TUTORIALS

PG Residency

Pediatrics

Pediatric Nephrology

Smart Notes. Better Scores.

Table of Content

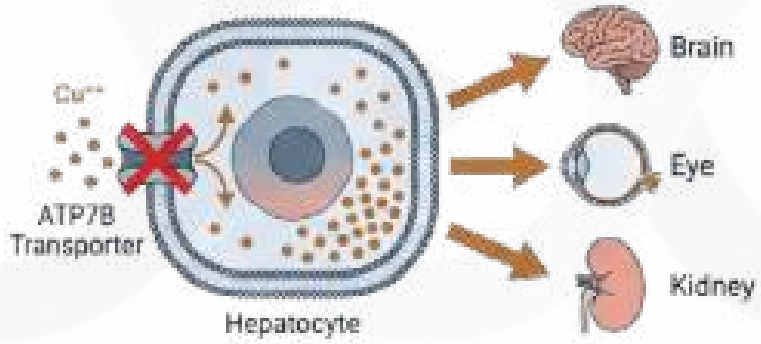
Wilson Disease: Genetic Basis & Pathophysiology

AutosomalRecessive I ATP7BMutation I Chromosome13

- **Definition**
  MutationinATP7B gene encoding transmembrane ATP-ase transport protein.
- **Mechanism**
  Failureofcoppertransport out of liver cells.
- **Result**
  Copperaccumulation in Liver, Brain, Eyes, and other organs causing oxidant damage.
- **Systemic Scope**
  Multi-systemic(Hepatic, Neuro, Psych, Cutaneous, Skeletal, Endocrine).

The Copper Blockade & Expression Spectrum

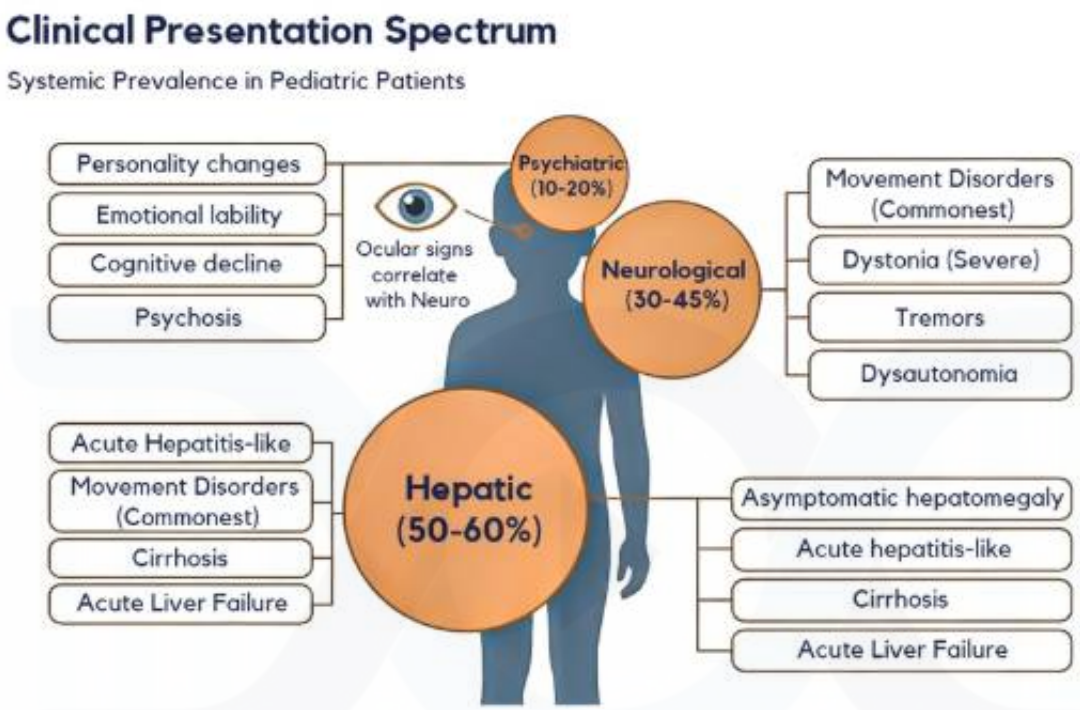
A)The Cellular Defect



B)Variable Expressivity

Clinical Presentation Spectrum

Systemic Prevalence inPediatric Patients



Study Structure

- Kidney
- Skeletal
- Endocrine (menstrual abnormalities)

