, 5
 - population-based study, Olmsted County, 5
- information, family doctor, 532–533
- key recommendations, practice, 532
- mild synovitis, 153
- model, SjS pathogenesis, 47
- mortality and causes, deaths
 - cyclophosphamide and glucocorticoids, 517–518
 - Geek patients, 517
 - hemodialysis, 517
 - lymphoproliferative disorders, 516
 - MALT, 516
- musculoskeletal manifestations, 151
- non-sicca manifestations, 525
- ocular dryness, 524
- PAH, 190
- pathogenetic factors
 - environmental factors, 44–46
 - genetic predisposition, 44
 - hormonal, 46
- patient diagnostic algorithm, AD, 325–326
- PBS/IC, 283–285
- prevalence
 - adult populations, 6
 - ethnic differences role, 7
 - systematic recording, diagnosed cases, 6
 - tricyclic antidepressants and antipsychotic agents, 7
- prognostic factors
 - cryoglobulins evaluation, 519
 - lymphoproliferative disorders, 518–519
 - parotid gland enlargement, 519–520
 - rheumatoid factors (RF), 518
 - serum monoclonality, 518
- pulmonary function tests
 - airway disease, 202
 - ILD, 208
- stress impact, 333
- symptoms
 - joint disease, 527
 - keratoconjunctivitis sicca, 525–526
 - systemic dryness, 527
 - vascular disease, 527
 - xerostomia, 526–527
- symptoms, impaired autonomic nervous function, 313
- tissue injury and repair
 - extracellular matrix and tissue damage, 42–44
 - functional impairment and autonomic nervous system, 41–42
- treatment
 - hydroxychloroquine, 532
 - keratoconjunctivitis sicca, 531
 - xerostomia, 530–531
- Primary SS (pSS). *See also* Primary Sjögren's syndrome (pSjS)
 - CNS, 292, 294
 - MRI, 292, 293
 - peripheral nervous system, 291
- Progressive multifocal leukoencephalopathy (PML)
 - and MS, 297
 - viral neurologic disease, 297
- pSS. *See* Primary Sjögren's syndrome
- Pulmonary arterial hypertension (PAH)
 - description, 190
 - management, pSS, 192
 - pSS treatment algorithm, 192, 193
- Pulmonary involvement
 - airway disease
 - imaging studies, 201
 - mucus secretion, 200–201
 - pathology, 201
 - pulmonary function tests, pSS-associated, 202

- Pulmonary involvement (*cont.*)
 diagnosis and management, 208–209
 ILD, 202–208
 pleuritis, 208
 respiratory system type, SS, 200
- Pure red cell aplasia (PRCA)
 description, 365
 erythropoietic-inhibitor IgG and cytotoxic lymphocytes, 365–366
- Q**
- QoL measurement, primary SjS
 autoantibodies, blood, 461–462
 concept, 462
 HRQoL
 measures, “disease-specific”, 464–466
 predictors, SF–36, 464–466
 predictors, WHOQoL, 467–469
 SF–36 questionnaire, 463–464
 therapeutic interventions, 469–470
 SF–36, patients, 463
 uses, 462–463
- R**
- RA. *See* Rheumatoid arthritis
 Raynaud’s phenomenon (RP), 325
- Renal tubular acidosis (RTA)
 dRTA, 274, 275, 278–279
 patients, proximal, 279
 patient, SS, 274
- Rheumatoid arthritis (RA)
 disease activity, 509
 KCS, 481
 lymphoproliferative disease, 510
 prevalence, sicca symptoms, 482
 pSS and RA-SS
 clinical and serological difference, 482–484
 genetic difference, 484
 seronegative, 152
 skin changes, SSc and idiopathic inflammatory myopathies, 171
 SLE and arthropathies, 153–154
 Tarpley’s classification, 482
- Rheumatoid factor (RF)
 and ANA, 334
 and anti-Ro/SS-A antibody, 359, 366–367
 patients, elevated ESRs, 358
- S**
- Saliva
 autoimmune process, 624
 intervention therapy, 626
 mean salivary flow rates, 624, 625
 submandibular/sublingual (SM/SL) flow rates, 626
- Salivary gland biopsy, primary SjS
 Daniels’ technique, 405
 focus score
 cigarette smoking, 408
 defined, 406
 histopathologic classification, 407
 multiple tissue levels, 408
 nodular lymphocytic infiltration, 407
 sensitivity and specificity, 406
 standardization, 406–407
- labial
 biopsy results, 409
 description, 404
 symptoms and positive serology, 408
- lacrimal, 406
 punch and major, 405
- Schirmer test, 112
- SCLE. *See* Subacute cutaneous lupus erythematosus
- Sensory ataxic neuropathy (SAN)
 causes, 307, 308
 defined, 307
 diagnostic and therapeutic algorithm, SS, 308
 paraneoplastic, 307
 treatment regimens, 308
- Serum proteins
 acute phase reactants, 358
 circulation, monoclonal immunoglobulins
 HCV infection, 361–362
 prevalence, 360, 361
 hypogammaglobulinemia and immunoglobulin deficiency, 360
 β_2 -microglobulin
 HLA, 362
 levels, 362–363
 polyclonal hypergammaglobulinemia, 359–360
- Sialadenitis, 98, 99
- SICCA. *See* Sjögren’s international collaborative clinical alliance
- Single photon emission computerized tomography (SPECT), 293

