

Ahmed H. Al-Salem

An Illustrated Guide to Pediatric Urology

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Ahmed H. Al-Salem
Pediatric Surgery
AlSadik Hospital
Qatif
Saudi Arabia

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Preface

The field of pediatric urology is rapidly growing and currently it is considered as a subspecialty. Pediatric urologists and pediatric surgeons care for newborns, infants, and children with congenital and acquired urological conditions. I have written this book with more than 20 years' experience in the care of infants and children with urological conditions and hope it will help all those involved in the surgical care of infants and children with urological problems. This book is written in a simple way and easy to read. It covers most areas in the field of pediatric urology with emphasis on the most important points relevant to the patient's presentation, diagnosis, and management. This book is well illustrated and includes clinical, operative, radiological, and hand-drawn illustrations. I hope it will be useful to consultant pediatric urologists, pediatric surgeons, specialists, fellows, and residents. This book should be useful also to general practitioners, general surgeons, accident and emergency doctors, pediatricians, neonatologists, trainees, medical students, and nurses.

Qatif, Saudi Arabia

Ahmed H. Al-Salem

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Contents

1	Congenital Urological Malformations	1
1.1	Introduction	1
1.2	Normal Embryology	2
1.3	Abnormalities of the Kidney	4
1.3.1	Renal Agenesis.	5
1.3.2	Renal Hypoplasia.	6
1.3.3	Supernumerary Kidneys.	7
1.3.4	Renal Dysplasia and Multicystic Kidney.	8
1.3.5	Polycystic Kidney Disease.	8
1.3.6	Simple (Solitary) Renal Cyst.	11
1.3.7	Renal Fusion and Renal Ectopia	12
1.3.8	Horseshoe Kidney	12
1.3.9	Crossed Fused Renal Ectopia.	15
1.3.10	Ectopic Kidney.	17
1.4	Abnormalities of the Ureter	18
1.5	Abnormalities of the Bladder.	22
1.6	Abnormalities of the Penis and Urethra in Males	33
1.7	Abnormalities of Female External Genitalia	36
	Further Reading	41
2	Hydronephrosis in Infants and Children	43
2.1	Introduction	43
2.2	Pathophysiology.	45
2.3	Etiology of Hydronephrosis.	49
2.4	Classification of Hydronephrosis.	55
2.5	Clinical Features	57
2.6	Investigations and Diagnosis	58
2.7	Treatment	63
2.8	Antenatal Hydronephrosis	65
	Further Reading	69
3	Pelviureteric Junction (PUJ) Obstruction	71
3.1	Introduction	71
3.2	Embryology	73
3.3	Pathophysiology.	73
3.4	Etiology of PUJ Obstruction	74
3.5	Clinical Features	76
3.6	Diagnosis and Investigations	79

3.7	Management of Newborns with PUJ Obstruction	88
3.8	Treatment	89
3.9	Post-operative Complications and Follow-Up	97
	Further Reading	98
4	Renal Tumors in Children	101
4.1	Introduction	101
4.2	Wilms' Tumor	102
4.2.1	Introduction	102
4.2.2	Etiology	104
4.2.3	Histopathology	105
4.2.4	Nephroblastomatosis	108
4.2.5	Clinical Features	110
4.2.6	Risk Factors for Wilms' Tumor	112
4.2.7	Staging of Wilms Tumor	113
4.2.8	Investigations	114
4.2.9	Prognosis and Complications of Wilms Tumor	121
4.2.10	Surgical Considerations	123
4.2.11	Surgical Complications	130
4.2.12	Prognosis and Outcome	130
4.2.13	Extrarenal Wilms' Tumors	131
4.3	Mesoblastic Nephroma	132
4.3.1	Introduction	132
4.3.2	Classification	133
4.3.3	Epidemiology	134
4.3.4	Histopathology	134
4.3.5	Clinical Features	136
4.3.6	Investigations	137
4.3.7	Treatment and Prognosis	138
4.4	Clear Cell Sarcoma of the Kidney (CCSK)	138
4.4.1	Introduction	138
4.4.2	Pathophysiology	139
4.4.3	Clinical Features	140
4.4.4	Investigations	140
4.4.5	Histopathology	140
4.4.6	Treatment	141
4.4.7	Prognosis	142
4.5	Malignant Rhabdoid Tumor of the Kidney	143
4.5.1	Introduction	143
4.5.2	Etiology and Pathophysiology	144
4.5.3	Histologic Findings	144
4.5.4	Clinical Features	145
4.5.5	Investigations and Diagnosis	146
4.5.6	Treatment and Outcome	149
4.5.7	Mortality/Morbidity	150
4.6	Renal Cell Carcinoma in Children	150
4.6.1	Introduction	150
4.6.2	Histopathology	151

4.6.3	Classification	153
4.6.4	Staging	153
4.6.5	Clinical Features	154
4.6.6	Investigations	155
4.6.7	Management	155
4.6.8	Prognosis	156
4.7	Angiomyolipoma of the Kidney	156
4.7.1	Introduction	156
4.7.2	Histopathology	157
4.7.3	Classification	157
4.7.4	Clinical Features	158
4.7.5	Investigations	158
4.7.6	Treatment and Prognosis	159
4.8	Renal Lymphoma	159
4.8.1	Introduction	159
4.8.2	Etiology and Pathogenesis	159
4.8.3	Diagnosis	160
4.8.4	Clinical Features	161
4.8.5	Treatment and Prognosis	162
4.9	Ossifying Renal Tumor of Infancy	162
4.10	Metanephric Adenoma	163
4.10.1	Introduction	163
4.10.2	Histopathology	164
4.10.3	Diagnosis	164
4.10.4	Clinical Features	164
4.10.5	Treatment	165
4.11	Multilocular Cystic Renal Tumor	165
	Further Reading	168
5	Multi Cystic Dysplastic Kidney (MCDK)	173
5.1	Introduction	173
5.2	Embryology	174
5.3	Etiology and Pathophysiology	175
5.4	Histologic Findings	175
5.5	The Natural History of Multicystic Dysplastic Kidney	178
5.6	Classification	179
5.7	Associated Anomalies	180
5.8	Clinical Features	182
5.9	Investigations	182
5.10	Treatment	183
	Further Reading	185
6	Congenital Ureteral Anomalies	187
6.1	Etiology	187
6.2	Clinical Features	188
6.3	Investigations and Diagnosis	188
6.4	Duplex (Duplicated) System	189
6.4.1	Introduction	189
6.4.2	Classification	192

