

# Index

## A

- AAMI. *See* Advancement of Medical Instrumentation (AAMI)
- AAP. *See* American Academy of Pediatrics (AAP)
- ACE I. *See* Angiotensin-converting enzyme inhibitor (ACE I)
- Acid-base homeostasis
  - medical management, AKI, 300
  - metabolic acidosis, 69
  - organic acids and bases, 69
- Acidosis, GFR
  - adverse effects, 133
  - alkali therapy, volume contraction, 133
  - dietary intake and osteoid calcification, 133
  - metabolic alkalosis, 133
- Acute kidney injury (AKI)
  - BUN and SCr levels, 289
  - case vignette, 288
  - complications, 326
  - Cook catheter set, PD, 324
  - definitions/classification systems
    - adults, 292
    - children, 292
    - neonates, 292
  - description, 288
  - dynamic and evolving process, 289
  - epidemiology, neonate, 289–291
  - etiology and incidence
    - acquired renal disorders, 322
    - congenital malformations, 322
    - neonatal ICU, 322
    - surgical complications, 322–323
  - history and risk factors, 297
  - ICU technologies and therapies, 293
  - indications, acute PD
    - CRRT, 323
    - dialysis, neonates, 323
    - inborn errors, metabolism, 323
  - intrinsic (*see* Intrinsic AKI)
  - laboratory and radiology findings, 298
  - medical management, neonate (*see* Medical management, AKI)
  - neonatal population, 288
  - neonate, etiologies, 293–295
  - newborns at risk, 293
  - novel biomarkers, 302
  - physical examination (*see* Physical examination, AKI)
  - postrenal/obstruction, 297
  - prerenal azotemia (*see* Prerenal azotemia)
  - prescriptions
    - acute setting, 326
    - advantages, manual exchanges, 325, 326
    - and APD, 326
    - CFPD, 325
    - lactate-based solutions, 326
    - neonatal and pediatric patients, 325
    - respiratory embarrassment, 325
  - published data outcomes, 326–327
  - renal replacement modality, 323–324
  - risk, dialysate leakage, 324–325
  - staging system, 292–293
  - Tenckhoff catheter, 324, 325
- ADH. *See* Antidiuretic hormone (ADH)
- ADPKD. *See* Autosomal dominant polycystic kidney disease (ADPKD)
- Advancement of Medical Instrumentation (AAMI), 312
- AFP. *See*  $\alpha$ -Fetoprotein (AFP)
- AKI. *See* Acute kidney injury (AKI)
- American Academy of Pediatrics (AAP)
  - diagnosis and management, initial UTI, 186
  - prophylactic antibiotic treatment, 193
  - VCUG, 194
- ANF. *See* Atrial natriuretic factor (ANF)
- Angiotensin-converting enzyme inhibitor (ACE I)
  - captopril, 295
  - first and second trimesters, 297
  - nephrotoxic medications, 296
  - NSAIDs, 297
  - prenatal findings and events, 297
- Anorectal malformation (ARM)
  - cloaca, 163
  - currarino's triad, 163
  - hydrocolpos, 163, 172
  - imperforated anus, 163, 173
  - longitudinal US, 163, 172
  - and VUR, 163
- ANP. *See* Atrial natriuretic peptide (ANP)
- Antenatal Bartter syndrome, 89
- Anteroposterior (AP), 157, 161

- Antibiotic therapy, UTI  
 bacterial susceptibility, antimicrobial agents, 192  
 description, 192  
 diagnosis, 192  
 genitourinary abnormality, infants, 193  
 older infants, 193  
 treatment, pyelonephritis, 193
- Anticancer therapy, infants  
 bone marrow, 255  
 dose reductions, 256  
 factors, enhanced chemotoxicity, 255  
 NWTs-2, 256  
 renal and hepatic metabolic function, 255  
 traditional chemotherapy drugs, 255
- Anticoagulation  
 CRRT  
 citrate and calcium titration protocols, 316  
 description, 315  
 heparin-based protocols, 315  
 historical adult-based protocols, 316  
 younger patients, 316  
 HD  
 calcium manipulation, 313  
 continuous veno-venous hemofiltration, 312–313  
 heparin, 312
- Antidiuretic hormone (ADH)  
 distal tubules, 81  
 immature intracellular second messenger system, 81  
 vasopressin receptor (V2R), 81  
 water permeability, 81
- AP. *See* Anteroposterior (AP)
- ARM. *See* Anorectal malformation (ARM)
- ARPKD. *See* Autosomal recessive polycystic kidney disease (ARPKD)
- Atrial natriuretic factor (ANF)  
 cardiac myocytes, 64–65  
 fetal urinary flow and electrolyte excretion, 65  
 physiologic adaptation, 65  
 somatic growth, 65
- Atrial natriuretic peptide (ANP), 79
- Autosomal dominant polycystic kidney disease (ADPKD)  
 clinical signs and symptoms, 269  
 diagnosis, 269  
 older children and adults, 268  
*PKD1/PKD2* genes, 268  
 prenatal ultrasonographic findings, 268–269  
 prognosis, 269  
 treatment, 269  
 vascular abnormalities, 268
- Autosomal recessive polycystic kidney disease (ARPKD)  
 affected infants, 267  
 CHF, 267  
 clinical diagnosis, 267  
 counseling, future pregnancies, 267  
 kidney pathology, 266, 267  
 molecular genetic testing, 267  
*pkhd1* gene, 266  
 primary cilia, 266  
 prognosis, 268  
 pulmonary hypoplasia and Potter's facies, 267  
 renal complications, newborn period, 267  
 treatment, 267–268
- B**
- Bartter syndrome, 110
- Bladder anomalies  
 congenital bladder diverticula, 222  
 dilated, 221  
 genitourinary, 221  
 nondilated, 221  
 urachal, 222  
 and urethral duplication, 222
- Bladder development  
 molecular mechanisms, 36–37  
 normal bladder morphogenesis, 36  
 ureterovesical junction  
 bladder embryology, 39  
 LAR phosphatase function, 42  
 morphological events, 38, 39  
 nephric duct morphogenesis, 40  
 retinoic acid-dependent signals, 41  
 trigone and bladder anti-reflux mechanism, 37  
 ureter-derived smooth muscle, 39  
 ureter epithelium, 39  
 ureteric bud outgrowth, 40  
 UVJ formation, 38  
 VUR, 37
- Bladder exstrophy  
 after closure, 225  
 and epispadias, female, 225  
 growth rates, closures, 226  
 maldeveloped structures, 225
- Bladder imaging  
 anechoic fluids, 153, 154  
 high intensity fluids, 153, 154  
 normal, distended urinary bladder, 153, 154  
 valsalva, 154, 155  
 VCUG, 153, 155
- Blood pressure measurement, neonates  
 alertness and calmness, 350–351  
 automated oscillometric methods, 350  
 complexities, 351  
 cuffs, 350  
 Doppler assessment, 350  
 features, standard protocol, 351  
 intra-arterial analysis, 350  
 Korotkoff sounds, brachial artery, 350  
 normal values, 351  
 parameters, postconceptual age, 352
- Blood urea nitrogen (BUN)  
 prerenal azotemia, 293  
 and urine output, 289
- Blood vessel imaging, 344
- Body fluids  
 ANP, 79  
 composition, 79–80  
 ECF, 78