Seizures:

5-10% of cases (Status epilepticus rare)

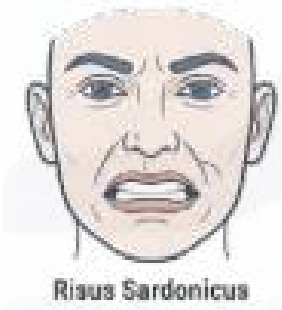
Neurological Subtypes: Denny Brown Classification

Distinct Phenotypes & Physical Exam Findings

Tremor-Predominant (70%)

- Wing-Beating Tremor (Pathognomonic)
- Elicited by proximal arm flexion
- Basal ganglia & dentate-rubro-thalamic pathway

Dystonia-Predominant (13-30%)



- Most severe form
- Focal, segmental, orgeneralized
- Hypomimia (expressionless) or Risus Sardonicus
- **WARNING:** Status Dystonicus = Emergency (Rhabdomyolysis/ AKI)

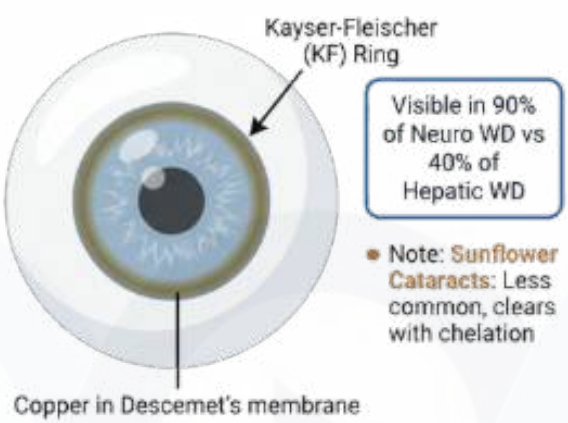
Nigrostriatal (11%)

- Rigidity & Bradykinesia
- Resting tremor
- Drooling, gait difficulty

Diagnostic Signs: Ocular & Neuroimaging

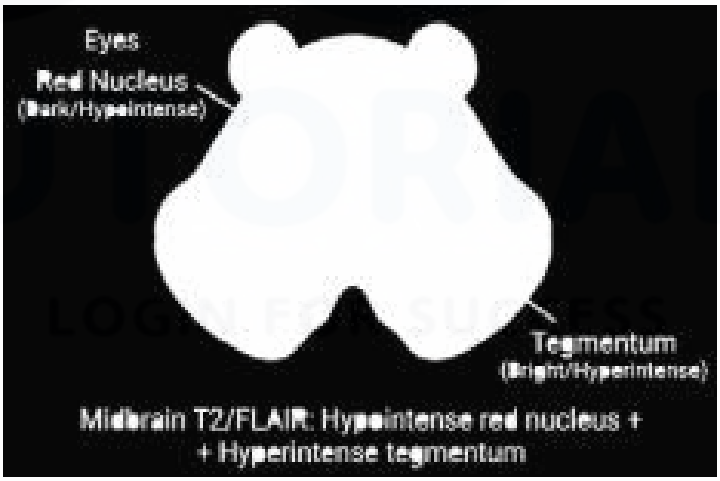
KayserFleischerRingsandThe'GiantPanda'Sign

The Eye (Ophthalmology)



The Brain (MRI)

Face of the Giant Panda Sign



Epidemiology & Diagnosis: Leipzig Scoring System

MeanPediatricOnset:7-9Years I Prevalence:3-10per100,000

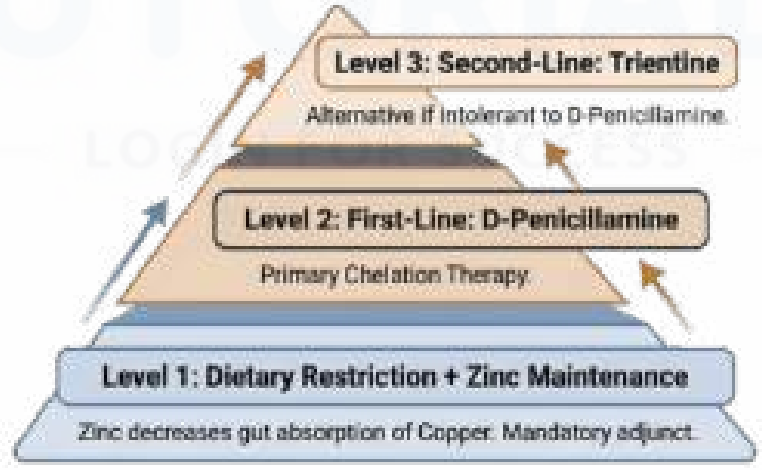
The Leipzig Diagnostic Calculator

	Kayser-Fleischer Rings	+2
	Neurological Symptoms	+1
	Serum Ceruloplasmin (< 20 mg/dL)	+1
	24hr Urine Copper (> 30 µg)	+1
	Liver Copper (Biopsy)	+2
	Coombs-negative hemolytic anemia	+1