6.4.3	Clinical Features	193
6.4.4	Investigations	193
6.4.5	Treatment and Prognosis	195
6.5	Ectopic Ureter	197
6.5.1	Introduction	197
6.5.2	Embryology and Pathophysiology	198
6.5.3	Clinical Features	199
6.5.4	Diagnosis	200
6.5.5	Surgical Treatment	200
6.6	Ureterocele	202
6.6.1	Introduction	202
6.6.2	Classification	203
6.6.3	Clinical Features	204
6.6.4	Investigations and Diagnosis	204
6.6.5	Treatment	205
6.7	Vesicoureteral Reflux (VUR)	208
6.8	Mega Ureter	212
	Further Reading	215
7	Congenital Megaureter	217
7.1	Introduction	217
7.2	Classification	218
7.3	Etiology and Pathophysiology	223
7.4	Clinical Presentation	225
7.5	Investigations and Diagnosis	225
7.6	Treatment and Prognosis	231
7.7	Complications	234
	Further Reading	234
8	Vesicoureteral Reflux (VUR) in Children	237
8.1	Introduction	237
8.2	Pathophysiology	240
8.3	Classification of VUR	243
8.4	Etiology of VUR	247
8.5	Clinical Features	250
8.6	Investigations	254
8.7	Management	257
8.7.1	Medical Treatment of VUR	259
8.7.2	Antibiotics Used for Prophylaxis	261
8.7.3	Anticholinergics	261
8.7.4	Surveillance	262
8.8	Surgical Therapy of VUR	262
8.8.1	Indications for Surgical Interventions	262
8.8.2	Indications for Surgical Interventions Based on Age at Diagnosis and the Presence or Absence of Renal Lesions	262
8.8.3	Endoscopic Injection	263
8.8.4	Surgical Management	266
8.9	Mortality/Morbidity	268
	Further Reading	269

9	Pediatric Urolithiasis	271
9.1	Introduction	271
9.2	Etiology	272
9.3	Classification of Urolithiasis	278
9.4	Clinical Features	278
9.5	Investigations	280
9.6	Complications of Urolithiasis	282
9.7	Management	282
	Further Reading	285
10	Persistent Müllerian Duct Syndrome (PMDS)	287
10.1	Introduction	287
10.2	Embryology of Persistent Müllerian Duct Syndrome	287
10.3	Etiology and Inheritance of PMDS	290
10.4	Classification of PMDS	291
10.5	Clinical Features	291
10.6	Treatment	292
10.7	Prognosis	293
	Further Reading	293
11	Neurogenic Bladder Sphincter Dysfunction	295
11.1	Introduction	295
11.2	Physiology and Bladder Function	296
	11.2.1 Micturition	301
11.3	Pathophysiological Changes of NBSD	301
11.4	Etiology and Clinical Features	302
11.5	Investigations and Diagnosis	310
11.6	Classification of Neurogenic Bladder	313
11.7	Management	314
11.8	Clean Intermittent Catheterization	316
11.9	Anticholinergics	317
11.10	Botulinum Toxin Type A	318
11.11	Tricyclic Antidepressant Drugs	318
11.12	Surgical Management	320
	Further Reading	321
12	Urinary Tract Infection in Infants and Children	323
12.1	Introduction	323
12.2	Etiology	324
12.3	Pathophysiology	325
12.4	Clinical Features	325
12.5	Investigations and Diagnosis	327
12.6	Management	330
	Further Reading	334
13	Bladder Exstrophy-Epispadias Complex	337
13.1	Introduction	337
13.2	Embryology	341
13.3	Epispadias	342
	13.3.1 Introduction	342
	13.3.2 Etiology	343
	13.3.3 Classification	344

13.3.4	Treatment	345
13.3.5	Surgical Repair of Male Epispadias	347
13.3.6	Female Epispadias	348
13.3.7	Surgical Repair of Female Epispadias	349
13.3.8	Prognosis	349
13.4	Bladder Exstrophy	350
13.4.1	Introduction	350
13.4.2	Associated Anomalies	354
13.4.3	Principles of Surgical Management of Bladder Exstrophy	355
13.4.4	Evaluation and Management	356
13.5	Cloacal Exstrophy	360
13.5.1	Introduction	360
13.5.2	Skeletal Changes in Cloacal Exstrophy	363
13.5.3	Etiology and Pathogenesis	363
13.5.4	Prenatal Diagnosis	364
13.5.5	Associated Anomalies	364
13.5.6	Clinical Features and Management	366
13.5.7	Surgical Repair of Cloacal Exstrophy	368
13.5.8	Surgical Reconstruction	370
13.5.9	Management of Urinary Incontinence	371
13.5.10	Prognosis	371
13.5.11	Complications	372
	Further Reading	372
14	Megacystis Microcolon Intestinal Hypoperistalsis Syndrome (Berdon Syndrome)	373
14.1	Introduction	373
14.2	Etiology	374
14.3	Clinical Features	376
14.4	Associated Anomalies	376
14.5	Diagnosis	376
14.6	Treatment and Prognosis	377
	Further Reading	379
15	Cloacal Anomalies	381
15.1	Introduction	381
15.2	Associated Anomalies	383
15.3	Classification	384
15.4	Clinical Features	384
15.5	Investigations	386
15.6	Management	387
	Further Reading	391
16	Urachal Remnants	393
16.1	Introduction	393
16.2	Embryology	393
16.3	Classification	395