- SIR. *See* Standardized Incidence Ratio
- Sjögren's international collaborative clinical alliance (SICCA), 440
- Sjögren's syndrome (SjS). *See also* Ocular involvement
- arguments, "T-cell disease", 68
 - axonally polyneuropathies, 305
 - B cells
 - chemokines and antibody production, 74–75
 - cytokines, 70–71
 - IgA autoantibodies and cytokines, 75–76
 - intrinsic abnormalities, 76
 - monoclonal expansion, 74
 - ontogeny, lymphocytes, 71–72
 - subpopulations, 72–73
 - B lymphocytes, 68
 - cancer
 - associated disorder, SLE, 500
 - autoimmune disease, 500
 - disease activity, 509
 - lymphoma, 501–510
 - malignancy and types, 500–501
 - reverse causality, 501
 - candidate genes and pathogenesis
 - allele frequencies, 23
 - antibody responses, 24–25
 - BAFF influences, 26
 - components, immune system, 18
 - early B cell factor 1 (EBF1), 26
 - FAM167A and BLK, 25
 - HLA association, 22
 - interferon (IFN) activation, 23
 - multifunctional proteins, 22
 - non-HLA association studies, 19–22
 - plasmacytoid dendritic cells (pDCs), 24
 - STAT4 protein, 24
 - TNFSF4, 25
 - description, 3, 291, 343, 358
 - gene expression profiling (GEP), 12
 - gene expression studies
 - cytokine and chemokine signaling, 27
 - "IFN signature", 27
 - mRNA, 26–27
 - genetic epidemiology
 - candidate genes, 12
 - human leukocyte antigen (HLA), 13
 - twin concordance rate, 13
 - genetics, transcriptomics and proteomics
 - central dogma, information flow, 14
 - DNA sequence variation, 15
 - genome-wide association (GWA) scans, 17, 18
 - human genome, 14, 15
 - imprecision, allele frequency, 18
 - linkage disequilibrium (LD), 16, 17
 - mapping genetic effects, 17
 - SNPs, 15–16
 - variant association calculation, 16
 - gynecological manifestations
 - dyspareunia and pruritus, 344
 - problems, patients, 344, 345
 - HCV-SjS patients, 362
 - hormonal treatment, 338–339
 - identification and treatment, skin changes, 159
 - lymphocytes, 68
 - mean prevalence, skin disorders, 162
 - patient, diagnostic algorithm, 298–299
 - peripheral neuropathy, 303
 - primary, 3–7, 163–170
 - and prolactin, 336–337
 - protein expression studies, 27–28
 - restless leg syndrome, 309
 - reticulosarcoma development, 360
 - secondary, 170–171
 - small fiber neuropathy patterns, 309
 - T-cell infiltrates, 68
 - T cells
 - germinal center evolution, 69
 - Th-1 and Th-2 cells, 70
 - T lymphocytes, 69
- Sjögren's syndrome and associations
- autoimmune diseases, 488–492
 - classification, 476
 - nomenclature, 476
 - RA (*see* Rheumatoid arthritis (RA))
 - SLE, 476–481
 - SS-SSc, 485–487
 - symptoms, sicca, 476
 - systemic/extraglandular, 475
- Sjögren's syndrome (SjS) and IgG4-related disease (IgG4-RD)
- autoimmune pancreatitis
 - AIP, 257–258
 - clinical features, 258–259
 - diagnostic criteria, 261