Traffic Light Scoring Logic

Management Strategy: Copper Chelation

Goal:Removeexcesscopperandpreventre-accumulation



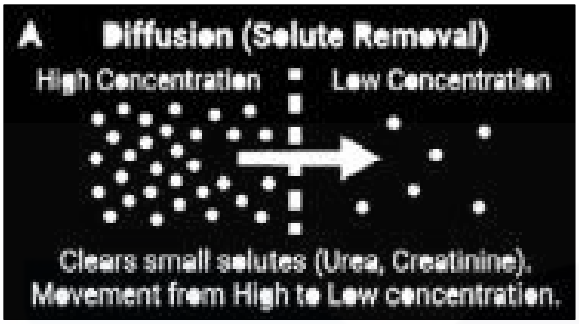
Prognosis

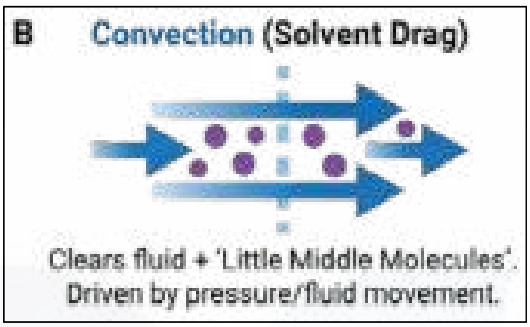
- Generally favourable if diagnosed and treated in time.
- Clinical signs (KF rings, cataracts) often clear with chelation.
- Genetics: Testing may be negative even with clear presentation (New genes emerging).

Pediatric RRT: Foundations & Mechanisms

ReplacesKidneyFunctioninAKI/CKI I RequiresPICU+Nephrology Team

Physics of Clearance



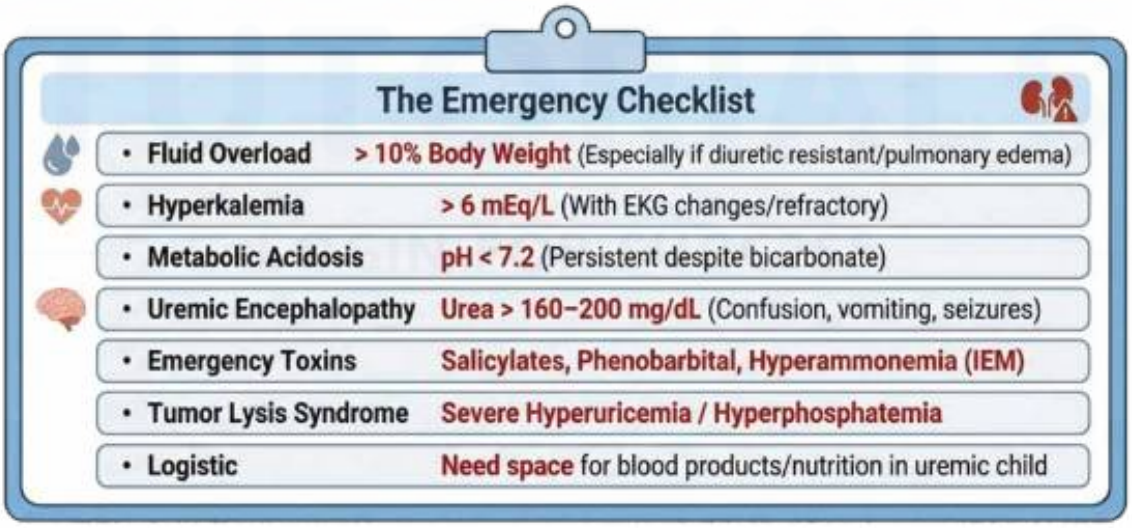


⚠ **Limitation:** Does not replace metabolic or hormonal kidney functions.

