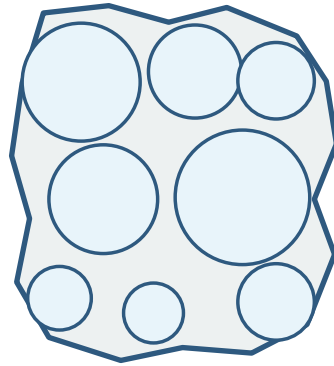


Multicystic renal dysplasia

Developmental pathology • gross and microscopic morphology • clinicopathologic interpretation



Dr Abu Suraih Sakhri

MBBS, MD (Pathology)

www.abusuraihsakhri.com

Learning outcomes

- Define multicystic renal dysplasia (MCDK) as a congenital maldevelopment rather than an acquired cystic disorder.
- Identify the gross and histologic features that establish renal dysplasia.
- Differentiate MCDK from polycystic disorders, hydronephrosis and cystic renal masses.
- Explain how laterality and the contralateral kidney determine clinical consequences.
- Distinguish the textbook surgical description from contemporary observation-based management.

What the term describes

Congenital renal dysplasia with multiple cysts of variable size and disordered nephron/collecting-system development.

- The affected organ is structurally abnormal, not simply a normal kidney bearing multiple cysts.
- Unilateral or bilateral involvement is possible.

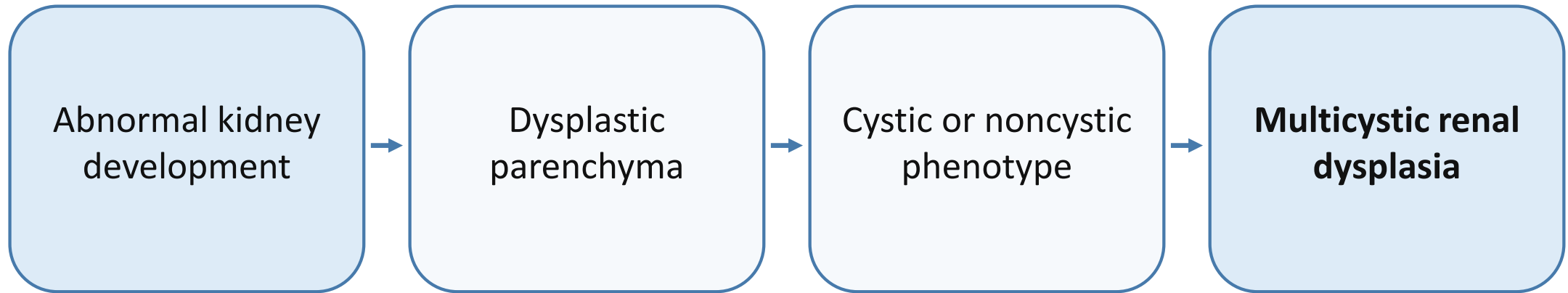
Dysplasia is a developmental diagnosis

Developmental dysplasia	Acquired cyst formation
Abnormal organization arising during organogenesis	Cysts arise in previously formed kidney tissue
Primitive ducts and undifferentiated stroma may persist	Primitive dysplastic tissue is not the defining feature
Frequently associated with urinary-tract malformations	Context depends on the acquired process

Why “multicystic” is only part of the name

- The cysts are conspicuous grossly and on ultrasound.
- The underlying disorder is abnormal renal differentiation and architectural organization.
- Primitive ducts, mesenchymal cuffs and sometimes cartilage give histologic specificity.
- Multiple cysts alone do not establish developmental dysplasia.