16.4	Clinical Features	396
16.5	Tumors and Urachal Remnants	398
16.6	Management	399
	Further Reading	400
17	Inguinal Hernias and Hydroceles	401
17.1	Introduction	401
17.2	Inguinal Hernia	401
17.2.1	Incidence	401
17.2.2	Etiology	402
17.2.3	Clinical Features	403
17.2.4	Variants of Hernia	404
17.2.5	Complications of Inguinal Hernias	406
17.2.6	Treatment	408
17.2.7	Complications of Inguinal Herniotomy	410
17.3	Hydrocele	410
17.3.1	Embryology	410
17.3.2	Classification of Hydroceles	411
17.3.3	Treatment	413
	Further Reading	414
18	Cloacal Exstrophy	415
18.1	Introduction	415
18.2	Etiology and Pathogenesis	415
18.3	Associated Anomalies	417
18.4	Clinical Features and Management	418
	Further Reading	421
19	Posterior Urethral Valve	423
19.1	Introduction	423
19.2	Embryology	424
19.3	Pathophysiology	425
19.4	Classification	426
19.5	Clinical Features	427
19.6	Investigations and Diagnosis	428
19.7	Management	433
19.8	Medications Used in Patients with PUV	437
19.9	Prognosis and Follow-Up	438
19.10	Long-Term Outcomes	439
19.10.1	Vesico-ureteric Reflux	440
19.10.2	Hydro-ureteronephrosis	440
19.10.3	Bladder Dysfunction	441
19.10.4	Renal Transplantation	441
19.10.5	Fertility	441
	Further Reading	442
20	Utricular Cyst (Prostatic Utricular Cyst)	443
20.1	Introduction	443
20.2	Embryology	445
20.3	Classification of Utricular Cysts	445

20.4	Clinical Features	446
20.5	Investigations	446
20.6	Treatment	447
20.7	The Müllerian Duct Cyst	448
	Further Reading	449
21	Hypospadias.	451
21.1	Introduction	451
21.2	Effects of Hypospadias	455
21.3	Embryology	455
21.4	Etiology of Hypospadias	456
21.5	Associated Anomalies	456
21.6	Classification of Hypospadias	458
21.7	Clinical Features of Hypospadias	462
21.8	Treatment	464
21.9	Urinary Diversion	472
21.10	Postoperative Complications	473
	Further Reading	476
22	Male Circumcision	477
22.1	Introduction	477
22.2	Anatomy and Pathophysiology	479
22.3	History of Circumcision	480
22.4	Pain Management	482
22.5	Indications for Circumcision	482
22.6	Contraindications to Circumcision	485
22.7	Surgical Procedure	486
22.8	Complications of Circumcision	492
	Further Reading	499
23	Priapism in Children	501
23.1	Introduction	501
23.2	Pathophysiology	503
23.3	Etiology	505
23.4	Classification of Priapism	507
23.5	Clinical Features	508
23.6	Investigations	509
23.7	Management	510
23.8	Prognosis	512
23.9	Priapism and Sickle Cell Disease	513
	23.9.1 Introduction	513
	23.9.2 Epidemiology	514
	23.9.3 Classification	514
	23.9.4 Pathophysiology	515
	23.9.5 Clinical Features	517
	23.9.6 Treatment	519
	23.9.7 Prevention of Stuttering Priapism	521
	23.9.8 Complications of Priapism and Prognosis	524
	Further Reading	524

24 Undescended Testes (Cryptorchidism)	527
24.1 Introduction	527
24.2 Embryology and Normal Testicular Development and Descent	530
24.3 Classification of Undescended Testes.	531
24.4 Causes of Undescended Testes and Risk Factors	531
24.5 Histopathology	532
24.6 Classification of Abnormal Testes	533
24.7 Clinical Features and Diagnosis	537
24.8 Treatment	538
24.8.1 Success of Surgical Treatment	541
24.9 Complications of Orchidopexy.	541
24.10 Infertility and Undescended Testes.	541
24.11 Undescended Testes and the Risk of Cancer	542
Further Reading	543
25 Varicocele	545
25.1 Introduction	545
25.2 Etiology	546
25.3 Pathophysiology	546
25.4 Grading of Varicoceles	547
25.5 Clinical Features.	548
25.6 Diagnosis	548
25.7 Treatment	549
25.8 Postoperative Complications	550
25.9 Prognosis	551
Further Reading	551
26 Testicular Torsion and Torsion of the Testicular or Epididymal Appendage	553
26.1 Introduction	553
26.2 Etiology and Risk Factors.	554
26.3 Diagnosis	555
26.4 Intermittent Testicular Torsion	556
26.5 Classification of Testicular Torsion	556
26.6 Effects of Testicular Torsion.	557
26.7 Clinical Features.	558
26.8 Treatment	560
26.9 Intra-uterine Torsion of Testes	562
26.9.1 Introduction	562
26.9.2 Etiology of Extravaginal Torsion	562
26.9.3 Clinical Features.	562
26.9.4 Treatment	563
26.10 Torsion of the Testicular or Epididymal Appendage	565
26.10.1 Introduction	565
26.10.2 Embryology	567
26.10.3 Clinical Features.	567
26.10.4 Investigations and Treatment	567
Further Reading	568