Risk: CKI incidence after pediatric AKI is 20-30%.

Indications for Initiation: The Non-Negotiables

Critical thresholds for emergency start



Modality Selection Matrix

ChoosingbetweenPD,IHD,andCRRT based on patient status

Peritoneal Dialysis (PD)

Patient Status: Stable OR Unstable

Speed: Continuous

Pros:

- Simple, Low Cost
- No Vascular Access

Cons:

- Poor solute clearance
- Infection risk (Peritonitis)
- Uncontrolled Ultrafiltration

Best For: Difficult access

- Neonates, Low resource

Intermittent Hemodialysis (IHD)

Patient Status: Stable Only

Speed: Rapid (Intermittent)

Pros:

- Rapid toxin removal
- Controlled fluid removal

Cons:

- Hypotension
- **DialysisDisequilibrium Syndrome (DDS)**

Best For: Rapid toxin clearance

Continuous (CRRT)

Patient Status: **Clinically Unstable**

Speed: Continuous (>24hr)

Pros:

- Hemodynamic stability
- Precise fluid control

Cons:

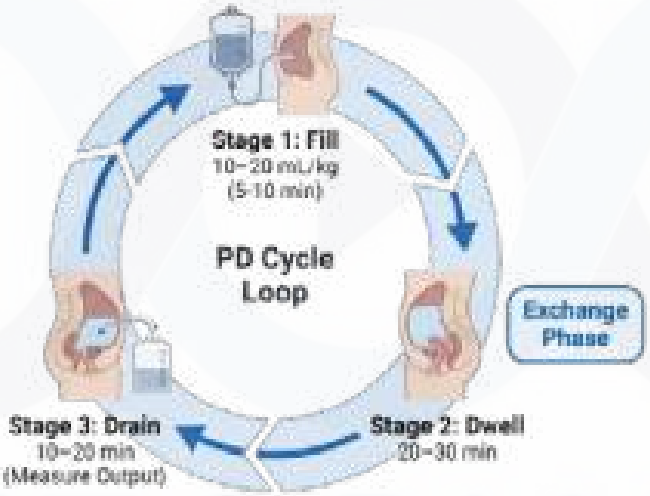
- Immobilization
- High cost
- Prolonged anticoagulation

Best For: Sepsis, Shock

- Multi-organ failure

Peritoneal Dialysis (PD): Procedure & Prescription

Cycle Logic and Dwell Times



Prescription Basics

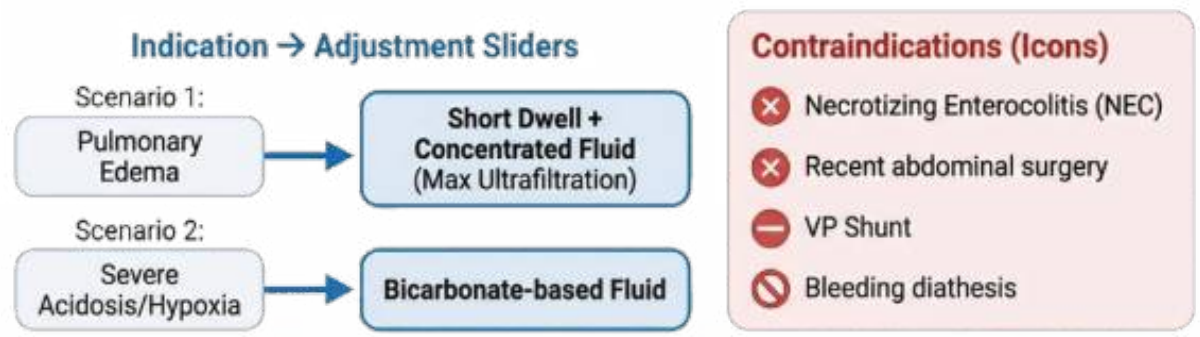
- **Access:** Abdominal catheter.
- **Fluid:** 1.7% Dextrose (Std) up to 2.5%. Lactate or Bicarb based.
- **Additives:** Heparin (500u/L), Potassium (2-4 mEq/L after stabilization).

Key Concept Box: Dwell Time Physics

- **Short Dwell:** Better Diffusion (Solute Clearance).
- **Prolonged Dwell:** Equilibrium reached = Less clearance, More fluid removal.