Renal dysplasia has a spectrum

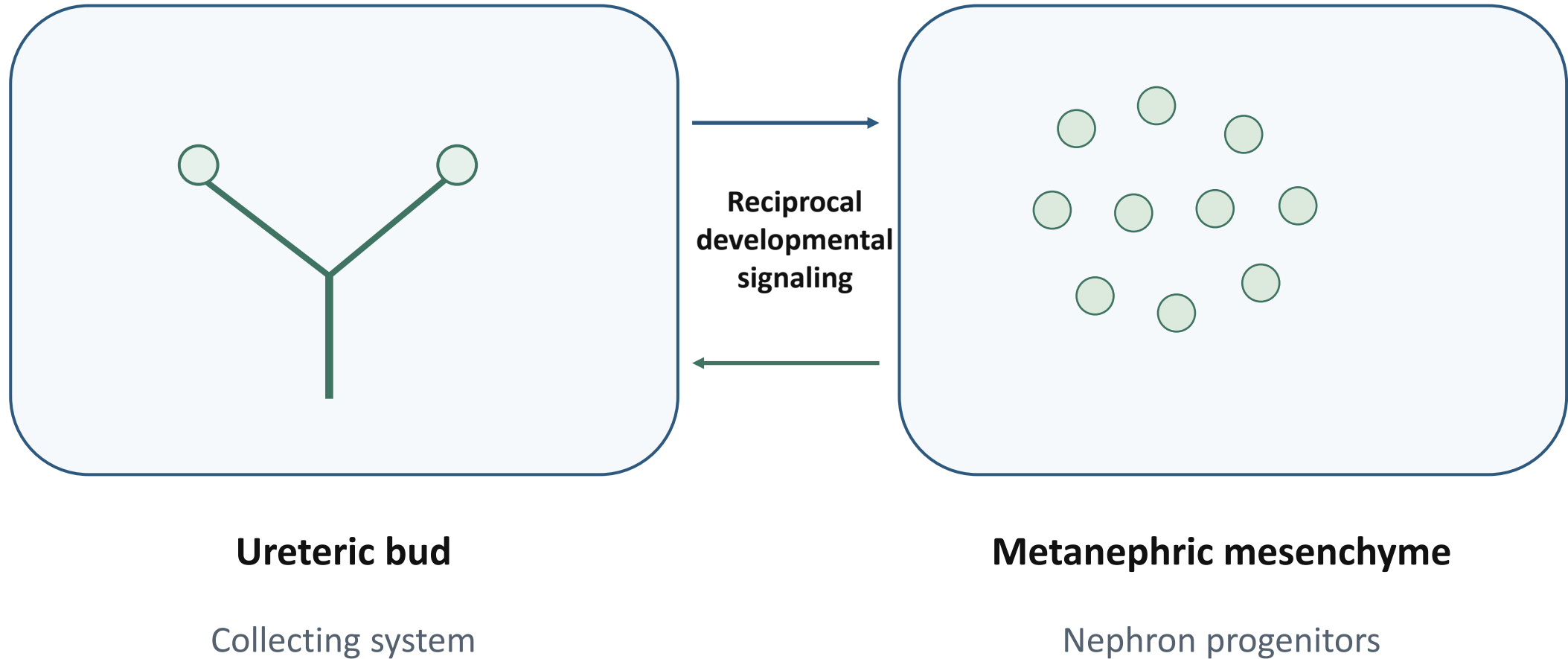


Conceptual sequence — original editable schematic

Affected tissue is not a normal functioning kidney

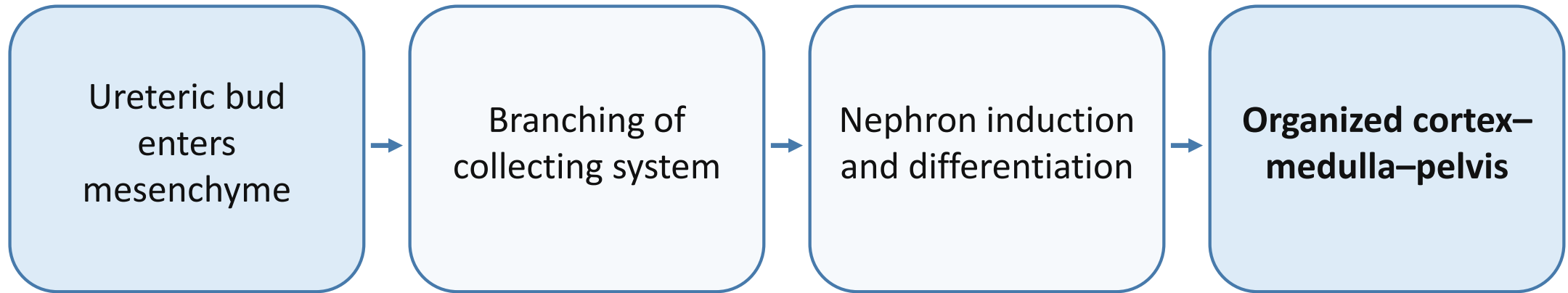
- Cysts displace or replace much of the functioning parenchyma in the classic phenotype.
- Some normal nephrons can still be identified microscopically.
- An affected MCDK often contributes little or no useful overall renal function.
- Clinical renal reserve depends on the functioning tissue elsewhere, especially in the opposite kidney.

Developmental context: Two interacting components



Original developmental schematic

Reciprocal induction builds the kidney

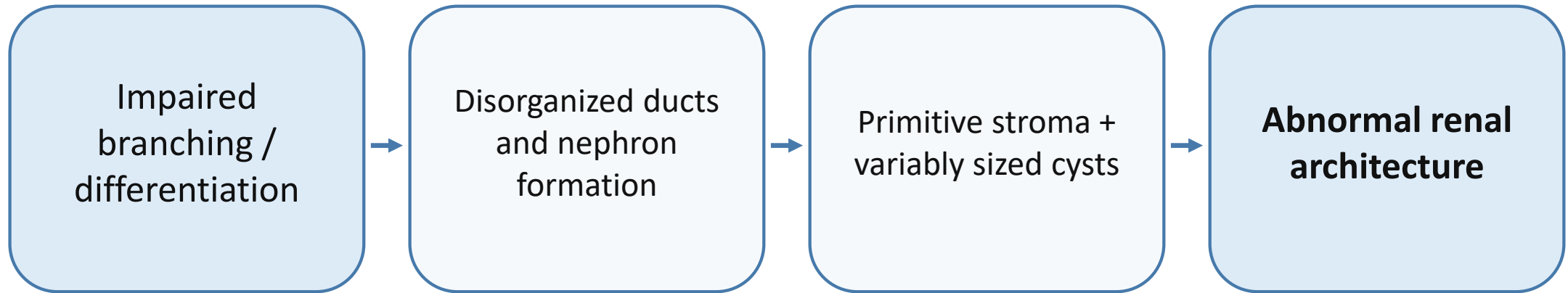


Conceptual sequence — original editable schematic

What normal renal architecture requires

Component	Normal result	Dysplastic contrast
Nephron differentiation	Mature tubules and glomeruli	Immature or aberrant structures may persist
Collecting-duct formation	Branching, organized drainage	Primitive ducts may be disordered
Stromal maturation	Ordered supportive interstitium	Persistent primitive mesenchyme
Renal organization	Recognizable parenchyma and pelvis	Architectural disorganization