27	Testicular Tumors in Children	569
27.1	Introduction	569
27.2	Classification of Testicular Tumors	573
27.3	Histologic Classification of Seminomas	576
27.4	Etiology of Testicular Tumors	576
27.5	Clinical Features	579
27.6	Staging	580
	27.6.1 Regional Lymph Node Staging	581
27.7	Investigations	582
27.8	Treatment	583
27.9	Yolk Sac Tumor	585
27.10	Teratoma	586
27.11	Mixed Germ Cell Tumor	587
27.12	Stromal Tumors	587
27.13	Simple Testicular Cyst	589
27.14	Epidermoid Cysts	589
27.15	Testicular Microlithiasis (TM)	589
27.16	Gonadoblastoma	590
27.17	Cystic Dysplasia of the Testes	590
27.18	Leukemia and Lymphoma	590
27.19	Paratesticular Rhabdomyosarcoma	591
27.20	Prognosis and Outcome	592
	Further Reading	592
28	Splenogonadal Fusion	595
28.1	Introduction	595
28.2	Etiology	596
28.3	Classification	596
28.4	Associated Anomalies	597
28.5	Clinical Features	597
28.6	Investigations	598
28.7	Treatment	598
	Further Reading	599
29	Acute Scrotum	601
29.1	Introduction	601
29.2	Torsion of Testes	601
	29.2.1 Introduction	601
	29.2.2 Classification	602
	29.2.3 Etiology	603
	29.2.4 Clinical Features	604
	29.2.5 Effects of Torsion of Testes	605
	29.2.6 Investigations	606
	29.2.7 Treatment	606
29.3	Torsion of the Testicular or Epididymal Appendage	608
	29.3.1 Introduction	608
	29.3.2 Embryology	609
	29.3.3 Clinical Features	609
	29.3.4 Investigations and Treatment	609

29.4	Epididymitis, Orchitis, and Epididymo-orchitis.	610
29.4.1	Introduction	610
29.4.2	Etiology	610
29.4.3	Clinical Features.	611
29.4.4	Investigations and Treatment	611
29.5	Idiopathic Scrotal Edema	612
29.6	Testicular Trauma	613
29.7	Other Causes of Acute Scrotum	613
29.8	Splenogonadal Fusion	614
	Further Reading	617
30	Hydrocolpos, Vaginal Agenesis and Atresia	619
30.1	Introduction	619
30.2	Imperforate Hymen	619
30.3	Vaginal Atresia	621
30.4	Classification	621
30.5	Associated Anomalies	622
30.6	Embryology	622
30.7	Clinical Features.	623
30.8	Investigations	625
30.9	Management.	628
	Further Reading	633
31	Disorders of Sexual Development	635
31.1	Introduction	635
31.2	Embryology	639
31.3	Sexual and Gonadal Differentiation	642
31.4	Classification	645
31.5	Evaluation of a Newborn with DSD.	651
31.6	Diagnosis and Investigations	653
31.7	Management of Patients with DSD	656
31.8	Surgical Corrections of DSD	657
31.9	Congenital Adrenal Hyperplasia (CAH)	659
31.10	Androgen Insensitivity Syndrome (Testicular Feminization Syndrome)	665
31.11	Deficiency of MIS (Persistent Müllerian Duct Syndrome). . .	670
31.12	5-Alpha-Reductase Deficiency.	672
31.13	Gonadal Dysgenesis	673
31.14	Deficient Testosterone Biosynthesis.	678
31.15	Ovotestis Disorders of Sexual Development	679
31.16	Other Rare Disorders of Sexual Development	682
	Further Reading	685
	Index.	687

1.1 Introduction

- The urinary system is comprised of two kidneys, two ureters, a bladder and a urethra.
- The kidneys contains the nephrons which are responsible to filter the blood as it passes through the kidney.
- It is estimated that the kidneys will filter around 190 l of water every day from the blood. Most of the water from the blood that is filtered is reabsorbed into the body and the remaining water is excreted as urine.
- This water travels down the ureters to the bladder which acts as a storage area for the urine.
- When the bladder reaches a certain volume, nerves in the walls of the bladder are stimulated and urination happens.
- The kidneys also play an important role in red blood cell production. Erythropoietin, which is produced in the kidneys stimulates the production of the red blood cells.
- Congenital anomalies of the kidney and urinary tract (CAKUT) are common in children.
- They represent approximately 30 % of all prenatally diagnosed malformations.
- Congenital anomalies of the kidney and urinary tract occur in 3–6 per 1,000 live births.
- They account for the most cases of pediatric end-stage kidney disease (ESKD), and predispose an individual to hypertension and cardiovascular disease throughout life.
- They are responsible for 34–59 % of chronic kidney disease (CKD) and for 31 % of all cases of end-stage kidney disease (ESKD) in children.
- Congenital anomalies of the kidney and urinary tract comprise a wide range of structural and functional malformations that occur at the level of:
 - The kidney
 - Collecting system
 - Bladder
 - Urethra
- With improved prenatal screening, many cases of CAKUT are diagnosed by antenatal ultrasonography performed on 18–20 weeks of gestation.
- Most common antenatal manifestations of CAKUT include oligohydramnios or variations in gross morphology of the kidney, ureter, or bladder.
- CAKUTs anomalies can be:
 - Sporadic
 - Familial
 - Syndromic
 - Nonsyndromic
- Syndromic CAKUTs:
 - They develop in association with other additional congenital abnormalities outside of the kidney and urinary tract.
 - They manifest clinically recognizable features of a known syndrome.