- human neonates, 79
- ICF, 78
- intrapartum hypoxia, 79
- TBW, 78, 79
- Body surface area (BSA)
  - age independent relationship, 331
  - neonate on PD, 331
- BSA. *See* Body surface area (BSA)
- BUN. *See* Blood urea nitrogen (BUN)
- C**
- CAKUT. *See* Congenital abnormalities of the kidney and urinary tract (CAKUT)
- Cancer syndromes
  - clinical characteristics, inherited, 246
  - heritable predisposition, malignancy, 246
  - “Li-Fraumeni syndrome”, 246
  - management, surveillance plan, 247
  - Wilms tumor, 247
- CCD. *See* Cortical collecting duct (CCD)
- CCSK. *See* Clear cell sarcoma of the kidney (CCSK)
- Cerebral demyelination, 105
- Chronic kidney disease (CKD)
  - acidosis, 133
  - calcium/phosphorus, 133–134
  - etiology, infancy, 129
  - forms, AKI, 125
  - nutrition and growth, 343
  - renal transplantation, 136
  - and scarring, 195–196
  - supplemental feeding, 134
- CIC. *See* Clean intermittent catheterization (CIC)
- CKD. *See* Chronic kidney disease (CKD)
- Clean intermittent catheterization (CIC)
  - and anticholinergics, 207
  - vesicostomy, 205
- Clean voided bag specimens, 190–191
- Clear cell sarcoma of the kidney (CCSK), 251
- Cloacal extrophy, 226
- CNS. *See* Congenital nephrotic syndrome (CNS)
- Computed tomography (CT), 173
- Congenital abnormalities of the kidney and urinary tract (CAKUT)
  - anatomical and functional defects, 3
  - clinical management, 1
  - diagnosis, 1
  - embryology, kidney (*see* Embryology, kidney)
  - end-stage renal disease, 1
  - gene expression and function, 1
  - genetic/molecular control mechanisms, 3
  - human congenital malformation syndromes, 1–3
  - human embryology, 1
  - origins, embryonic, 3–7
  - renal, ureter and bladder malformations, 1, 2
  - stereotypic processes, 2
- Congenital anomalies
  - hydronephrosis (*see* Hydronephrosis)
  - RCD (*see* Renal cystic disease (RCD))
  - renal ectopia, 156–158
- Congenital malformation syndromes
  - cystic dysplastic disorders, 270
  - diffuse cystic dysplasia, 271
  - human, associated with CAKUT, 2
- Congenital nephrotic syndrome (CNS)
  - case vignette, 275–276
  - classification
    - genetic forms, 279
    - nongenetic forms, 280
    - NPHS2 (*see* Podocin gene (NPHS2) mutations)
    - NPHS1 mutation, 278
  - clinical presentation, 280
  - defined, 134, 284
  - diagnosis
    - AFP, 280
    - immunohistochemistry methods, 280
    - NPHS1 and NPHS2 mutations, 281
    - renal biopsy, 280
    - renal function, 280
  - differential diagnosis, 281
  - epidemiology, 280
  - genetic defects, Neph1 gene, 136
  - genetics and pathophysiologic factors, 276
  - guidance and education, families, 284
  - medical management (*see* Medical management, CNS)
  - pathophysiology
    - adapter proteins, 276–277
    - components, glomerular filtration barrier, 277
    - glomerular basement membrane, 276
    - glomerular filtration barrier, 276
    - heterogeneous group, renal diseases, 276
    - renal filtration system, 276, 277
    - structural and regulatory proteins, 278
  - primary and secondary forms, 276
  - prognosis, 284
  - protein intake, 136
  - “stepwise treatment approach”, 281
  - surgical management (*see* Surgical management)
  - vitamin D supplementation, 136
- Conservative care, neonatal AKI
  - “humanized” concentrations, electrolytes, 137
  - indirect calorimetry, 137
  - infant’s anabolic drive, 136
  - modular additions, formulas and diets, 131, 136
  - pediatric subspecialty care, 136
- Continuous renal replacement therapy (CRRT). *See also* Hemodialysis (HD)
  - anticoagulation (*see* Anticoagulation)
  - dialysis, neonatal AKI, 137
  - extracorporeal therapies, 318
  - hypotension, 317
  - machines (*see* Machines, CRRT)
  - medication dosing, 318
  - nutritional losses and drug dosing, 317
  - nutritional stealing, 318
  - pediatric and infant groups, 318
  - prescription, 316–317
  - social supports, 319
  - solutions used for, 315

- Continuous renal replacement therapy (CRRT) (*cont.*)  
 tandem therapies, 318  
 thermic instability, 317  
 vascular access, 318  
 vasopressor agents, 318
- Continuous venovenous hemodiafiltration (CVVHDF), 288
- Cortical collecting duct (CCD), 89
- CRRT. *See* Continuous renal replacement therapy (CRRT)
- Cryptorchidism  
 assessment and management, 229–230  
 definitions, 229  
 male endocrine gland disorder, 229  
 testicular descent, 229
- CT. *See* Computed tomography (CT)
- CVVHDF. *See* Continuous venovenous hemodiafiltration (CVVHDF)
- Cystic dysplasia  
 description, 270  
 diffuse (*see* Diffuse cystic dysplasia)  
 MCDK (*see* Multicystic dysplastic kidney (MCDK))  
 renal cysts, neonates, 272
- Cystic kidney diseases  
 case vignette, 263–264  
 description, 264  
 developmental considerations, 264  
 differential diagnosis, 265–266  
 with dysplasia (*see* Cystic dysplasia)  
 inherited (*see* Inherited cystic kidney diseases)  
 morbidity and mortality, newborn, 264  
 in neonate (*see* Neonates, cystic kidney disease)  
 polycystic (*see* Polycystic kidney diseases)
- Cystography  
 bladder, 152  
 prenatal hydronephrosis, 152  
 VUR, 152
- D**
- Denys-Drash syndrome, 15, 276
- DHT. *See* Dihydrotestosterone (DHT)
- Dialysis  
 access, newborn  
 avoidance, lower body circulation, 344  
 interventional radiological techniques, 344  
 PD and HD, 343  
 filters and machines  
 Fresenius F3, 311  
 gallop rhythm warrants, 311  
 Gambro Polyflux 2H, 311  
 heat loss, 311–312  
 manual system, 312  
 proper pump speeds, 311  
 volumetric ultrafiltration monitoring systems, 311  
 neonatal AKI  
 CRRT, 137  
 energy and protein intakes, infants, 137  
 intrauterine renal failure and oliguria, 137  
 pediatric and adult patients, 301  
 renal support therapy, 302  
 RRT, 301
- Differential diagnosis, cystic kidney disease  
 congenital obstructive uropathies, 266  
 in neonate, 265, 266
- Diffuse cystic dysplasia  
 congenital malformation syndromes, 271  
 description, 270  
 isolated/familial, 270–271
- Dihydrotestosterone (DHT)  
 deficiency, 235  
 testosterone conversion, 234
- Dilated bladder anomalies, 221
- Dimercaptosuccinic acid (DMSA), 152
- Disorders of sex development (DSD)  
 diagnostic imaging, 236  
 endoscopy, 236  
 evaluation, neonates, 235  
 female assignment, 236  
 gonadal dysgenesis, 235  
 history, pregnancy, 235–236  
 initial laboratory evaluations, 236  
 intersex disorders, 233  
 management, gender assignment, 236  
 medical conditions, 233  
 normal sexual differentiation, 233–234  
 ovotesticular DSD, 235  
 revised nomenclature, 234  
 support and counseling, parents, 233  
 46XX DSD, 234, 235  
 46XY DSD, 234–235
- Divalent ion physiology  
 fetus, 90–91  
 neonates  
 calcium homeostasis, 91  
 magnesium, 92  
 phosphorus, 91–92
- DMSA. *See* Dimercaptosuccinic acid (DMSA)
- DSD. *See* Disorders of sex development (DSD)
- Duct collection system  
 branch formation, 17  
 embryonic process, 16  
 fate, tip and trunk cells, 17, 18  
 human kidney development, 17  
 kidney organ culture, 19  
 morphogenetic process, 17  
 reciprocal inductive interactions, 16  
 renal branching morphogenesis, 16  
 renal medulla and pelvicalyceal system  
 cortical growth, 22  
 embryonic kidney development, 22  
 medullary collecting duct patterning, 23  
 OCD, 22  
 renal cortex and medulla, 22  
 renin-angiotensin system, 23  
 signaling mechanisms, 23