A developmental route to dysplasia

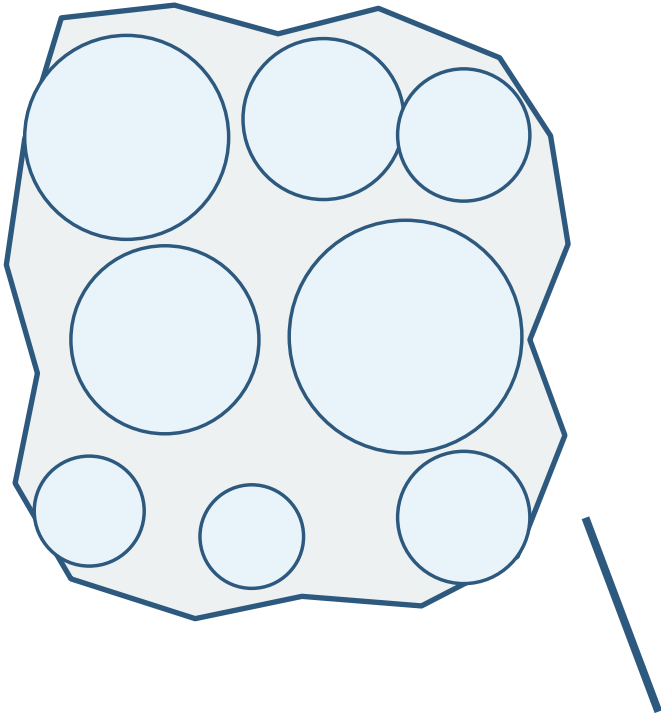


Conceptual sequence — original editable schematic

Obstruction and dysplasia frequently coexist

- Many cases with ureteropelvic obstruction, ureteral agenesis or ureteral atresia.
- Other lower urinary tract anomalies may accompany the dysplastic kidney.
- The association is important in a congenital anomalies of the kidney and urinary tract (CAKUT) evaluation.
- An associated obstructive lesion does not establish the same sequence of causation in every case.

The ureter is part of the pathology story



Kidney: Cystic disorganization

Ureteropelvic region

Ureter: Absent / atretic / obstructed

Sporadic cases and genetic heterogeneity

- Renal dysplasia as typically sporadic.
- Genetic syndromes and inherited susceptibility can nevertheless occur, as described by NIDDK and developmental reviews.
- Do not equate an isolated sporadic presentation with an invariably nongenetic disorder.
- Look for additional anomalies before assuming an isolated renal phenotype.

Why the CAKUT framework matters

Kidney morphology

Ureter and pelvis

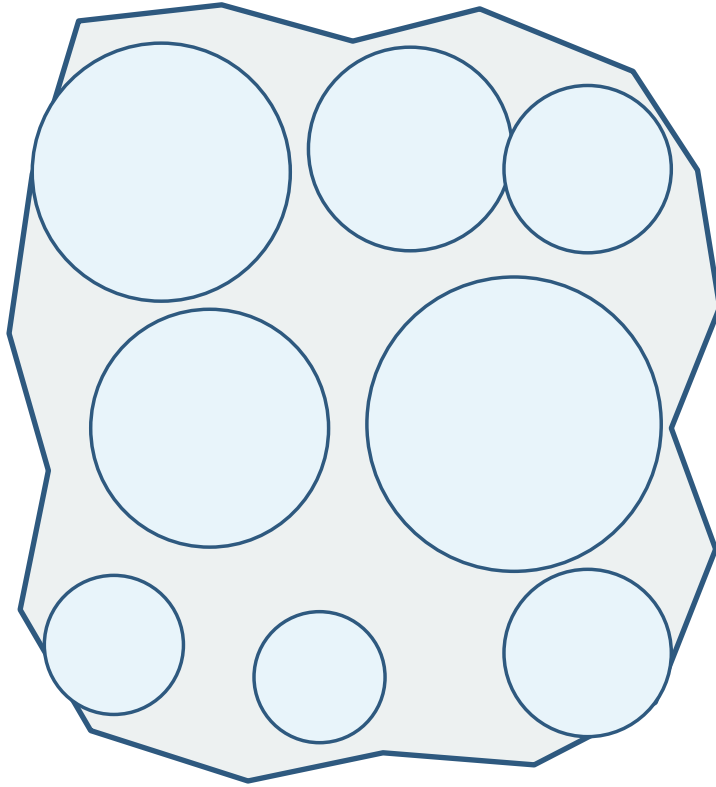
Bladder / outlet

Contralateral kidney

Laterality defines the principal clinical split

Unilateral MCDK	Bilateral MCDK
One dysplastic kidney	Both kidneys dysplastic
Contralateral kidney may maintain adequate function	Overall renal function is severely compromised
Usually favorable when the other kidney is healthy	Renal failure and serious neonatal consequences
Investigate additional urinary-tract anomalies	Requires specialist perinatal / renal planning