- Nonsyndromic CAKUT:
 - Congenital structural anomalies confined only to the kidney and urinary tract.
- Genetic causes have been identified in association with the syndromic forms of congenital malformations.
- The spectrum of congenital anomalies includes more common anomalies such as vesicoureteral reflux and, rarely, more severe malformations such as bilateral renal agenesis.
- These congenital anomalies can be unilateral or bilateral, and sometimes different defects coexist in the same child.
- It is important to recognize and diagnose these anomalies early as early treatment will minimize renal damage, prevent or delay the onset of ESRD, and provide supportive care to avoid complications of ESRD.

1.2 Normal Embryology

- The two most common congenital bladder abnormalities are:
 - Bladder exstrophy
 - Congenital diverticula
- An exstrophic bladder is one that is open to the outside and turned inside out, so that its inside is visible at birth, protruding from the lower abdomen.
- A diverticulum is an extension of a hollow organ, usually shaped like a pouch with a narrow opening.
- The terminal portion of the hindgut is called the cloaca.
- The cloaca forms the future urinary and gastrointestinal tracts.
- The cloaca is formed early in fetal life by incorporation of the allantois to form a common distal channel for the primitive urinary and gastrointestinal systems.
- The most caudal portion of the cloaca is the cloacal membrane.
- This separates the cloaca from the amniotic cavity.
- During the fourth to seventh weeks of development, the cloaca is divided by the urorectal septum into anterior and posterior portions to form the urogenital sinus and anal canal, respectively.
- The urogenital sinus further differentiates into three anatomic components:
 - The vesical (cranial) portion
 - The pelvic (middle) portion
 - The phallic (caudal) portion
- The pelvic and phallic portions will form the urethra and genitals, respectively.
- The vesical portion forms most of the bladder and is continuous with the allantois.
- The allantois normally constricts to a thick fibrous cord, the median umbilical ligament, and extends from the apex of the bladder to the umbilicus.
- The trigone portion of the bladder is formed from the caudal ends of the mesonephric ducts, which are incorporated into the developing bladder wall.
- The ureters are formed from the ureteral buds.
- The ureteral bud is an outgrowth of the mesonephric duct near its entrance into the cloaca. This will elongate and develop into the ureter.
- The more distal ureteral bud undergoes a complex interaction with the primitive kidney to induce differentiation of the renal parenchyma and formation of the renal pelvis, calyces, and collecting tubules.
- As the kidneys develop and ascend, traction on the ureters causes the ureteral orifices to move superolaterally, resulting in an oblique course through the muscular wall at the base of the bladder.
- The kidneys are paired organs located retroperitoneally. Their vascular supply comes from the renal arteries, and they drain into the renal veins. Each kidney excretes into a ureter, which will in turn empty into the urinary bladder. Its functional unit is the nephron.
- The urogenital system arises from intermediate mesoderm which forms a urogenital ridge on either side of the aorta.
- The urogenital ridge develops into three sets of tubular nephric structures (from head to tail): the pronephros, the mesonephros, and the metanephros.
- During the development of the kidney, there are three main structures initially, which

derive from intermediate mesoderm. These structures are pronephros, mesonephros and metanephros.

- The development of the kidney proceeds through a series of three successive phases, each marked by the development of a more advanced kidney:
 - The pronephros
 - The mesonephros
 - The metanephros
- The pronephros:
 - This is the most immature form of the kidney.
 - During approximately day 22 (fourth embryonic week) of human gestation, the paired pronephros appear towards the cranial end of the intermediate mesoderm.
 - It develop as a condensation of intermediate mesoderm in the lower cervical and upper thoracic regions extending to the cloaca, and almost entirely regresses in gestational week 4.
 - It appears as seven to ten cell groups and arrange themselves in a series of tubules called nephrotomes and join laterally with the pronephric duct.
 - The pronephric duct, which arises from dorsal and caudal evaginations of the pronephros, is preserved and ultimately will give rise to the mesonephric duct.
- The mesonephros:
 - The mesonephros begins to develop as the pronephros is regressing (fourth week).
 - The development of the pronephric duct proceeds in a cranial-to-caudal direction.
 - As it elongates caudally, the pronephric duct induces nearby intermediate mesoderm in the thoracolumbar area to become epithelial tubules called mesonephric tubules.
 - It starts as a series of S-shaped tubules.
 - Each mesonephric tubule receives a blood supply from a branch of the aorta, ending in a capillary tuft analogous to the glomerulus of the definitive nephron.
 - The tubules around the glomerulus will form a Bowmann's capsule around the capillary tuft.
- Together this will lead to the formation of a renal corpuscle allowing for filtration of blood.
- Laterally, the tubule enters the mesonephric collecting duct (wolffian duct).
- This filtrate flows through the mesonephric tubule and is drained into the continuation of the pronephric duct, now called the mesonephric duct or Wolffian duct.
- The nephrotomes of the pronephros degenerate while the mesonephric duct extends towards the most caudal end of the embryo, ultimately attaching to the cloaca.
- These mesonephric tubules carry out some kidney function at first, but then many of the tubules regress. However, the mesonephric duct persists and opens to the cloaca at the tail of the embryo.
- In both sexes, the ureters, renal pelvis, and bladder trigone are derived from the mesonephric duct.
- In males, the mesonephric duct also gives rise to the vasa deferentia, epididymides, and seminal vesicles; the former is part of the duct itself, while the latter two structures arise as a result of ductal dilatation or outpouching.
- Once the mesonephric duct comes in contact with the cloaca at the caudal aspect of the embryo, it then grows cranially as the ureteric bud until it comes in contact with the metanephric mesenchyme, forming the metanephros.
- The ureteric bud and metanephric mesenchyme reciprocally induce growth, forming the kidney.
- The metanephros:
 - This gives rise to the definitive kidney.
 - The metanephros develops from several components:
 - An outgrowth of the caudal mesonephric duct
 - The ureteric bud
 - A condensation of nearby renogenic intermediate mesoderm, the metanephric blastema
 - The metanephros appears during the fifth week of intrauterine life.

