Approach to Renal Tubular Acidosis

Dr. Jalpa Dave

Pediatric Nephrologist

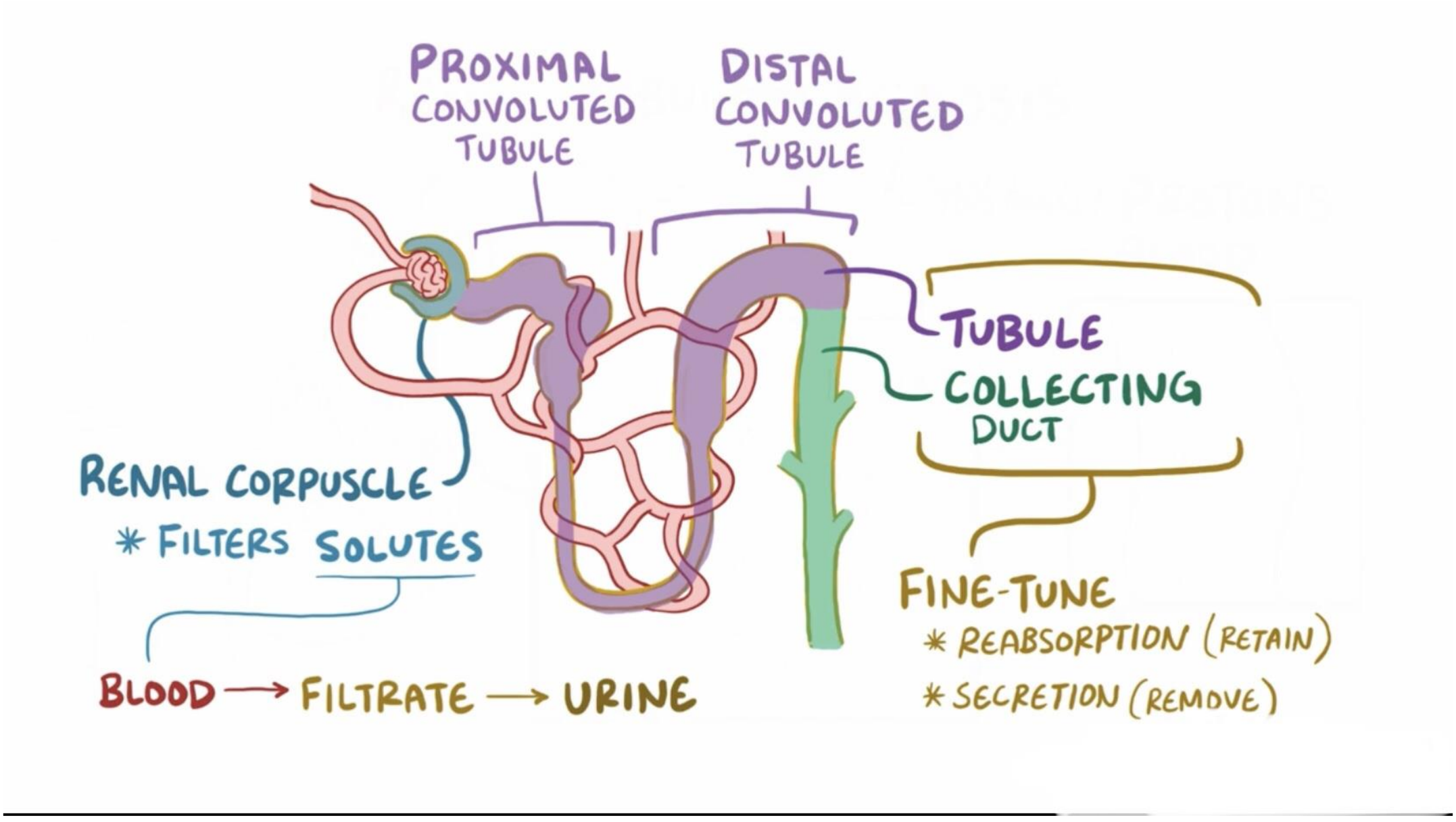
Vadodara

ECHO India

Project ECHO®

KIDNEY FOUNDATION FOR CHILDREN

Renal acid base homeostasis



Definition

- Renal tubular acidosis is a group of **acquired or hereditary** renal tubular disorders which occurs due to defect in