Gross morphology: Extremely irregular outline



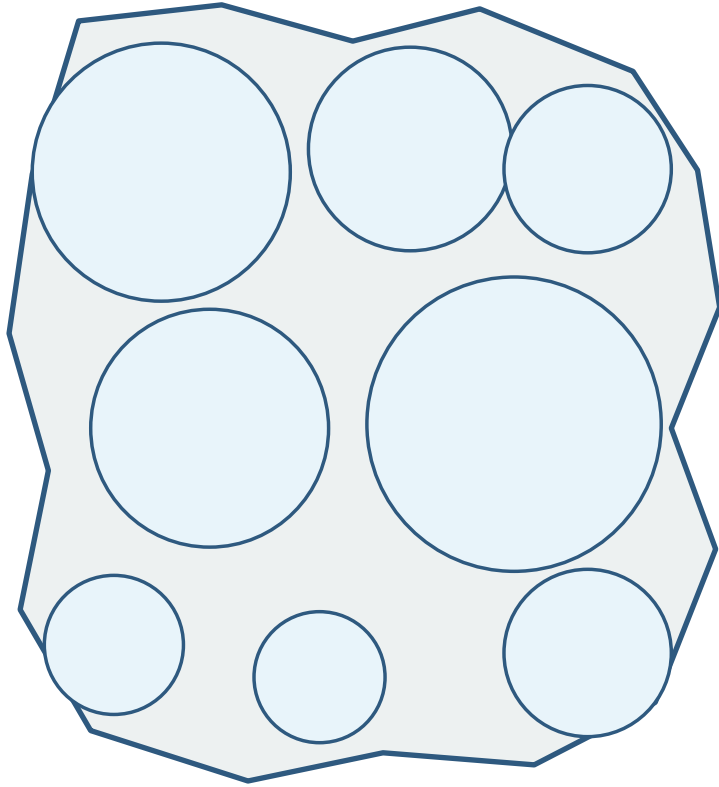
Original anatomical schematic

Irregular renal contour

Lobulated multicystic surface

Distorted parenchymal outline

Gross morphology: Varied cyst diameters



Original anatomical schematic

Small cysts

Intermediate cysts

Large cysts

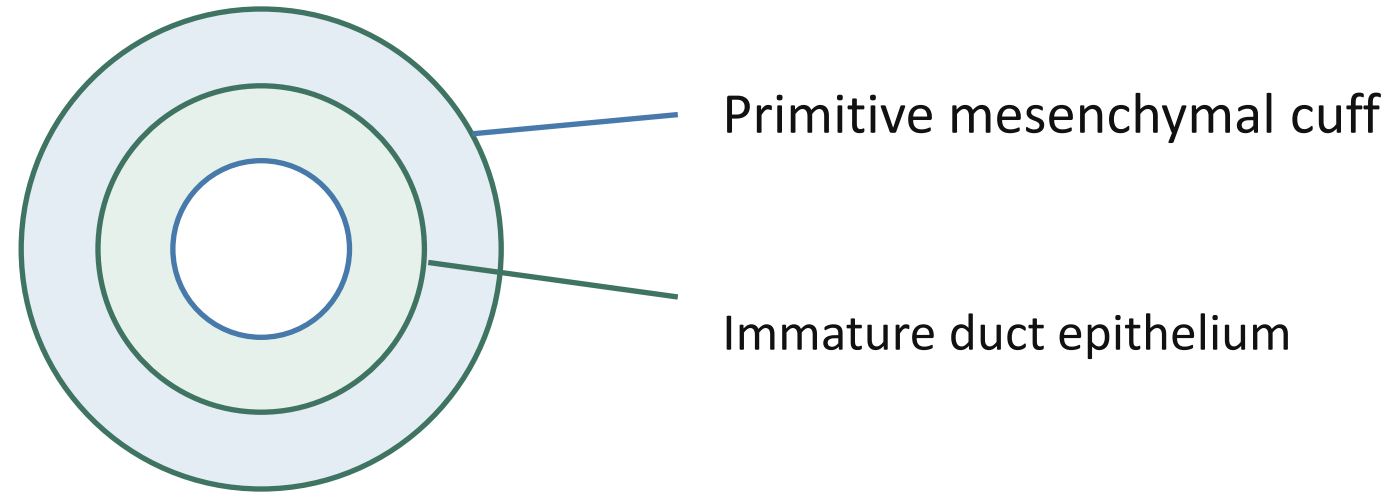
Gross appearance reflects lost organization

- The kidney is commonly enlarged and grossly distorted.
- Cysts of unequal diameter interrupt normal architecture.
- The cut surface does not resemble a simply dilated collecting system.
- Gross morphology suggests dysplasia, but histology provides defining developmental evidence.

Microscopic cyst lining

- Cystic cavities can be lined by flattened epithelium.
- The flattened lining is a nonspecific consequence of cystic dilatation.
- Diagnostic confidence rises when epithelial cysts coexist with disorganized, immature renal tissue.

Primitive ducts are central to histology



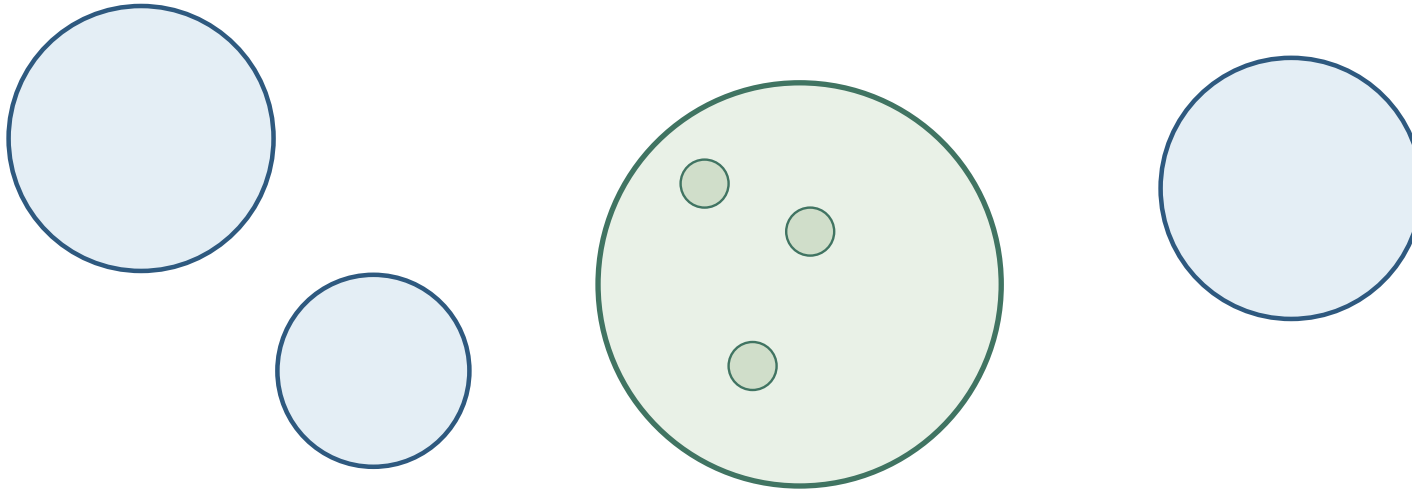
Central white space: Duct lumen

Schematic section—not an H&E specimen

Undifferentiated mesenchyme is diagnostic context

- Islands of undifferentiated mesenchyme among the cysts.
- Primitive mesenchyme reflects incomplete renal tissue differentiation.
- Cuffs surrounding immature ducts help distinguish dysplasia from ordinary cystic dilatation.
- Read mesenchyme and ducts together, not as isolated findings.