PD Safety: Modifications & Complications

Monitoring Requirements and Contraindications



Non-Negotiable Monitoring

Fluid Check

- Color (turbid=infection), fibers/threads

Labs

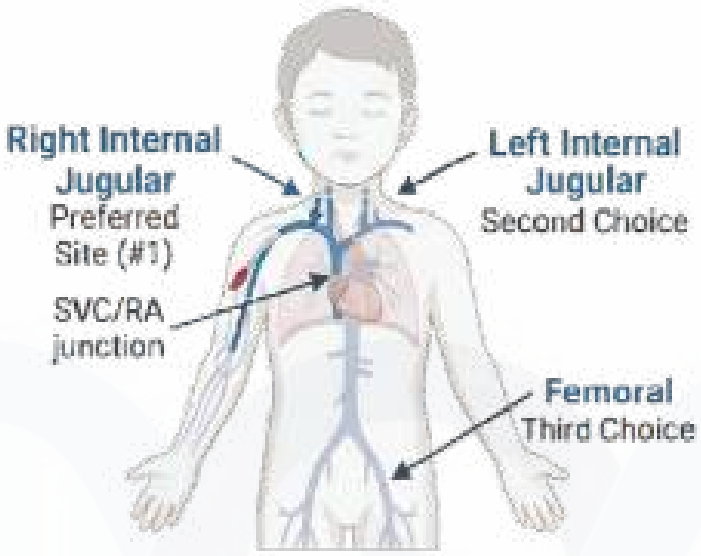
- Electrolytes/Sugar q12h (Risk: Hypoglycemia/Hyperglycemia).

Complications

- Peritonitis, Leakage, Hypothermia, Poor drainage.

Intermittent Hemodialysis (IHD): Technical Setup

Access Sites & Equipment Sizing Rules



Critical Equation

Dialyzer Surface Area ≤ Patient Body Surface Area (BSA)

- Smallest size available: 0.2 m2

Safety Note: Prevents hemodynamic collapse.

Critical Equation

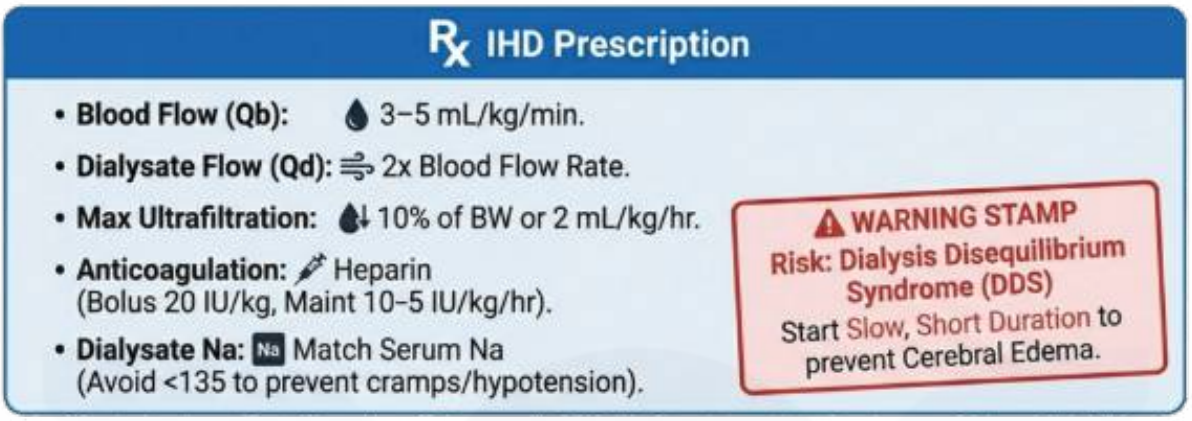
- Dual flow.
- Heparin lock (1000u/mL).

Contraindications

- Hemodynamic instability,
- Small infants (technical limits).