- The kidney has two parts:
 - The collecting system
 - The excretory system
- The collecting system develops from the ureteric bud which is an outgrowth of the mesonephric duct.
 - The ureteric bud penetrates the metanephric tissue.
 - The bud then dilates, forming a renal pelvis.
 - The renal pelvis will differentiate into the major calyces.
 - The major calyces will further differentiate and subdivide for 12 or more generations to form the minor calyces.
 - By the fifth generation, the renal pyramids are formed.
- The excretory system is formed because a metanephric tissue cap is induced by the collecting tubules to form renal vesicles.
- The vesicles form an s-shaped tubules which is covered in capillaries, giving rise to glomeruli.
- The tubules and the glomeruli form the nephron.
- Continues expansion of the tubules will form the convoluted tubules of the kidney and the loop of Henle.
- At birth, approximately 750,000–1 million nephrons are present in each kidney; postnatally, renal size may increase, owing to elongation of the proximal convoluted tubules.
- With differential longitudinal growth of the embryo, the kidney ascends from its initial location in the pelvis to its final location in the upper retroperitoneum.
- During ascent, transient blood vessels serially arise and degenerate; these arteries persist in ectopic kidneys as well as in some orthotopic renal units.
- Concurrently, the kidneys rotate around their vertical and horizontal axes so that their final orientation is one in which the upper poles are slightly more medial and anterior than the lower poles.
- The urogenital sinus can be further subdivided into cranial (future bladder) and caudal (future prostate, urethra, and external genitalia) portions.
 - The vesical epithelium is entirely derived from the endodermal layer of the urogenital sinus.
 - The mesonephric duct gives rise to the ureter.
 - With continued caudal growth of the embryo, the proximal end of the mesonephric duct is progressively absorbed caudally and the common portion of the mesonephric duct is absorbed into the bladder trigone and urogenital sinus.
 - The discrete branches of the mesonephric duct which will become the male genital ducts and ureters becomes distinct entities attached to the urogenital sinus.
 - The nonepithelial layers of the detrusor (non-trigone) portion of the bladder arise from condensations of splanchnic mesenchyme.
 - The lumen of the allantois, which connects the bladder and the anterior abdominal wall, closes over time, yielding the urachus. Over time, the urachus becomes more fibrotic and becomes the median umbilical ligament.
 - The prostate gland develops around 9–11 gestational weeks from the urogenital sinus, as endoderm invaginates into surrounding mesenchyme.
 - Prostate development is an androgen-dependent process.
 - It appears that the mesenchyme, rather than the endoderm, must be androgen-sensitive in order for normal prostatic development to occur.
 - The urethra develops from the urogenital sinus, with endoderm giving rise to the epithelium and splanchnic mesenchyme giving rise to the surrounding soft tissue.
 - In males, the most distal part of the urethra (the glanular portion) appears to arise from an ectodermal invagination which then joins with the endodermal epithelium of the proximal urethra to create a continuous channel.

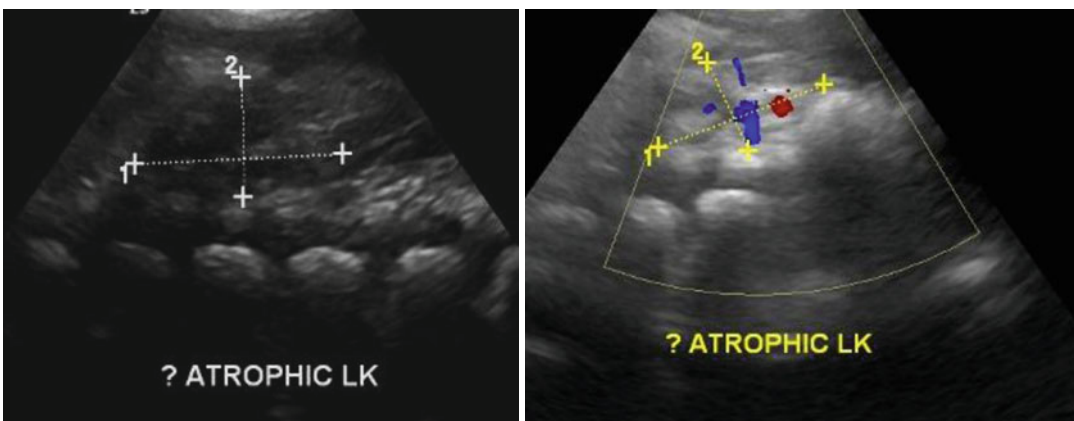
1.3 Abnormalities of the Kidney

- Normal renal development depends upon the interaction between the ureteric bud and metanephric mesenchyme, which induces organogenesis resulting in the formation of the 600,000–2 million nephrons and the collecting system of each kidney.

- The kidney is the most common site of congenital abnormalities.
- Renal malformations are often associated with other congenital defects such as a grossly deformed pinna with ipsilateral abnormalities of the facial bones.