Cartilage signals aberrant differentiation



Immature duct / cyst

Focal cartilage island

Native schematic—not histology

Cartilage is helpful—not required

- Cartilage is a characteristic *possible* finding in renal dysplasia.
- Its absence does not exclude the diagnosis.
- Primitive ducts with mesenchymal collars and lobar disorganization are more reliable morphologic anchors.
- Do not label focal cartilage itself as evidence of malignancy.

Normal nephrons may persist within dysplasia

Residual normal structures	Dysplastic structures
Some formed glomeruli / nephrons may be present	Immature or poorly organized ducts
Function may be limited by overall abnormal organ	Large cysts disrupt parenchymal continuity
Finding a normal tubule does not negate dysplasia	Dysplasia is judged by the composite architecture

Morphologic constellation—not a single clue

Irregular multicystic kidney

Flattened epithelial cyst lining

Primitive ducts + mesenchyme

Cartilage can be present

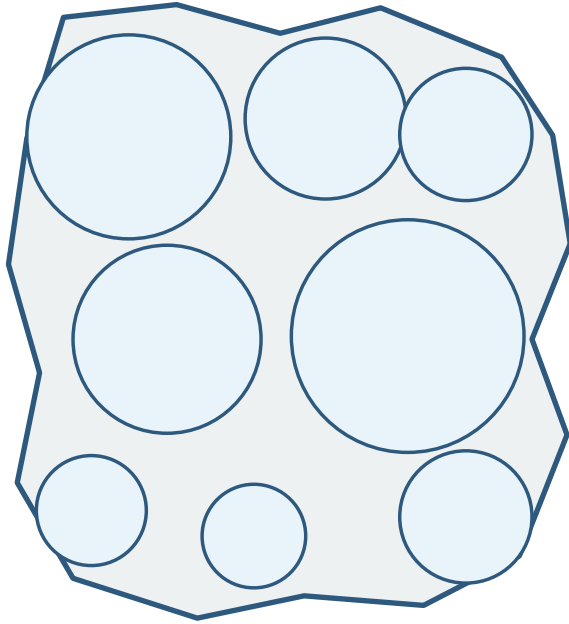
Gross and microscopic findings reinforce each other

At gross examination	At microscopy
Uneven, enlarged multicystic kidney	Varied cyst sizes and epithelial linings
Distorted renal architecture	Lobar disorganization and immature ducts
Suspicion of a developmental anomaly	Primitive stroma and possible cartilage substantiate it

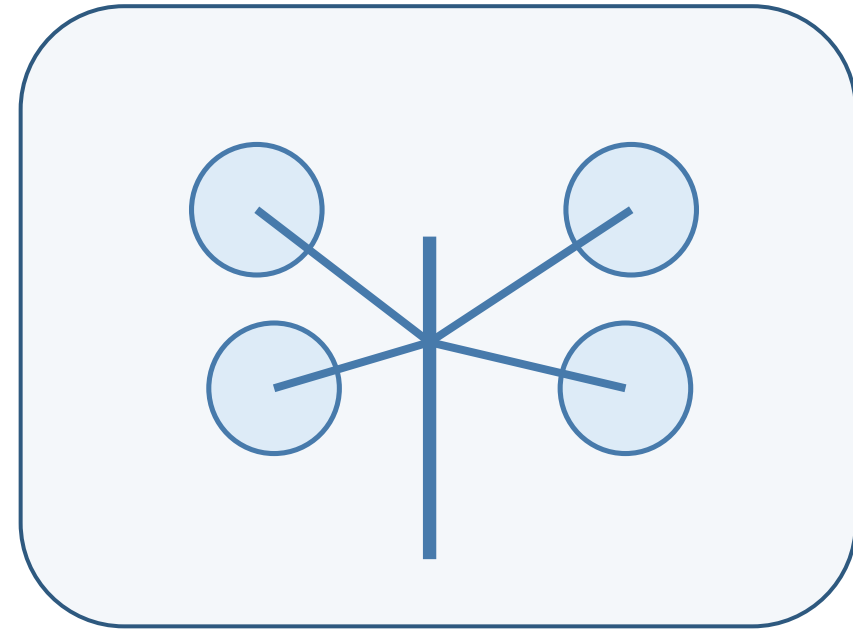
Prenatal ultrasound often begins the work-up

- Many cases are recognized before delivery during prenatal ultrasonography.
- Assessment includes the cystic kidney, the opposite kidney and the urinary drainage system.
- The amount of normal parenchyma and the pattern of cysts guide interpretation.
- Postnatal ultrasound can confirm the suspected diagnosis.