- stimulatory renal branching morphogenesis
  - (*see* Stimulatory renal branching)
- terminal differentiation, 23–24
- ureteric bud branching, 17
- Dysplasia, renal
  - hydronephrosis, 168, 174
  - MAG3, 169, 175
  - MCDK, 168, 175
  - VCUG, 169, 175
  - VUR, 169, 175
- E**
- Early urinary tract embryology
  - embryonic time table, nephrogenesis, 3, 4
  - nephric duct and urogenital sinus, 3
  - post-gastrulation embryo, 3
- ECF. *See* Extracellular fluid (ECF)
- Echogenic fluid (EF), 160, 167
- EF. *See* Echogenic fluid (EF)
- Electrolyte disorders, newborn
  - hyperkalemia, 111–114
  - hyponatremia (*see* Hyponatremia)
  - hypokalemia, 109–111
  - hyponatremia (*see* Hyponatremia)
- Embryology, kidney
  - development, collecting system (*see* Duct collection system)
  - induction, MM, 14–15
  - morphogenesis, 7–8
  - nephron development (*see* Nephron development)
  - UB outgrowth (*see* Ureteric bud (UB) outgrowth)
- End-stage renal disease (ESRD)
  - chronic setting, 329
  - comorbidities, 328
  - complications, 334
  - dialysis care advancement, 329
  - families and pediatric nephrology, 327–328
  - health-care providers, 328
  - ideal placement, PD catheter, 330
  - incidence and etiology, neonates/infants, 327
  - Kaplan-Meier survival curve, 335
  - long-term PD, 329
  - neonatal management, infants, 200
  - nonrenal abnormalities, 328
  - nutrition in neonates
    - assessments, 332
    - initiation and advancement tube feedings, 333
    - IPPN, 333
    - negative consequences, protein depletion, 332
    - neonatal patient survival, 334
    - NG/gastrostomy tube feeding, 333
    - oral and occupational therapy, 333
    - postnatal brain growth, 332
    - post-renal transplant period, 333
    - Stamm gastrostomy procedure, 334
    - suboptimal care and/or complications, 332
  - pediatric caregivers, 328, 330
  - prescriptions
    - BSA, 331
    - Icodextrin, 331
    - lactate-buffered solutions, glucose, 331
    - pediatric peritoneal membrane, 331
    - phosphate removal, 331
    - qualitative targets, dialysis adequacy, 332
    - urea kinetic modeling ( $Kt/V_{\text{urea}}$ ), 332
  - presence, oliguria/anuria, 329
  - PUV management, infants with, 202–203
  - renal replacement modality, 329
  - renal transplantation, 201
  - RRT and PUV, 203–204
  - Tenckhoff catheters, 329–330
  - withhold/withdraw dialysis, 328
- Enlarged echogenic kidneys, 264, 265
- ERF. *See* Established renal failure (ERF)
- ESRD. *See* End-stage renal disease (ESRD)
- Established renal failure (ERF)
  - cardiorespiratory assessment, 342
  - PD and HD, 342
  - preemptive kidney transplantation, 342
  - renal dysplasia, 342
  - RRT, 342
  - vaccinations against infectious disease, 342
- Etiologies, neonatal hypertension
  - antenatal steroids and bronchodilators, 355
  - aortic coarctation, 354
  - autosomal recessive polycystic kidney, 353–354
  - bronchopulmonary dysplasia, 354
  - causes, 353–354
  - congenital renal parenchymal disorders, 353
  - congenital renal tumors, 355
  - endocrinologic causes, 354
  - Gordon syndrome, 354–355
  - “high” vs. “low” placed catheters, 353
  - Liddle syndrome, 354
  - renal artery stenosis, 353
  - renovascular disease, 353
  - surgeries, 355
  - urologic abnormalities, 354
- Exstrophy-epispadias complex
  - bladder exstrophy (*see* Bladder exstrophy)
  - cloacal exstrophy, 226
  - defects, 224–225
  - dehydration, malnutrition and electrolyte abnormalities, 226
  - etiology, exstrophy, 225
  - female epispadias, 226
  - fertility and sexual function, 227
  - kidney damage, 227
  - maldevelopment, lower abdominal wall, 224
  - prenatal diagnosis, 225
  - sexual function and libido, 227
  - staged reconstruction, closures, 227
  - urinary continence, 227
- Extracellular fluid (ECF)
  - cell membrane, 78
  - gestation, 78
  - neonatal weight loss, 79
- Extraneal®, 331
- Extravaginal torsion, 233

**F**

- Female epispadias, 226  
 FeMg. *See* Fractional excretion of magnesium (FeMg)  
 FeNa. *See* Fractional excretion of sodium and urea (FeNa)  
 Fetal renal function  
   assessment, 123–124  
   fluid and electrolyte balance, 61  
   GDNF, 60  
   glomerular and tubular function in utero, 65–71  
   hemodynamics  
     blood flow, 61–62  
     renin-angiotensin system, 62  
     small molecule mediators in utero, 64–65  
     SNS, 62–64  
   morphogenesis, 60  
   pronephric tubules, 59  
   proper nephrogenesis, 61  
   proto-oncogene *n-myc*, 61  
   sequential steps, nephrogenesis, 60  
   transitional developmental structures, 59  
 Fetal urology  
   bladder outlet obstruction, 141  
   conservative management, 148  
   cortical cysts, 143  
   cystoscopy, 146–147  
   embryology, 142  
   fast scanning MRI, 143  
   imaging, 142–143  
   megacystis, 141  
   normal nephrogenesis, 147  
   obstructive uropathy, 147  
   oligohydramnios, 147  
   open fetal vesicostomy, 144–145  
   patient selection, 144  
   prenatal ultrasound scan, male patient, 143  
   prenatal ultrasound screening, 142  
   prognostic power, 144  
   pulmonary hypoplasia, 142  
   randomization, 148  
   renal outcomes and bladder function, 147  
   treatment, lower urinary tract obstruction, 142  
   urinalysis, 143  
   urine electrolyte levels, 144  
   vesicoamniotic shunt, 145–146  
 Fetal water acquisition  
   active transport mechanisms, 80  
   aquaporins (AQPs), 80–81  
   polyhydramnios, 81  
   water flux, 80  
   water movement, 80  
 $\alpha$ -Fetoprotein (AFP), 280  
 Fluid and electrolyte balance, tubular regulation  
   acid-base homeostasis, 69–70  
   phosphate, 70  
   potassium  
     glucose, 69  
     secretory pathways, 69  
   sodium  
     gestational age, 67  
     natriuretic factors, 67

- total fractional reabsorption, 67, 68  
     transport, eukaryotic cells, 68  
 Fluid and electrolyte physiology  
   cell growth and differentiation, 78  
   distribution and composition, 78–80  
   divalent ions, 90–92  
   intrauterine and perinatal life, 78  
   organ formation and function, 78  
   potassium, 88–90  
   sodium balance, 82–88  
   water balance, 80–82  
 Focal bacterial nephritis  
   acute lobar nephronia, 188  
   renal abscess, 189  
 Fractional excretion of magnesium (FeMg), 122  
 Fractional excretion of sodium and urea (FeNa), 122
- G**  
 Galloway-Mowat syndrome, 279  
 GCKD. *See* Glomerulocystic kidney disease (GCKD)  
 Genitourinary tract anomalies, 221  
 GFR. *See* Glomerular filtration rate (GFR)  
 Glial-derived neurotrophic factor (GDNF)  
   UB outgrowth, 61  
   ureteric bud and mesenchyme, 60  
 Glomerular and tubular function in utero  
   concentration capacity, kidney development, 70–71  
   development, renal tubules, 66  
   fluid and electrolyte balance, 67–70  
   kidney, fluid and electrolyte homeostasis, 66  
   neonatal adversity, 65  
   postnatal environment, 66  
   renal tubular function, 66–67  
   vasoactive forces, 66  
 Glomerular filtration rate (GFR)  
   conditions with decreased  
     acidosis, 133  
     calcium/phosphorus, 133–134  
     growth hormone, 134  
     nutritional intake and growth, 129–131  
     potassium, 132  
     protein, 131–132  
     sodium, 132  
     supplemental feeding, 134, 135  
     water, 133  
   conditions with normal  
     congenital NS, 134–136  
     tubular lesions, 136  
   creatinine-based, 120–121  
   cystatin C, 121  
   description, 120  
   estimation, 120  
   exogenous substrate-based, 122  
   function-based dosing, 120  
   infants and children, 120  
 Glomerular proteinuria, 119  
 Glomerulocystic kidney disease (GCKD)  
   and ADPKD, 270

cysts appearance, Bowman's capsule, 270  
*HNF-1 $\beta$  gene*, 270  
 Gonadal dysgenesis, DSD, 235  
 Gordon syndrome, 354–355