- Sjögren's syndrome (SjS) and IgG4-related disease (IgG4-RD) (*cont.*)
- imaging, 259
 - pathology, 259–260
 - serology, 259
 - chronic pancreatitis, 254
 - clinical characteristics, 254, 255
 - clinical manifestations
 - CT, 262, 264
 - submandibular, parotid and lacrimal glands, 262, 263
 - diagnostic criteria, 266
 - exocrine and endocrine portions, 254
 - nomenclature, 261
 - pancreas involvement
 - autoantibodies, 256
 - clinical presentation, 255–256
 - imaging studies, 257
 - pancreatic enzymes, 256
 - pathology, 256–257
 - pancreatic injury and exocrine dysfunction, 266–267
 - pathogenesis, 266
 - pathology
 - lymphoplasmacytic infiltrate, 263, 265
 - positive staining, IgG4-bearing plasma cells, 265
 - serological issues, 263
- SjS. *See* Sjögren's syndrome
- SLE. *See* Systemic lupus erythematosus
- SS associated with systemic sclerosis (SS-SSc)
- comparison, patients, 487
 - cryoglobulinemia, 486
 - ENA testing, 487
 - extra glandular features, 486
 - patient, RA, 485
 - prevalence and parotid enlargement, 485
 - salivary gland biopsy, 486
 - symptoms, sicca, 487
- SSc. *See* Systemic sclerosis
- SS disease activity index (SSDAI), 448, 449, 454–456
- SS disease damage index (SSDDI), 448–450
- SS-SSc. *See* SS associated with systemic sclerosis
- Standardized Incidence Ratio (SIR)
- malignancy, 510
 - non-hematological tumors, 510–511
 - patient cohort, 502
 - primary SS, 501–502
- Stem cell therapy
- submandibular gland function, 636
 - tissue formation, 635
- Subacute cutaneous lupus erythematosus (SCLE)
- annular lesions, 170
 - and RP, 162
- Systemic lupus erythematosus (SLE)
- anti-Ro and-La antibodies, 479
 - aplastic anemia, 365
 - associated disorder, 500
 - and CNS, 337
 - comparison, 477
 - diseases
 - autoimmune, 509
 - lymphoproliferative, 510
 - exposure, mothers, 347–348, 352
 - hormonal and reproductive factors, 511
 - immune-neuroendocrine communication, 332
 - lymphocytic filtration, 478
 - β_2 -microglobulin, increased serum levels, 362
 - and MS, 294, 299
 - perivascular infiltration, 481
 - PML, 297
 - pSS, systemic autoimmune diseases, 187
 - RA and arthropathies, 153–154
 - Raynauds phenomenon and CNS involvement, 481
 - salivary duct dilatation and acinar tissue destruction, 479
 - serological difference, 481
 - sicca, clinical expression, 480
 - and SS, 333–334
 - and SS, relationship, 476
- Systemic sclerosis (SSc)
- PBC, 246
 - skin changes, 171
- T**
- Tissue injury and repair, SjS
- extracellular matrix and tissue damage
 - cytokines, 43–44
 - Fas/Fas-ligand apoptotic pathway, 43
 - IFNg, 43
 - matrix metalloproteinase, 43
 - functional impairment and autonomic nervous system involvement
 - animal models, 41–42

aquaporins (AQP), 42
 exocrine secretion, 41
 M3 muscarinic receptors, 42
 peripheral tissue, 41

U

University of Pennsylvania Smell Identification Test, 126
 Usual interstitial pneumonia (UIP)
 characterization, 204
 NSIP, 204

V

Vasculitis and Sjögren's syndrome (SS)
 clinical determinations, 180
 description, 175–176
 epidemiology, 176–177
 histopathology
 acute necrotizing, 178, 179
 endarteritis obliterans, 179
 leukocytoclastic, 177, 178
 lymphocytic, 178
 laboratory determinations, 179
 and mortality, 183–184
 other organs, 183
 pathogenesis, 179–180
 peripheral and CNS
 biopsy specimens, 183
 neuropathy, 182–183
 skin
 nodules, 180, 181
 palpable purpura, hyperpigmentation,
 180, 181
 urticaria, 182
 treatment, 183–184

Viruses, SS

coxsackieviruses
 cross-reaction, 62
 lymphocytes, 61
 hepatitis B virus (HBV)
 lymphotropism and sialotropism,
 59, 60
 prevalence, 59–60
 systemic autoimmune diseases, 59
 hepatitis C virus (HCV)
 autoimmune features, 57
 B-cell hyperactivity, 59
 chronic infection, 59
 lymphotropism, 58
 sicca syndrome, 58
 herpes viruses, 62
 HTLV-I, 61
 human immunodeficiency virus (HIV)
 HAART, 60–61
 salivary gland infiltration, 60
 human parvovirus B19
 B19-DNA, 63
 leukopenia and thrombocytopenia, 62

X

Xerostomia
 causes, 88–89
 dry mouth and maxillofacial
 diseases, 90
 dry mouth symptoms, 526–527
 hyposalivation, 86
 measurement, salivary flow, 529
 older patients, 87
 pilocarpine and cevimimine, 531–532