Imaging concept: Separate cysts versus dilated pelvis



Separate cystic spaces



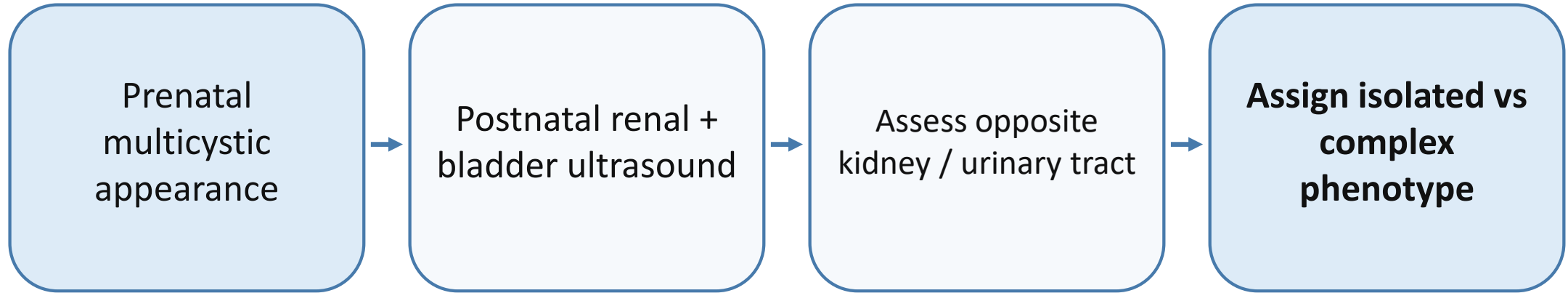
Communicating dilated collecting system

Schematic—NOT diagnostic ultrasound

Why cyst communication matters

MCDK pattern	Hydronephrotic pattern
Multiple cystic spaces, usually not communicating	Calyceal spaces often connect to renal pelvis
Disorganized architecture	Obstructed collecting-system dilation
May coexist with urinary-tract anomalies	Obstruction is itself the principal lesion

Postnatal confirmation is contextual

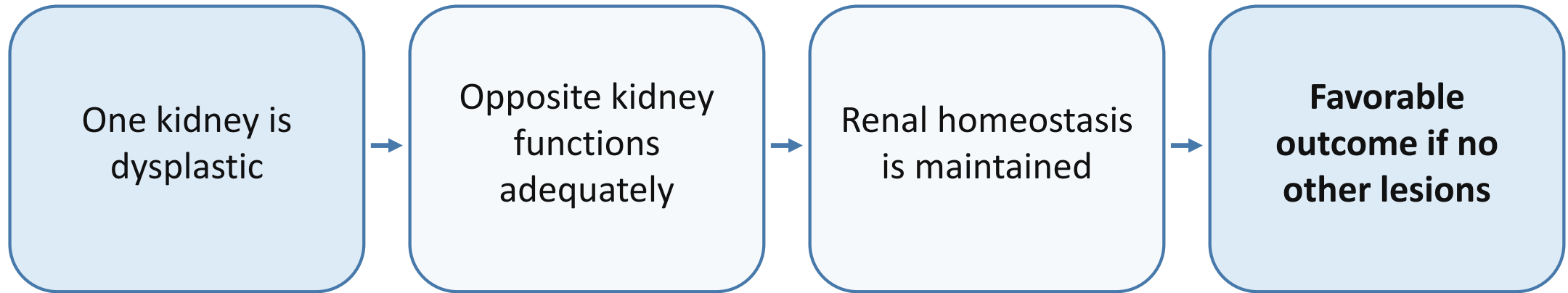


Conceptual sequence — original editable schematic

Do not overlook the opposite kidney

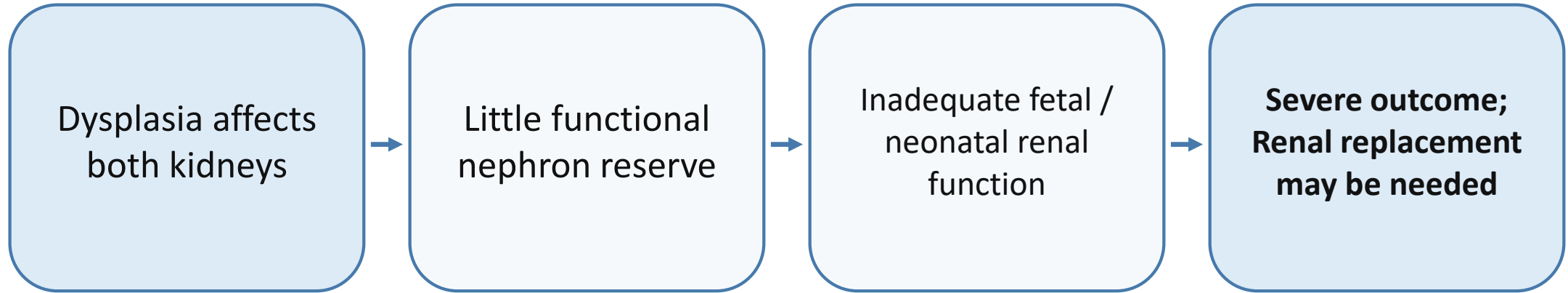
- It is the major determinant of renal reserve in unilateral MCDK.
- Ultrasound assesses presence, structural integrity and compensatory growth.
- Associated reflux or obstruction elsewhere can change follow-up needs.
- The term “unilateral” does not automatically mean the urinary tract is otherwise normal.