## H

HD. *See* Hemodialysis (HD)

Hemodialysis (HD)

- anticoagulation (*see* Anticoagulation)
- blood flow rates, 312
- catheter longevity, 344
- clotting/blood loss, 313
- congenital renal abnormalities, 309
- CVVHD/CAVHD, 309
- dialysate flow rates, 312
- dialysis filters and machines, 311–312
- disequilibrium and hypotension, 313
- dysplasia, 308
- electrolytes, 313
- extracorporeal membrane oxygenation therapy, 308
- growth, 314
- hemodynamic changes, 343
- hyperammonemia, 309
- infants initiating dialysis, 313
- location of therapy, 309
- “medical ATN”, 308, 314
- NAPRTCS, 308
- and PD, 308, 342
- pediatric nephrological care, 308
- prescriptions, 312
- self-reported NAPRTCS database, 314
- theophylline toxicity, 309
- vascular access (*see* Vascular access)
- water supply, 312

Hernias

- assessment, 231
- indirect inguinal hernias, 231
- management, 231
- processus vaginalis, 230
- risk factors, congenital inguinal, 231

Horseshoe kidney

- neonatal MR, 156, 159
- renal parenchyma, 156, 159
- VCUG, 156, 160
- VUR, 156, 160

HUN. *See* Hydroureteronephrosis (HUN)

Hydroceles

- assessment, 230
- classification, 230
- description, 230
- management, 230
- spectrum, anatomic varieties, 230

Hydronephrosis

- ARM, 162–164
- differential diagnosis, 158
- fetus and neonate, 158
- nonoperative management
  - preoperative and postoperative differential renal function, 216

- renal ultrasound and diuretic renograms, 216
- surgical intervention, 216

operative management, without hydroureter

- infection and urinary tract obstruction, 217
- intravenous antibiotics and resuscitation, 217
- percutaneous nephrostomy tube placement, 217
- pyeloplasty, 217
- UPJ obstruction and differential renal function, 216–217

PUV, 161–162

UPJO, 158–159

ureterocele/ureteral

- bladder, 161, 168
- ectopic ureters, 159–161
- EF, 160, 167
- fluid-filled distal ureter, 161, 169
- longitudinal US, 160, 167–169
- pelvis, 160, 168, 169
- renal moieties, 159
- retroperitoneum, 160, 168
- VCUG, 160, 167, 168

VUR, 158

Hydroureteronephrosis (HUN)

nonoperative management

- description, 216
- renal ultrasound and diuretic renograms, 216
- ureteroceles with nonobstructed duplex systems, 216

operative management

- primary obstructed megaureter, 217–218
- primary obstructed ureterocele, 218

Hyperkalemia

causes, 111

clinical effects

- cardiac conduction, 112
- electrocardiographic features, 112, 113

definition, 111

neonatal, 112

pseudohyperkalemia, 112

serum potassium, 112

treatment

- emergency management, 113
- non-oliguric, 114
- sodium polystyrene, 113

Hypernatremia

breastfeeding-associated, 106

clinical manifestations, 107

definition, 106

diagnosis, 106–107

diarrheal dehydration, 106

pathogenesis, 106

potassium homeostasis, 108–109

treatment

- etiology, 108
- fluid resuscitation, 108
- hypotonic fluid infusion, 108
- idiogenic osmoles, 108
- long-term neurologic sequelae, 108
- management, 107, 108
- symptoms, hypernatremic encephalopathy, 108
- volume depletion, 108

- Hypokalemia**  
 Bartter syndrome, 110  
 causes, 109, 110  
 clinical effects, 109  
 definition, 109  
 treatment, 110–111
- Hyponatremia**  
 cerebral demyelination, 105  
 definition, 99  
 diuretics, 103–104  
 encephalopathy  
   cellular swelling, 104  
   clinical manifestations, 104  
   hypoxia, 104  
   symptomatic, treatment, 104, 105  
 evaluation, 100, 101  
 hospital-acquired hyponatremia, 102  
 hyponatremic encephalopathy, 104–105  
 impaired neonatal growth and development, 99  
 multifactorial disorder, 102  
 oral water intoxication, 103  
 parenteral fluid therapy, 99  
 pathogenesis, 99–100  
 prevention, hospital-acquired, 102  
 SIADH, 102–103
- Hypospadias**  
 classification, 227, 228  
 description, 227  
 distal/coronal, hypospadias, 228  
 DSD, 228–229  
 incidence and etiology, 227  
 management, 229  
 proximal/penoscrotal, hypospadias, 228  
 urethral meatus, 228
- I**
- ICF.** *See* Intracellular fluid (ICF)
- Infant urinary system**  
 anticancer therapy, infants, 255–256  
 CCKS, 251  
 mesoblastic nephroma, 247–248  
 MRT (*see* Malignant rhabdoid tumor (MRT))  
 RCC, 253  
 rhabdomyosarcoma and sarcoma botryoides, 253–255  
 Wilms tumor (*see* Wilms tumor)
- Inherited cystic kidney diseases**  
 GCKD, 270  
 JN, 269–270
- Innate immunity, UTI, 188**
- Intracellular fluid (ICF)**  
 fetal development, 79  
 osmolality, 80
- Intravaginal spermatic cord torsion, 233**
- Intrinsic AKI**  
 causes, newborn, 295  
 hypoxic-ischemic and nephrotoxin-related, 295  
 ischemic, 296  
 nephrotoxic, 296–297  
 pathophysiologic pathways, 295  
 renal vein thrombosis, 295
- Ischemia/reperfusion injury, 296**
- Ischemic AKI, 296**
- Isolated/familial diffuse cystic dysplasia**  
 ARPKD and ADPKD, 271  
 histologic confirmation, diagnosis, 270–271  
*HNF-1B/TCF2* gene, 271  
 “hypoplastic” GCKD, 271
- J**
- JN.** *See* Juvenile nephronophthisis (JN)
- Juvenile nephronophthisis (JN)**  
 clinical signs and symptoms, 270  
 description, 269  
 diagnosis, 270  
 “infantile” presentation, 269–270  
 treatment, 270
- K**
- Kallikrein-Kinin system, 64**
- Kayexalate®, 132**
- L**
- Leukocyte esterase, 191**
- Liddle syndrome, 354**
- Li-Fraumeni syndrome, 246**
- Lower urinary tract obstruction (LUTO), 141**
- LUTO.** *See* Lower urinary tract obstruction (LUTO)
- M**
- Machines, CRRT**  
 catabolic stem cell transplant patients, 314  
 CVVH and CVVHD modes, 314  
 FDA, 314  
 inborn errors, metabolism, 315
- MAG3.** *See* Mercaptoacetyltriglycine (MAG3)
- Magnetic resonance imaging (MRI)**  
 fetal, 151, 155  
 fluid intensity, 153  
 renal ultrasound, 160
- Magnetic resonance urography (MRU), 195**
- Malignant rhabdoid tumor (MRT)**  
 aggressive clinical nature, 252  
 description, 251  
*hSNF/INI1* gene, 22q11.2 chromosome, 252  
 kidney and brain, 2-month-old infant, 252  
 treatment regimens, combined therapies, 252–253
- MCDK.** *See* Multicystic dysplastic kidney (MCDK)
- Medical management**  
 AKI  
   acid-base homeostasis, 300  
   complications, hypertonic saline administration, 299  
   description, 298  
   dialysis, 301–302  
   electrolyte abnormalities, 299