IHD Prescription & Safety Parameters

Dosing, Flow Rates, and AvoidingDDS

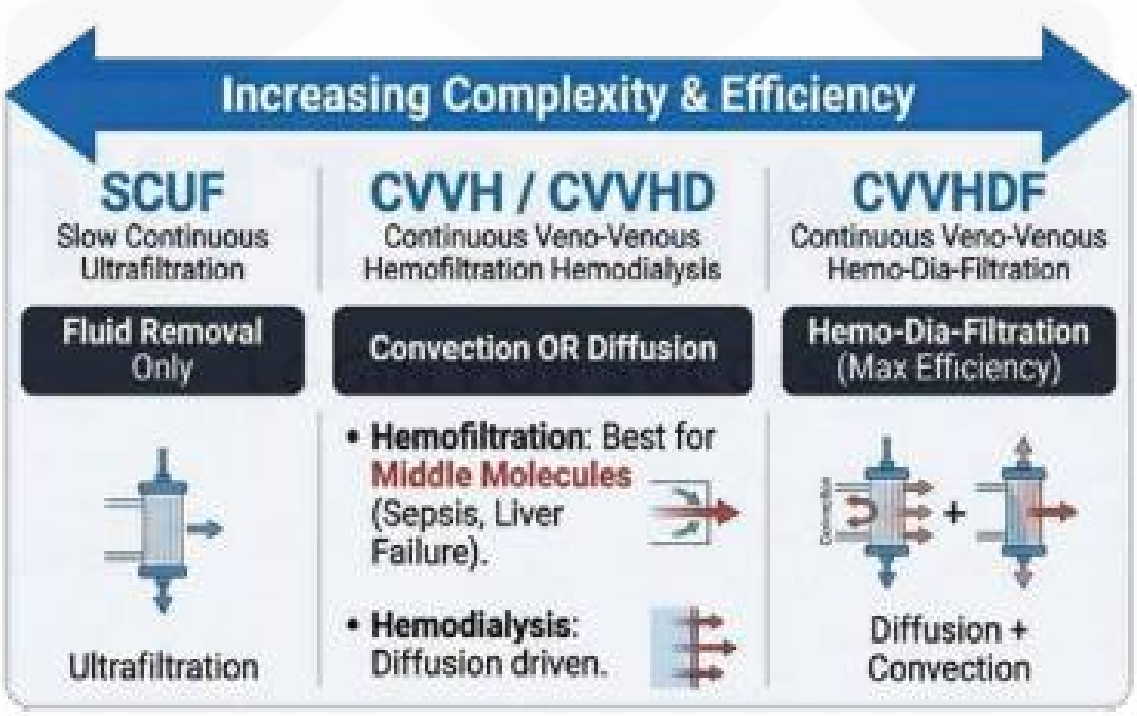


Target Kt/V: 1.2-1.4

** Measure of dialysis adequacy.

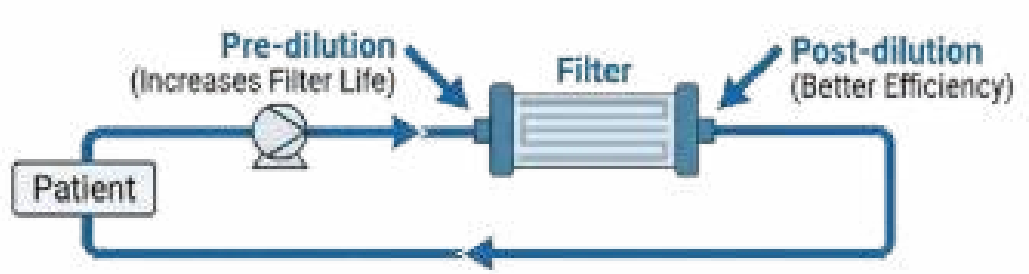
CRRT: Modalities & Indications

ContinuousTherapy forUnstable Patients



CRRT: Prescription Logic

Dosing, Dilution, and Priming



1. Dosing (Effluent Dose)

- Standard: 20-25 mL/kg/hour.
- Hyperammonemia: > 50 mL/kg/hour.

2. Flow Rates

- Blood Flow: 3-5 mL/min.
- Filtration Fraction: < 25%.

3. Priming

- Saline or Albumin.
- **Whole Blood:** Required for small infants/neonates to prevent hemodilution.

Notes:

DOC TUTORIALS

MADE FOR PG RESIDENTS · FOCUSED ON YOUR SUCCESS

PG Residency + NEET SS 2 + 1 Combo NEW

One Plan.
Two Goals.
Your Best Future.

Designed for PG Residents preparing for both PG Residency & NEET SS.

Structured Learning
Subject-wise & Systematic

Consistent Practice
High-yield QBank + Regular Tests

Smart Revision & Recall
Revision Notes + Memory Boosters

FREE Access in PG Year 1

Pay Only for PG Year 2 & Year 3

Lock Today's Price

2 Goals. 1 Plan.

Finish Strong in PG Residency.
Crack NEET SS.

Subscribe Now