Unilateral MCDK: Mechanism of favorable prognosis



Conceptual sequence — original editable schematic

Bilateral disease: Why function fails

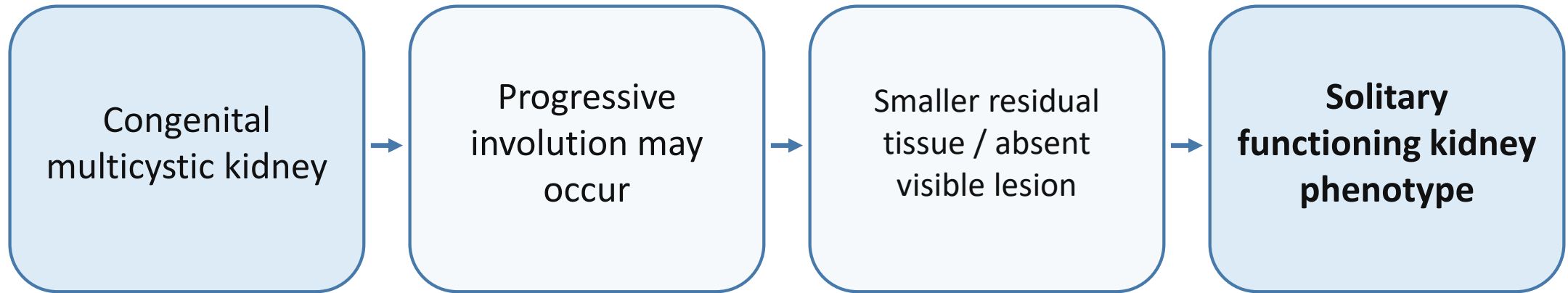


Conceptual sequence — original editable schematic

Antenatal bilateral involvement raises different concerns

- Both kidneys may lack sufficient functioning tissue.
- Severe fetal renal impairment can affect amniotic-fluid production and pulmonary development.
- Counseling must incorporate the entire fetal phenotype, not kidney appearance alone.
- Perinatal/nephrology input is appropriate; Prognosis depends on functional severity.

Spontaneous involution changes the longitudinal picture



Conceptual sequence — original editable schematic

Consequences of a single functioning kidney

- Adequate early renal function does not make follow-up irrelevant.
- Blood pressure, urine protein and kidney function may need periodic assessment.
- The contralateral kidney should be evaluated for congenital anomalies.
- An uncomplicated course is common, but monitoring is individualized.

Differential: Autosomal recessive PKD

MCDK	Autosomal recessive PKD
Usually asymmetric / often unilateral	Usually bilateral, enlarged kidneys
Cysts uneven, multiple sizes	Small collecting-duct dilations in radial pattern
Primitive ducts + stroma / cartilage	Uniform cuboidal-lined collecting-duct cysts
Other urinary anomalies may accompany	Congenital hepatic fibrosis / bile duct lesions

Differential: Autosomal dominant PKD

MCDK	Autosomal dominant PKD
Congenital dysplastic development	Inherited progressive cyst expansion
Variable cysts in disorganized kidney	Multiple tubule-derived cysts of varying linings
Frequently unilateral	Characteristically bilateral
Primitive immature tissue can be diagnostic	Polycystin gene abnormalities are characteristic

Differential: Nephronophthisis

MCDK	Nephronophthisis
Often enlarged, irregular multicystic organ	Kidneys typically small, with corticomedullary cysts
Primitive ducts and mesenchymal islands	Tubular atrophy, basement-membrane changes, fibrosis
Congenital anomaly presentation	Progressive tubulointerstitial dysfunction in childhood
Ureteral malformations may coexist	Ciliopathy gene associations

Differential: Medullary sponge kidney

MCDK	Medullary sponge kidney
Disorganized renal development	Collecting-duct cystic dilations limited to medulla
Grossly irregular / variable cyst sizes	Small cysts of papillary ducts
Functional tissue often markedly deficient	Renal function usually preserved
Primitive ducts and stroma	No defining primitive mesenchymal lesions

Differential: Acquired and simple cysts

MCDK	Acquired / simple cystic disease
Present from abnormal development	Arises in established kidney
May replace kidney and disrupt architecture	Simple cysts often leave rest of kidney intact
Primitive ducts / stromal dysplasia	No obligatory immature stroma or cartilage
Often associated with urinary anomalies	Acquired disease often related to chronic dialysis

A renal cystic mass is not automatically a tumor

- An enlarged irregular multicystic kidney can be mistaken for a neoplasm.
- The clinical differential includes congenital cystic lesions and less commonly cystic tumors.
- Imaging evaluation of solid elements, architecture and age matters.
- Uncertain atypical findings warrant expert evaluation, not a reflex assumption of benignity.

The comparative diagnostic matrix

Entity	Core distribution	Decisive clue
MCDK	Often unilateral; Variable-sized cysts	Primitive ducts + stroma
ARPKD	Bilateral small collecting-duct cysts	Hepatic fibrotic/biliary lesions
ADPKD	Bilateral progressive cysts	Familial / polycystin association
Nephronophthisis	Corticomedullary cysts	Tubular atrophy + interstitial fibrosis
Hydronephrosis	Collecting-system dilation	Connected pelvis and calyces

Triage a cystic fetal kidney systematically

Count the involved kidneys



Assess cyst size and communication



Look for collecting-system continuity



Inspect ureter / bladder / opposite kidney



Correlate anatomy with function

A pathology specimen: What to document

- Kidney size and contour; Number and heterogeneity of cysts.
- Whether a recognizable pelvis and ureter are present.
- Degree of renal parenchymal disorganization.
- Primitive ducts and cuffs of undifferentiated mesenchyme.
- Cartilage or other differentiation; Residual mature renal elements.

Histologic pitfall: Cartilage is not obligatory

Unsound conclusion	Better interpretation
No cartilage → not dysplasia	Assess ducts, stroma, architecture and cysts
A cyst proves dysplasia	Cystic change alone is nonspecific
Normal nephrons exclude dysplasia	Residual normal tissue may coexist
“Dysplasia” means premalignant change	Here it means aberrant organogenesis

A modern management distinction is essential

Robbins discussion	Contemporary pediatric approach
Unilateral cases may undergo nephrectomy	Observation is common when diagnosis is secure and asymptomatic
Focuses on excellent post-nephrectomy prognosis	Focuses on opposite kidney, involution and renal reserve
Historical clinical morphology framing	Surgery reserved for selected clinical problems

Conservative care when unilateral and uncomplicated

- Confirm that the multicystic lesion is typical and the other kidney is healthy.
- Observation is generally favored over automatic nephrectomy.
- Monitor for issues related to a solitary functioning kidney.
- Specific imaging and clinic schedules depend on local pediatric nephrology/urology protocols.