- etiology, 298–299
- fluid management, 299
- hyperkalemia, 300
- hyperphosphatemia, 300
- hypocalcemia, 300
- hyponatremia, 299
- medications, 301
- nutrition, 300
- therapeutics, 301
- urine outflow, 299
- CNS
  - albumin and diuretics, 282
  - antiproteinuric therapies, 282
  - immunoglobulin levels, 283
  - nutrition and growth deficits, 281–282
  - risk, hypothyroidism, 283
  - thyroxine dosing, 283
  - warfarin, 283
- PUVs
  - adequate bladder emptying, 206
  - Cincinnati Children’s Urology algorithm, 205–206
  - infants with ESRD, 205
  - poor bladder compliance, 205
  - positive bladder outcomes, 206
- Mercaptoacetyl triglycine (MAG3)
  - diuretic renogram, 158, 163–166
  - and DMSA, 152
  - nuclear medicine renogram, 175
  - renal scan, 159, 176
- Mesenchymal-to-epithelial transformation (MET)
  - beta-catenin, 28
  - cell-autonomous fashion, 28
  - epithelialization defects, 28
  - morphogenetic sequence, 29
  - nephron epithelial cell progenitors, 30
  - Notch receptor signaling, 29
  - podocyte terminal differentiation, 30
  - primary glomerulogenesis defect, 30
  - segmentation, nephron, 29
  - S-shaped stage, 31
  - tubular development, 30
  - VEGF, 31
- Mesoblastic nephroma
  - benign hamartomas, 247–248
  - clinical signs/symptoms, 248
  - histologic characteristics, renal tumors, 248
  - homogeneous rubbery fibroid-like character, 248
  - young infant, 248
- Metanephric mesenchyme (MM)
  - canonical Wnt signaling, 15
  - cell survival and cell-cell adhesion, 16
  - characteristics, 14
  - epithelial cell differentiation, 16
  - epithelial diverticulum, 4
  - induction, UB outgrowth, 8
  - mesenchymal cell fate, 14
  - multipotent epithelial progenitors, 14
  - nephron progenitor cells, 15
  - positive feedback mechanism, 14
  - reciprocal inductive interactions, 4
  - signaling peptides, 13
  - survival genes, 16
- MIS. *See* Mullerian inhibitory substance (MIS)
- Mitochondrial myopathies and mutations, LAMB3, 279
- MRI. *See* Magnetic resonance imaging (MRI)
- MRT. *See* Malignant rhabdoid tumor (MRT)
- MRU. *See* Magnetic resonance urography (MRU)
- <sup>99m</sup>Tc-DMSA. *See* Technetium-labeled dimercaptosuccinic acid (<sup>99m</sup>Tc-DMSA)
- Mullerian inhibitory substance (MIS)
  - initial laboratory evaluations, 236
  - internal genitalia and gonadal differentiation, 235
  - and testosterone, 233–234
- Multicystic dysplastic kidney (MCDK)
  - contralateral kidneys, 272
  - cystic dysplasia, 271
  - developmental genes, 271
  - diagnosis, 272
  - hypertension, 175
  - nephrectomy, 172
  - renal dysplasia, 168
  - and VUR, 272
- Myelomeningocele
  - assessment, 223
  - hydrocephalus, 223
  - management, 223–224
  - mesoderm in-growth, spinal cord, 222–223
  - neural injury and lower urinary tract dysfunction, 223
  - neuropathic bladder dysfunction, children, 222
  - spinal deformity, 223
- N**
- Nail-patella syndrome, 279
- NAPRTCS. *See* North American Pediatric Renal Transplant Collaborative Studies (NAPRTCS)
- NEC. *See* Necrotizing enterocolitis (NEC)
- Necrotizing enterocolitis (NEC), 203
- Neonatal AKI
  - conservative care, 136–137
  - dialysis, 137
- Neonatal hypertension
  - blood pressure measurement (*see* Blood pressure measurement, neonates)
  - case vignette, 349
  - with chronic lung disease, 359
  - de novo development, 359
  - diagnostic evaluation
    - catecholamines/endocrinopathies, 357
    - chest X-ray determination, 356
    - etiologies, 353–355
    - evaluation, medical history, 355
    - laboratory and radiologic testing, 356
    - noninvasive angiography, 357
    - pertinent physical exam findings, 356
    - plasma renin activity levels, 357
    - serum electrolytes, BUN and creatinine, 356
    - symptoms, 355

- Neonatal hypertension (*cont.*)  
 etiologies (*see* Etiologies, neonatal hypertension)  
 morbidity and mortality, 359  
 neonatologists and pediatric nephrologists, 350  
 prevalence, 352  
 short-term NICU follow-up, 359  
 treatment  
   ACE inhibitors, 358–359  
   antihypertensive effects, 357–359  
   ARPKD, 359  
   beta-blockers, 359  
   diuretics, 357–358  
   end-organ injury, 357  
   reversible causes, blood pressure, 357
- Neonatal intensive care unit (NICU), 175
- Neonatal management, PUVs  
 description, 201  
 in infants with ESRD  
   care of patients, 202  
   dialysis, 203  
   fetal care centers, 202  
   genitourinary tract, 203  
 in infants with normal renal function  
   clinical significance, upper tract dilation, 202  
   endoscopic valve ablation, 202  
   measurement, SCr, 202  
   progression to ESRD, 202  
   renal ultrasound, 201–202  
   treatment, 201
- Neonatal obstructive uropathy  
 case vignette, 211–212  
 description, 212  
 ESRD, newborns and children, 212  
 history  
   associated ureteral dilation, 213  
   degree of dilation, 213  
   hydronephrotic kidneys, 213  
   oligohydramnios, 214  
   prenatal ultrasonography, 213  
   sonographic appearance, bladder, 213  
   ureterocele identification, 214  
 imaging  
   cystic dysplasia, 214  
   echogenic debris, 215  
   nuclear imaging renogram, 215  
   percutaneous antegrade pyeloureterography, 215–216  
   radionucleotide tracers, 215  
   renal ultrasound, 214  
   VCUG, 215  
 physical examination, 214  
 SFU, grading system, 213  
 treatment  
   hydronephrosis (*see* Hydronephrosis)  
   hydroureteronephrosis (*see* Hydroureteronephrosis)  
   radiographic finding, 216  
   unilateral ureteral obstruction, 216  
 UPJ and UVJ, 213
- Neonatal UTI. *See* Urinary tract infection (UTI)
- Neonates, cystic kidney disease  
 enlarged echogenic kidneys, 264, 265  
 palpable abdominal masses, 264  
 Potter's facies, 264, 265  
 renal macrocysts, 264, 266
- Neoplasms, imaging  
 axial T2 and coronal T1, 178, 180  
 chromosomal analysis, 177  
 coronal MR images, 179, 181  
 echogenicity mass, 178, 179, 181  
 mesoblastic nephroma, 177, 179  
 MRI, 178  
 nephroblastomatosis, 177–180  
 tumor, 178, 180
- Nephric duct morphogenesis  
 formation, nephrogenic primordia, 4, 5  
 kidney and ureter morphogenesis, 4  
 mesoderm-derived ancestors, 4  
 mesonephric degeneration, 4  
 molecular pathways  
   BMP4, 5  
     genetic studies, mice, 5  
   Hox genes, 6  
   *Osr1*, 6  
     urinary tract development, 6  
   urogenital sinus, 6–7
- Nephrin (NPHS1) mutation  
 components, glomerular filtration barrier, 277, 278  
 description, 278  
 Fin-major and Fin-minor, 278  
 massive proteinuria, 278
- Nephron development  
 actin-based cytoplasmic extensions, 27  
 angiogenesis, 27  
 cessation, nephrogenesis, 31  
 endothelial and mesangial cells, 26  
 epithelialization, 25  
 first functioning nephrons, 24  
 glomerular filtration barrier, 28  
 glomerulogenesis, 26  
 Henle loop, 25  
 hypertension and renal disease, 24  
 and MET, 28–31  
 nephron segmentation, 25  
 pre-tubular aggregation, 24  
 stereotypic sequence, 24, 25  
 tubular domains and glomerular, 25
- Nephrotoxic AKI  
 acyclovir, 297  
 amphotericin B, 296  
 gentamicin, 296–297  
 indomethacin, 296  
 medications, 296  
 NSAIDs/ACE inhibitors, 297  
 pharmacologic agents, 296
- Neuropathic bladder  
 myelomeningocele (*see* Myelomeningocele)  
 sacral agenesis, 224  
 spina bifida occulta, 224