1.3.1 Renal Agenesis

- Renal agenesis is a congenital malformation in which one (unilateral) or both (bilateral) fetal kidneys fail to develop.
 - Renal agenesis can be unilateral or bilateral but almost always unilateral.
 - Unilateral renal agenesis is a relatively common congenital urinary malformation.
 - It is usually diagnosed during fetal ultrasonography or incidentally on ultrasound done for other reasons.
 - Some cases of unilateral renal agenesis may represent involution of a previous multicystic disease of the kidney (Figs. 1.1, 1.2, and 1.3).
 - Up to 40% of women with a urogenital tract anomaly also have an associated renal tract anomaly.
 - Adults with unilateral renal agenesis have considerably higher chances of hypertension.
 - The annual incidence of unilateral renal agenesis is estimated at around 1 in 2,000 live newborns.
- The male to female ratio is around 1.2:1.
 - Approximately 56% of unilateral renal agenesis occurs on the left side.
 - Most patients with unilateral renal agenesis are asymptomatic if the other kidney is fully functional.
 - However, hypertension, proteinuria and renal failure may develop in the long term follow-ups (20–50% of cases at the age of 30), which may be based on glomerular hyperfiltration.
 - Due to this increased risk of hypertension and/or proteinuria, long-term follow-up of these patients is important.
 - Associated malformations:
 - Unilateral renal agenesis may be an isolated congenital malformation or may be associated with chromosomal abnormalities or a variety of nonchromosomal syndromes including the VACTERL and MURCS associations.
 - Congenital cardiac malformations are the most common malformations associated with unilateral renal agenesis.
 - Girls with unilateral renal agenesis should have a pelvic ultrasound to look for abnormalities in the müllerian structures.
 - Vesicoureteral reflux is the most common abnormality noted in the contralateral kidney.
 - It is associated with an increased incidence of Müllerian duct abnormalities, and can



Figs. 1.1 and 1.2 Abdominal ultrasound showing left atrophic kidney. This is most likely following involution of a previous multicystic kidney

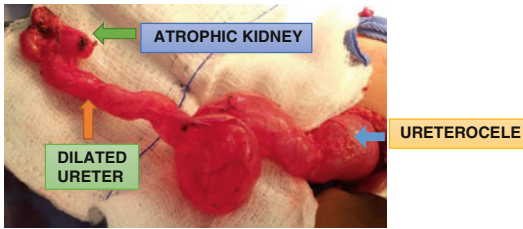


Fig. 1.3 Intraoperative photograph showing a very small atrophic kidney. Note also the massively dilated ureter secondary to an obstructive ureterocele

be a cause of infertility, hematocolpos, increased need for Caesarean sections, or other problems.

- Herlyn-Werner-Wunderlich syndrome is one such syndrome in which unilateral renal agenesis is combined with a blind hemivagina and uterus didelphys.
- Renal agenesis is occasionally associated with genital tract anomalies on the same side such as seminal vesicle hypoplasia and absence of the vas deferens.
- Other anomalies occur in up to 40% of patients, mainly cardiac (such as atrial or ventricular septal defects) and gastrointestinal (such as anorectal agenesis).
- Renal agenesis results from a development failure of the ureteric bud and the metanephric mesenchyme.
- Unilateral renal agenesis can be caused by mutations in many genes, such as *RET* (10q11.2), *BMP4* (14q22-23), *FRAS1* (4q21.21), *FREM1* (9p22.3 or UPK3A (22q13.31), *PAX2* (10q24.31), *HNF1B* (17q12), *DSTYK* (1q32).
- Unilateral renal agenesis can occur as part of multi-organ syndromes, several of which have defined genetic bases, including Kallmann syndrome, branchio-oto-renal syndrome, diGeorge syndrome, Fraser syndrome, MURCS association, Poland syndrome, renal cysts and diabetes syndrome, and Williams-Beuren syndrome.
- Maternal diabetes mellitus or use of specific drugs during pregnancy can also result in renal agenesis.
- Prenatal suspicion of unilateral renal agenesis is confirmed by postnatal ultrasound showing

an empty renal fossa, followed by renography to confirm the presence of a solitary functioning kidney.

- The size of the solitary functioning kidney is increased in the majority of patients.
- A voiding cystourethrogram should be considered in order to detect vesicoureteral reflux (VUR).
- The differential diagnoses include:
 - Extreme unilateral renal dysplasia
 - Involuted multicystic dysplastic kidney
 - Renal ectopia
- In most familial cases, unilateral renal agenesis is inherited in an autosomal dominant manner with incomplete penetrance.
- Unilateral renal agenesis can occur with dysplasia or hypoplasia of the solitary functioning kidney which makes the prognosis more serious.

1.3.2 Renal Hypoplasia

- Renal hypoplasia is a common congenital malformations.
- It is poorly understood and commonly used to describe a congenitally small kidneys with a reduced number of nephrons but normal architecture.
- There are however two distinct conditions:
 - Oligomeganephronia:
 - This is a type of renal hypoplasia that results from a quantitative defect of the renal parenchyma with a reduced number of nephrons.
 - Simple renal hypoplasia:
 - This is characterized by reduction in the renal mass but the number of nephrons is normal.
- It was estimated that renal hypoplasia affect about 2.2% of the population.
- The exact incidence of renal hypoplasia is not known but it is estimated to occur 1 in 400 live births.
- This however is not a true incidence of pure renal hypoplasia because the majority of congenitally small kidneys also exhibit evidence of renal dysplasia.