When does surgery enter consideration?

- NIDDK notes nephrectomy may be considered for a persistently problematic nonfunctioning kidney.
- Examples include troublesome infection, symptoms, or hypertension attributable to the abnormal kidney.
- Suspicion of another diagnosis may require further assessment.
- Size or absence of involution alone does not universally mandate removal.

“Simple” versus “complex” MCDK

Simple unilateral phenotype	Complex phenotype
Typical MCDK and unaffected opposite kidney	Associated urinary/renal abnormalities
Compensatory contralateral growth	Potential impairment of renal reserve
May leave specialist urologic follow-up after confirmation	Requires individualized urology/nephrology follow-up

Follow-up targets: Function, not just cyst size

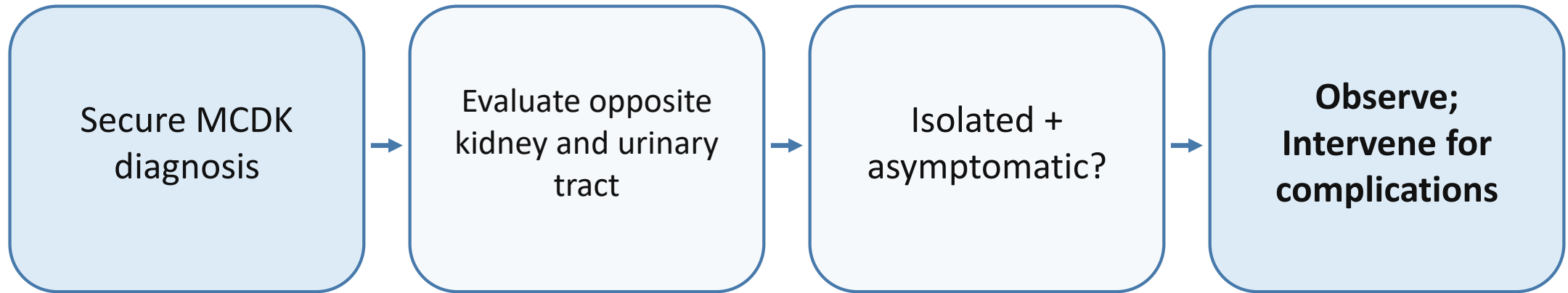
Blood pressure

Urine protein

Renal function

Healthy kidney growth

Management is a phenotype-driven decision



Conceptual sequence — original editable schematic

Case 1: Antenatal cystic kidney

A routine fetal scan detects one irregular kidney containing numerous cysts of different sizes. The other kidney appears structurally normal.

Discussion questions

- What developmental diagnosis is most likely?
- What additional urinary-tract features should be assessed?
- Which finding would most strongly alter predicted renal function?

Case 1: Diagnostic reasoning

- Working diagnosis: Unilateral multicystic renal dysplasia.
- Cyst heterogeneity and loss of organized renal architecture support it.
- Check collecting-system continuity, ureter/bladder and contralateral kidney.
- Final risk assessment depends on preserved functional renal tissue in the opposite kidney.

Case 2: A surgical pathology specimen

An irregular nephrectomy specimen shows large and small cysts. Sections reveal flattened cyst epithelium and immature ducts surrounded by primitive stroma.

Discussion questions

- Which microscopic findings identify a developmental process?
- Does absence of cartilage invalidate the diagnosis?
- Why might the gross specimen mimic a cystic mass?

Case 2: Morphologic diagnosis

- Multicystic renal dysplasia: Epithelial cysts plus immature, disorganized ducts and mesenchymal cuffs.
- Cartilage is supportive but not obligatory.
- The lobulated multicystic gross mass explains potential confusion with neoplasia.
- Describe associated abnormalities of the ureter and remaining renal tissue.

Case 3: Bilateral cystic enlargement

A neonate has bilateral markedly abnormal kidneys. Both lack organized functioning parenchyma on evaluation.

Discussion questions

- Why is prognosis unlike isolated unilateral MCDK?
- How should bilateral involvement influence consultation?
- Which differential with bilateral collecting-duct dilatation is important?

Case 3: Function and differential

- Bilateral disease can leave inadequate functioning nephron mass and cause renal failure.
- Urgent multidisciplinary assessment is appropriate.
- ARPKD is a key alternative: Bilateral small ductal dilatations and associated hepatic lesions.
- Do not diagnose a specific entity from “bilateral cysts” alone.

Three diagnostic anchors

Developmental disorganization

Primitive ducts + mesenchyme

Cysts of varying size

Evaluate the other kidney

Four clinically actionable principles

- Gross cysts are conspicuous; Histologic dysplasia is the defining process.
- Unilateral and bilateral involvement have profoundly different functional implications.
- Assess the entire urinary tract, not the affected kidney in isolation.
- For typical uncomplicated unilateral MCDK, modern care commonly observes rather than removes the lesion.

References and source distinctions

Robbins, Cotran & Kumar. Pathologic Basis of Disease. 11th ed. 2026; Chapter 20, p. 859 (primary morphologic source).

Jain S, Chen F. Developmental pathology of congenital kidney and urinary tract anomalies. Clin Kidney J. 2019;12:382–399.

NIDDK. Multicystic Dysplastic Kidney. Updated January 2023.

Psooy K. MCDK in the neonate: The role of the urologist. Can Urol Assoc J. 2016.

UC Davis Department of Pathology. Renal dysplasia case discussion. January 2022.