- NICU. *See* Neonatal intensive care unit (NICU)
- Nissen fundoplication, 134
- Nondilated bladder anomalies, 221
- Nonsteroidal anti-inflammatory drugs (NSAIDs)
- indomethacin and ACE I, 295
  - nephrotoxic medications, 296
  - tocolysis, 297
- Normal sexual differentiation
- gonads and external genitalia, 233
  - MIS and testosterone, 233–234
- North American Pediatric Renal Transplant Collaborative Studies (NAPRTCS)
- chronic PD, 325
  - description, 308
  - height SDS scores, 329
  - Tenckhoff catheters, 329–330
- Novel biomarkers, AKI, 302
- NPHS1. *See* Nephhrin (NPHS1) mutation
- NPHS2. *See* Podocin gene (NPHS2) mutations
- NSAIDs. *See* Nonsteroidal anti-inflammatory drugs (NSAIDs)
- Nutritional intake and growth, GFR
- average energy requirements, infancy, 129, 131
  - common neonatal and infant formula, 129, 130
  - dialysis progression, 129
  - human milk, 129
  - modular additions, formulas and diets, 129, 131
  - peritoneal dialysis, 131
- Nutrition, newborn with renal disease
- case vignette, 127–128
  - etiology, CKD in infancy, 129
  - GFR (*see* Glomerular filtration rate (GFR))
  - growth velocity, 128–129
  - linear growth rates, 128
  - maintenance and growth, 128
  - NAPRTCS, 128
  - neonatal AKI (*see* Neonatal AKI)
  - overlapping phases, 128
  - SDS, 128
- O**
- Ovotesticular DSD, 235
- P**
- PD. *See* Peritoneal dialysis (PD)
- Percutaneous shunting in lower urinary tract obstruction (PLUTO), 148
- Peritoneal dialysis (PD)
- AKI (*see* Acute kidney injury (AKI))
  - dialysis, children, 343
  - ESRD (*see* End-stage renal disease (ESRD))
  - fluid removal, 322
  - and HD, 342
  - PEG, children, 343
  - postoperative recovery, 343
  - treatment modality, neonates, 322
- Phospholipase C epsilon 1 (PLCE1), 279
- Physical examination, AKI
- cumulative fluid balance, 297–298
  - dehydration, 297
  - infants phototherapy, 298
  - urine output, 298
  - vital signs, 297
- Pierson syndrome, 279
- PLCE1. *See* Phospholipase C epsilon 1 (PLCE1)
- PLUTO. *See* Percutaneous shunting in lower urinary tract obstruction (PLUTO)
- Podocin gene (NPHS2) mutations
- components, glomerular filtration barrier, 277, 279
  - description, 278
  - lipid raft-associated protein, 279
  - microscopic findings, 279
- Polycystic kidney diseases
- ADPKD (*see* Autosomal dominant polycystic kidney disease (ADPKD))
  - ARPKD (*see* Autosomal recessive polycystic kidney disease (ARPKD))
  - description, 266
- Posterior urethral valves (PUVs)
- and (*see* Renal replacement therapy (RRT))
  - anatomy, 200–201
  - bladder compliance, 204
  - bladder cycling and upper tracts stabilization, 201
  - fetal surgery, 161
  - hydronephrosis and hydroureter, 162, 171
  - medical management (*see* Medical management, PUVs)
  - neonatal ESRD management, 200
  - neonatal management (*see* Neonatal management, PUVs)
  - noncompliant bladder, 205, 206
  - obstructive uropathy, males, 200
  - paradoxical situation, newborn, 344
  - prenatal imaging, 161
  - prenatal ultrasound, 200
  - renal dysplasia, 200
  - renal parenchyma, 162, 171
  - renal transplantation, 201
  - sagittal T2-weighted, 162, 171
  - structural and functional abnormalities, GU tract, 204
  - surgical management (*see* Surgical management, PUVs)
  - transverse and longitudinal US, 161, 169
  - uropathic ERF population, 344
  - and VCUG, 161, 162, 170
  - VUR and bladder diverticula, 204
  - 24-year-old G2P0A1 patient, 199
- Potassium
- fetus, 88
  - homeostasis, hyponatremia
    - chronic perturbations, 109
    - extracellular potassium concentration, 109
    - gastrointestinal tract, 109
    - urinary excretion, 109

- Potassium (*cont.*)  
 neonate  
   antenatal Bartter syndrome, 89  
   basal physiological flow rate, 89  
   CCD, 89  
   developmental changes, 89, 90  
   loss of function genetic mutations, 90  
   non-oliguric hyperkalemia, 88  
   postnatal growth, 88, 89  
   TALH, 89
- Potter's facies, 264, 265
- Predisposing host factors, UTI  
   innate immunity, 188  
   VUR (*see* Vesicoureteral reflux (VUR))
- Prerenal azotemia  
   causes, 295  
   in newborns, 293  
   NSAIDs, 295  
   renal blood flow, 293
- Pretransplant urodynamics  
   dry augment, 345  
   native nephrectomy, 345  
   non-invasive studies, 344  
   PUV, 344  
   safe and unsafe, 344  
   VUR, 345
- Prophylactic antibiotic therapy, UTI, 193
- Protein, GFR  
   energy requirements, 131  
   macronutrient intakes, normal infants, 131, 132  
   nitrogen balance, 131
- Proteinuria  
   albumin, 119  
   glomerular, 119  
   tubular, 119
- Pseudohyperkalemia, 112
- Pseudohypoaldosteronism type 1 (PHA-1), 112
- PUVs. *See* Posterior urethral valves (PUVs)
- R**
- Radionuclide renography  
   DMSA, 152  
   MAG3, 152
- Radionuclide scintigraphy, 195
- RBUS. *See* Renal-bladder ultrasound (RBUS)
- RCCs. *See* Renal cell carcinomas (RCCs)
- RCD. *See* Renal cystic disease (RCD)
- Recombinant human growth hormone (rhGH), 343
- Renal-bladder ultrasound (RBUS)  
   description, 186  
   detection, anatomic abnormalities, 194  
   renal parenchyma, 194  
   ureterocele, 194  
   and VCUG, 192
- Renal cell carcinomas (RCCs)  
   cancer predisposition syndromes, 253  
   chemotherapy and radiotherapy, 253  
   infancy and childhood, 253  
   palpable abdominal masses, 253
- Renal cystic disease (RCD)  
   ciliopathies, 166  
   cystogram, 172  
   dysplasia, 168–169  
   echogenicity, 167, 173  
   etiologies, 164  
   ipsilateral genitourinary anomalies, 170–171  
   longitudinal US images, 167, 173  
   MAG3 scan, 172, 176  
   MCDK, 170  
   metanephric blastema, 170  
   nongenetic causes, 166  
   renal fossa, 171  
   unilateral and bilateral, 169–170  
   VCUG, 172, 176
- Renal ectopia  
   AP radiograph, 157, 161  
   contralateral metanephric blastema, 156  
   fetus development, 156  
   horseshoe kidney, 156  
   pancake kidney, 157, 162  
   pelvic kidney, 157, 161  
   renal fossa, 156, 160  
   retroperitoneum, 156
- Renal function  
   biomarkers, 124–125  
   case vignette, 117–118  
   fetal (*see* Fetal renal function)  
   GFR (*see* Glomerular filtration rate (GFR))  
   proteinuria, 119  
   tubular function, 122–123  
   UA (*see* Urine analysis (UA))
- Renal macrocysts, 264, 266
- Renal replacement modality  
   AKI  
     dialysis selection, 324  
     metabolic toxins, 323  
     patients with cardiovascular instability, 323  
     PD and CRRT, 323  
     pediatric nephrologists, 323  
     relative contraindications, PD, 323
- ESRD  
   chronic PD, 329  
   infectious and mechanical complications,  
     HD, 329  
   long-term HD access, neonate, 329  
   NAPRTCS data, 329
- Renal replacement therapy (RRT)  
   comorbidity, 342  
   and ERF, 342  
   PUVs  
     acidosis, stomach, 203  
     catheter placement, PUVs, 204, 205  
     gastrocystoplasty vessels, greater curvature, 204  
     G-tube placement, 203  
     infant hemodialysis patients, 203  
     mitrofanoff and ureter, 203  
     NEC, 203  
     neonatal time period growth, 203  
     peritoneal dialysis, 203