- Severe bilateral reductions in nephron numbers that are characteristic of renal hypoplasia/dysplasia are the leading cause of childhood end stage renal disease.
- A much less reduction in nephron number caused by mild bilateral renal hypoplasia, have been associated with the development of adult-onset hypertension and chronic renal failure.
- The diagnosis of hypertension in patients with unilateral hypoplasia/dysplasia is an indication for nephrectomy.
- Oligomeganephronia:
 - This results from arrested development of the metanephric blastema at 14–20 weeks' gestation, with subsequent hypertrophy of glomeruli and tubules in the kidney.
 - This hypertrophy and hyperfiltration results in further nephron injury and sclerosis. Eventually, this progressive loss of nephrons leads to end-stage renal disease (ESRD).
 - Oligomeganephronia is usually found in infants in their first year of life and presents with anorexia, vomiting, and failure to thrive.
 - After the first year of life, individuals with oligomeganephronia most often present with short stature, polyuria and polydipsia, or proteinuria.
- In some cases the separation of the reduplicated organ is incomplete forming fused supernumerary kidney.
- Associated anomalies:
 - Urogenital anomalies such as fusion anomalies, ectopic ureteric opening, vaginal and urethral atresia, urethral or penile duplication.
 - Non-urogenital anomalies such as coarctation of aorta, imperforate anus, ventricular septal defect and meningomyelocele.
- Supernumerary kidneys are most commonly located on the left side of the abdomen.
- A supernumerary kidney may be of same size as, larger than, or more commonly smaller than the usual kidney.
- It functions normally, possess a normal shape and capsule, and is either not attached to or loosely attached to the normal kidney but in an abnormal location.
- A supernumerary kidney may be located in front, below, above, or behind the normal kidney. They can also be found in the iliac region or anterior to the sacral promontory.
- The supernumerary kidney is thought to be an accessory organ with a separate arterial supply, venous drainage, collecting system, and distinct encapsulated tissue.
- It may have either a separate ureter or more commonly bifid ureters (50%). Rarely the ureter of the supernumerary kidney may have an ectopic opening.

1.3.3 Supernumerary Kidneys

- Supernumerary kidneys are a rare congenital anomaly of the urogenital system, where there are one or two accessory kidneys.
- A third kidney may be confused with the relatively common unilateral duplication of the renal pelvis.
- Supernumerary kidney results from the aberrant division and splitting of the nephrogenic blastema into two metanephric blastemas or from separate metanephric blastemas into which partially or completely reduplicated ureteral stalks enter to form separate encapsulated kidneys.
- The end result is two kidneys in association with a partially or completely duplicated ureteral bud.
- Symptoms have been noted in approximately two-thirds of the reported cases of supernumerary kidney. When symptomatic they may cause fever, pain, or palpable abdominal mass.
- The diagnosis of supernumerary kidney can be made by:
 - IVU
 - Ultrasonography
 - Nuclear scintigraphy
 - CT
 - MRI
- Bilateral supernumerary kidney is extremely rare.
- Surgery is indicated when supernumerary kidneys are affected by pathologic conditions and become symptomatic

1.3.4 Renal Dysplasia and Multicystic Kidney

(Figs. 1.4, 1.5, 1.6, 1.7, 1.8, 1.9, 1.10, 1.11, and 1.12)

- Renal dysplasia is characterized by the presence of malformed renal tissue elements, including primitive tubules, interstitial fibrosis, and/or the presence of cartilage in the renal parenchyma.
 - Multicystic dysplastic kidney (MCDK), a variant of renal dysplasia, is one of the most frequently identified congenital anomalies of the urinary tract.
 - Other terms used to describe this condition include multicystic kidney and multicystic renal dysplasia.
 - Multicystic kidney of the newborn is normally seen in only one kidney as an irregularly lobulated mass of cysts and usually absent or atretic ureter.
 - Multicystic dysplastic kidney is the most common cause of an abdominal mass in the newborn and is the most common cystic malformation of the kidney in infancy.
 - Renal dysplasia is considered the leading cause of end-stage renal disease in children.
 - Multicystic dysplastic kidney is characterized by:
 - The presence of multiple, noncommunicating cysts of varying size separated by dysplastic parenchyma and the absence of a normal pelvocaliceal system.
 - It is associated with ureteral or ureteropelvic atresia
 - The affected kidney is nonfunctional
 - Frequently, it is associated with contralateral abnormalities, especially ureteropelvic junction obstruction.
 - Dysplasia of the renal parenchyma is seen with urethral obstruction or reflux present early in pregnancy, or obstructed ureter.
- der in which abnormal cysts develop and grow in the kidneys.
- It is characterized by multiple cysts typically involving both kidneys.
 - About 15–17% of cases initially present with multiple cysts in one kidney, progressing to bilateral disease in the majority.
 - Polycystic kidney disease is one of the most common hereditary diseases.
 - It is the cause of nearly 10% of end-stage renal disease and affects males, and females equally.
 - Signs and symptoms of polycystic disease include:
 - High blood pressure
 - Headaches
 - Abdominal pain
 - Hematuria
 - Polyuria
 - Pain in the back
 - There are two types of polycystic kidney disease:
 - Autosomal dominant polycystic kidney disease (ADPKD)
 - Autosomal recessive polycystic kidney disease (ARPKD)
 - Autosomal dominant polycystic kidney disease (ADPKD):
 - This is the most common of all the inherited cystic kidney diseases
 - The incidence is 1:500 live births
 - It is estimated that about 10% of end-stage kidney disease (ESKD) patients being treated with dialysis were initially diagnosed and treated for ADPKD.
 - There are three genetic mutations with similar phenotypical presentation:
 - PKD-1
 - PKD-2
 - PKD3
 - Gene PKD1 is located on chromosome 16 and codes for a protein involved in regulation of cell cycle and intracellular calcium transport in epithelial cells.
 - Gene PKD1 is responsible for 85% of the cases of ADPKD.
 - Gene PKD2 is located on chromosome 4 and codes for a group of voltage-linked calcium channels.

1.3.5 Polycystic Kidney Disease

- Polycystic kidney disease, also known as polycystic kidney syndrome is a genetic disorder