- Renal tubular function  
 aldosterone, 66  
 ANF, 67  
 prostaglandins, 67  
 renin-angiotensin system, 66–67
- Renal vein thrombosis (RVT), 175
- Rhabdomyosarcoma  
 alveolar histology, 254–255  
 description, 254  
 embryonal tumors, 254  
 genitourinary system, 253  
 head/neck region, 253  
 mesenchymal progenitors, 253  
 postsurgical adjuvant chemotherapy, 255
- rhGH. *See* Recombinant human growth hormone (rhGH)
- RRT. *See* Renal replacement therapy (RRT)
- RVT. *See* Renal vein thrombosis (RVT)
- S**
- Sacral agenesis  
 defined, 224  
 neurologic deficit, 224  
 vertebral deformity, 224
- Sarcoma botryoides  
 description, 254  
 genitourinary rhabdomyosarcoma, 253  
 hematuria, 254
- SDS. *See* Standard deviation score (SDS)
- Sequential tubular segments  
 distal and collecting duct, 87–88  
 Henle loop, 87  
 proximal convoluted  
 cellular pathway, 87  
 electroneutral Na<sup>+</sup>/H<sup>+</sup> exchanger, 86  
 mRNA/protein expression, 86  
 sodium and potassium transport, 85, 86
- SFU. *See* Society of fetal urology (SFU)
- SIADH. *See* Syndrome of inappropriate antidiuretic hormone production (SIADH)
- Small molecule mediators, renal blood flow in utero  
 ANF, 64–65  
 endothelin, 65  
 Kallikrein-Kinin system, 64  
 nitric oxide, 64  
 prostaglandins, 64
- SNS. *See* Sympathetic nervous system (SNS)
- Society of fetal urology (SFU)  
 hydronephrosis grading system, 213  
 neonatal hydronephrosis and renal parenchyma, 214  
 “well-tempered” renogram, 215
- Sodium physiology  
 fetus, 83, 84  
 neonate  
 ANP, 84  
 developmental changes, transport mechanisms, 84, 85  
 gestational age, 83  
 physiologic natriuresis, 83–84  
 positive balance, 83  
 renal epithelial cells, 85  
 thyroid hormone, 85  
 renal tubules, 85–88
- Spina bifida occulta, 224
- Standard deviation score (SDS)  
 infants with CKD, 129  
 newborn length measurements, 128  
 supplemental enteral feeding, 134
- Stimulatory renal branching  
 cell movements, 20  
 FGF family members, 20  
 GDNF-RET signaling, 19  
 HGF, 20  
 high-resolution imaging systems, 20  
 inhibitory pathways  
 ALKs, 21  
 bone morphogenetic proteins, 21  
 branch formation, 20–21  
 dysplastic kidneys, 21  
 embryonic mice, 21  
 Gli activator, 22  
 renal hypoplasia, 22  
 Sonic Hedgehog signaling, 21  
 sprouty genes, 19  
 stimulatory and inhibitory signals, 19, 20  
 UB cell proliferation, 19
- Suprapubic aspiration  
 bladder, 189  
 minor complications, 190  
 reported success rates, 190
- Surgical management  
 CNS  
 cyclophosphamide and plasmapheresis, 284  
 peritoneal dialysis, 283  
 recurrence, nephrotic syndrome, 283  
 renal transplantation, 283  
 uni- and bilateral nephrectomies, 283
- PUVs  
 bladder appearance, VCUG before transplant, 207  
 CIC and anticholinergics, 207  
 dilating VUR, 206  
 DTPA and UPJO, 207  
 ipsilateral native nephrectomy and ureterectomy, 207  
 “pop-off” mechanism, 207  
 reimplantation, ureters, 206  
 urine output and upper tract function, 208
- Sympathetic nervous system (SNS)  
 α<sub>2</sub>-adrenoceptors, 63–64  
 β<sub>2</sub>-adrenoceptors, 63–64  
 catecholamines, 62  
 fetal adaption, 62–63  
 fetal renal vasculature, 63  
 renal stimulation, 63
- Syndrome of inappropriate antidiuretic hormone production (SIADH)  
 adrenal insufficiency, 103  
 causes, 102, 103  
 chemotherapeutic drugs, 103  
 fluid restriction, 103  
 treatment, vasopressin 2 antagonists, 103

**T**

- TBW. *See* Total body water (TBW)
- Technetium-labeled dimercaptosuccinic acid ( $^{99m}\text{Tc}$ -DMSA), 195
- Testicular torsion  
 “acute scrotum”, 232–233  
 etiology and assessment, 233  
 extravaginal torsion, 233  
 intravaginal spermatic cord torsion, 233  
 management, 233
- Testis and scrotum abnormalities  
 cryptorchidism (*see* Cryptorchidism)  
 hernias (*see* Hernias)  
 hydroceles (*see* Hydroceles)  
 testicular torsion (*see* Testicular torsion)  
 varicoceles (*see* Varicoceles)
- Thick ascending limb loop of Henle (TALH)  
 apical surface, 89  
 $\text{K}^+$  reabsorption, 89  
 reabsorptive capacity, 89
- Total body water (TBW), 78, 79
- Townes-Brock syndrome, 15
- Transplantations, newborn  
 blood vessel imaging, 344  
 case vignette, 341  
 CKD, 343  
 complications, 345  
 description, 345  
 dialysis access (*see* Dialysis)  
 ERF (*see* Established renal failure (ERF))  
 graft survival post-transplant, 345  
 pretransplant management, urinary tract (*see* Pretransplant urodynamics)  
 vascular thrombosis (*see* Vascular thrombosis)
- Trans-tubular potassium gradient (TTKG), 122–123
- Transurethral bladder catheterization, 190
- TRP. *See* Tubular reabsorption of phosphorus (TRP)
- TTKG. *See* Trans-tubular potassium gradient (TTKG)
- Tubular function  
 FeMg, 122  
 FeNa, 122  
 TRP, 123  
 TTKG, 122–123  
 UCa/Cr, 123
- Tubular lesions, GFR, 136
- Tubular proteinuria, 119
- Tubular reabsorption of phosphorus (TRP), 123

**U**

- UA. *See* Urine analysis (UA)
- UCa/Cr. *See* Urinary calcium to creatinine (UCa/Cr) ratio
- Ultrasound (US). *See* Urological imaging
- Undescended testes (UDT). *See* Cryptorchidism
- UPJO. *See* Ureteropelvic junction obstruction (UPJO)
- Urachal anomalies  
 bladder drainage, 222  
 cyst, 222  
 infection/abscess, organisms, 222  
 ultrasound, 222  
 umbilical-urachal sinus, 222  
 vesicourachal diverticulum, 222
- Ureter development  
 Cajal-like cells, 34  
 differentiated smooth muscle cells, 33  
 HCN3, 34  
 Hedgehog signaling, 34  
 molecular control, 34–35  
 normal and abnormal ureter morphogenesis, 32  
 Pallister-Hall syndrome, 34  
 smooth muscle cell differentiation, 33  
 stages, 32, 33  
 structure, 31, 32  
 UPJ, 33  
 urine flow, 33
- Ureteric bud (UB) outgrowth  
 cellular basis, 8  
 cellular rearrangements, 8  
 kidney development, 8  
 negative control  
*Bmp4*, 13  
*Grem1*-deficient mouse embryos, 14  
 inhibitory molecular mechanisms, 13  
 kidney organ culture studies, 13–14  
 ligand-receptor interactions, 12  
 renal agenesis, 13  
 sprouty proteins, 12  
 ureteric bud formation and branching, 11–12
- positive control  
 cell lineage, 11  
 DNA-binding properties, 10  
*Etv4* and *Etv5*, gene network, 9  
 Gata-binding protein 3 (*Gata3*), 10  
 GDNF-Ret signaling, 8, 10  
 gene mutations, 9  
 mouse models, renal agenesis, 9, 11  
 nephric duct extension, 10  
 normal kidney development, 9  
 promoting/suppressing signals, 9, 10  
 RTK signaling, 9, 11  
 stages, 11, 12  
 Wnt-mediated signals, 10
- Ureteropelvic junction obstruction (UPJO)  
 bladder decompression, 207  
 diuretic renography, 158–159  
 MAG3 renogram, 158, 165  
 management, kidney, 217  
 megaureters, 213  
 pelvocaliectasis, 158  
 percutaneous nephrostogram, 212  
 and VUR, 158
- Ureterovesical junction (UVJ)  
 distal ureters, 159  
 longitudinal US, 159, 166  
 MAG3 diuretic renogram, 159, 166  
 obstruction, 213
- Urethra imaging  
 AP projection, 155, 157  
 perineal sonogram, 154, 156

- US, 154
- VCUG, 154, 156
- verumontanum, 155, 157
- Urinalysis, 191
- Urinary calcium to creatinine (UCa/Cr) ratio, 123
- Urinary tract infection (UTI)
  - AAP guidelines, 186
  - antibiotic therapy, 192–193
  - bacterial illnesses, febrile young infants, 186
  - blood culture and spinal tap, 192
  - case vignette, patient
    - <sup>99m</sup>Tc-DMSA scan, 183, 185
    - renal ultrasound, 183, 184
    - VCUG, 183, 184
  - circumcision, 193–194
  - color doppler US, 172, 178
  - cortical scintigraphy, 172, 177
  - and CT, 173
  - cystosonography, 175
  - diagnosis, specimen culture, 189
  - DMSA renal scan, 172
  - epidemiology, 187
  - focal bacterial nephritis, 188–189
  - imaging studies
    - MRU, 195
    - radionuclide scintigraphy, 195
    - ultrasound, RBUS, 194
    - VCUG (*see* Voiding cystourethrogram (VCUG))
  - laboratory tests, 192
  - microbiology, 188
  - pathophysiology
    - agent virulence, 188
    - biological susceptibility, 187
    - mechanisms, infection to upper tract, 187
    - predisposing host factors (*see* Predisposing host factors, UTI)
  - posteromedial cortex, 172, 178
  - prognosis
    - risk of recurrence, 195
    - scarring and CKD, 195–196
  - prophylactic antibiotic therapy, 193
  - pyelonephritis, 172, 178
  - and RBUS, 186
  - renal pelvis, 172
  - “significant” bacteriuria, urine culture, 191–192
  - symptoms, young infants, 188
  - urinalysis, 191
  - urine collection methods (*see* Urine collection methods)
  - urine interleukin-1 $\beta$  and serum procalcitonin, 192
  - and VCUG, 174, 186
  - and VUR, 174–175, 186
- Urinary tract malformation syndromes
  - autosomal dominant
    - inheritance patterns, 44
    - polycystic kidney disease, 42–43
  - gene discovery, humans, 43
  - gene mutations, 43
  - genetic testing, 44
  - GWAS, 43
  - inheritance, 42
  - syndromic malformations, 42
  - web resources, genetic associations with human disease, 43
- Urinary tract malignancies
  - adults vs. children, 241, 242
  - cancer among infants
    - diagnosis, 241, 244, 245
    - infant ALL, 241
    - neuroblastoma, 241, 245
    - N-myc* oncogene, 245
    - pediatric cancers and renal tumors, 241, 243
    - radiologic and pathologic diagnostic interpretations, 245
    - renal tumors, 245
    - retinoblastoma, fibrosarcoma and hepatoblastoma, 241
    - solid tumors, 241, 245
  - cancer syndromes (*see* Cancer syndromes)
  - case vignette
    - baby’s pediatrician, 239
    - NWTSG, 240
    - pathologic examination, 240
    - Wilms tumor, 3-month-old male, 239, 240
  - clinical presentation, in infants
    - abdominal tumors, 245
    - BUN and creatinine, 246
    - implications, 246
    - magnetic resonance imaging, 245
    - prenatal detection, renal mass, 245, 246
  - disorders, 240
  - function, 240
  - tumors, infant urinary system (*see* Infant urinary system)
- Urine analysis (UA)
  - blood cells, 119
  - components types, 118
  - crystals, 119
  - description, 118
  - hematuria, 119
  - ketone bodies, 118
  - macroscopic assessment, 118
  - nitrites, 118
  - pH and glucose, 118
  - reagent strip, 118
- Urine collection methods
  - clean voided bag specimens, 190–191
  - description, 189
  - suprapubic aspiration, 189–190
  - transurethral bladder catheterization, 190
- Urological imaging
  - abnormalities, 151
  - bladder, 153–154
  - congenital anomalies (*see* Congenital anomalies)
  - cystography, 152
  - fetal imaging, 155–156, 158
  - hypertension
    - chronic lung disease, 175
    - diagnosis, 176
    - echogenicity, 176, 179

Urological imaging (*cont.*)

- neonatal, 175
- NICU, 175
- and refractory, 176, 179
- RVT, 175
- thrombosis, 176, 179
- kidney, 153, 154
- MRI, 151
- MR urography, 153
- neoplasms (*see* Neoplasms, imaging)
- radionuclide renography, 152
- urethra, 154–155
- US, 151
- UTI, 172–175

UTI. *See* Urinary tract infection (UTI)

UVJ. *See* Ureterovesical junction (UVJ)

**V**

## Varicoceles

- assessment and classification, 232
- associated pathology, 232
- etiology, 232
- management, 232

## Vascular access

- catheter tips, 309–310
- CRRT patients, 309
- dialysis, 309
- femoral catheters, 310
- 5-French catheters, 310
- hemodialysis catheters, neonates, 310
- impair proper blood flow, 311
- single-lumen access, 310
- TPA, 310
- uncuffed catheters, 310

## Vascular thrombosis

- pediatric recipients, 345–346
- recipient risk factors, 346

VCUG. *See* Voiding cystourethrogram (VCUG)

## Vesicoureteral reflux (VUR)

- acquired renal injury, 187
- cystogram, 174
- distal ureters, 151
- and ERF, 342
- hydronephrosis, 158
- intrarenal reflux, 163
- longitudinal US, 158, 162
- MAG3 diuretic renogram, 158, 163, 164
- mannose resistance, P-fimbriae, 188
- prophylactic antibiotics, patients, 193
- pyelonephritis, 187
- recurrent infection, stones and high-grade, 345
- renal damage, children, 187–188
- renal dysplasia, 169
- and UTI, 345
- and UVJ, 152
- and VCUG, 158, 163
- VCUG, patient, 183, 184

## Voiding cystourethrogram (VCUG)

- abnormal axis, 156, 160

- and abnormal RBUS, 192
  - bladder ears, 154, 155
  - febrile illness, 194–195
  - high-grade (IV–V) V VUR, 194
  - HUN, 158, 163
  - patient, 183, 184
  - posterior urethra, 161, 170
  - prevention, UTI recurrence, 194
  - prophylactic antimicrobial therapy, 194
  - PUV, 161, 170
  - splenic flexure, 176
  - and VUR, 158, 174
- VUR. *See* Vesicoureteral reflux (VUR)

**W**

## Water balance

- in fetus, 80–81
- in newborn, 81–82

## Wilms tumor

- age distribution, 249
- defined, bilateral, 250
- Denys-Drash syndrome, 15
- favorable histology, 250
- malignant kidney tumor, children, 248
- medical attention, 249
- “nephrogenic rests”, 249
- pediatric-inherited cancer syndromes, 249
- staging, 250
- surgery and chemotherapy, 250
- therapy, 249–250
- treatment, stage V, 250–251
- unfavorable histology, 250
- vincristine and dactinomycin, 250

Wilms tumor suppressor gene (WT1), 279

WT1. *See* Wilms tumor suppressor gene (WT1)

**X**

## 46XX DSD

- ACTH production, 234
- congenital adrenal hyperplasia, 234
- deficiency, estrogen synthetase, 234
- enzyme deficiency, congenital adrenal hyperplasia, 234, 235
- prenatal diagnosis, 234

## 46XY DSD

- androgen insensitivity syndrome, 235
- DHT deficiency, 234–235
- male infants, 234
- testosterone conversion, 234

**Y**

Y-connector, single-lumen access, 310

**Z**

Zellweger syndrome, 2