- **Net acid excretion or bicarbonate reabsorption** or both
- With normal or near **normal glomerular filtration rate**
- Leading to **non anion gap hyperchloremic metabolic acidosis**

- Anion gap = Serum (Na) - Serum (Hco3 +Cl-)

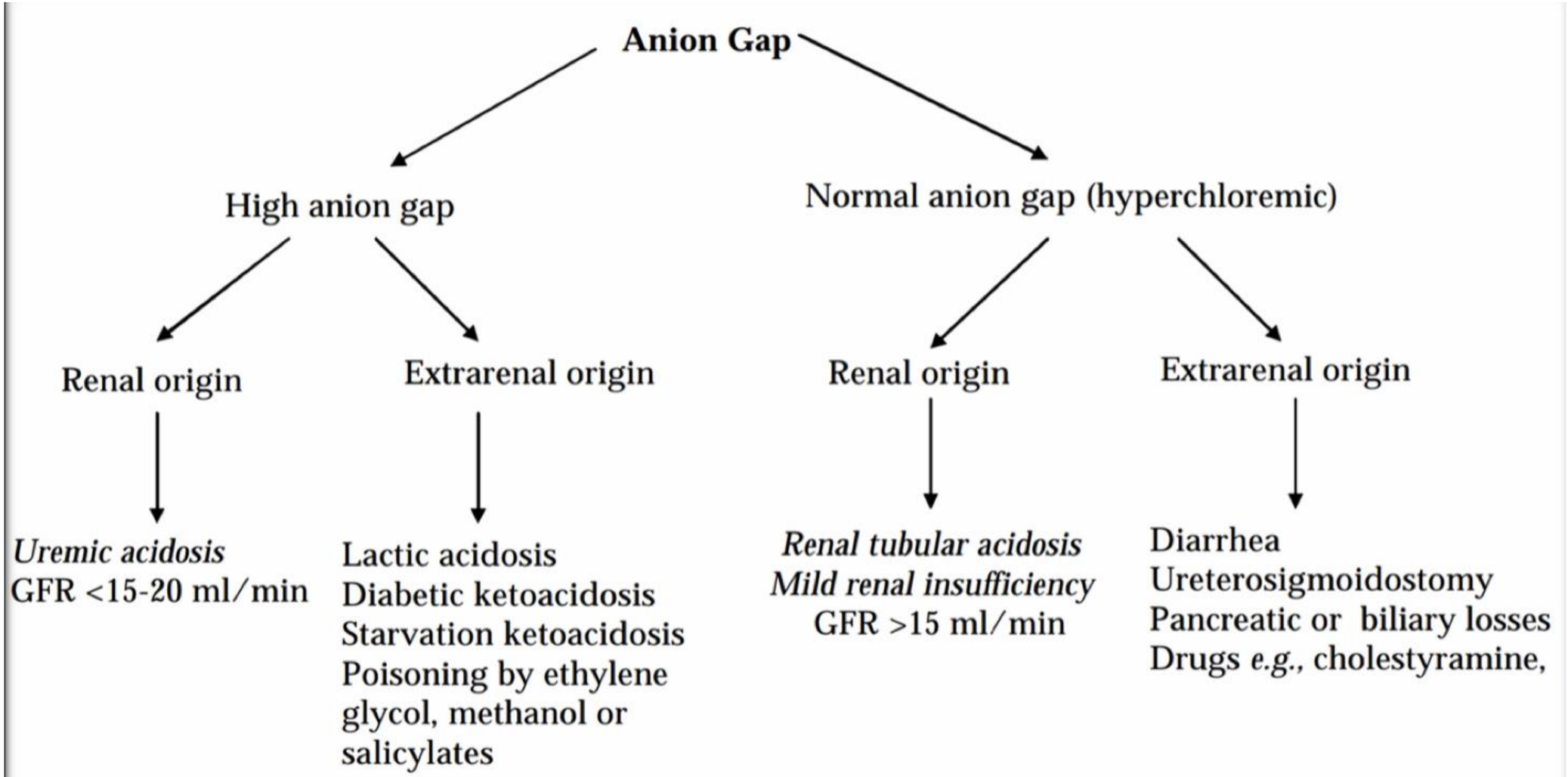
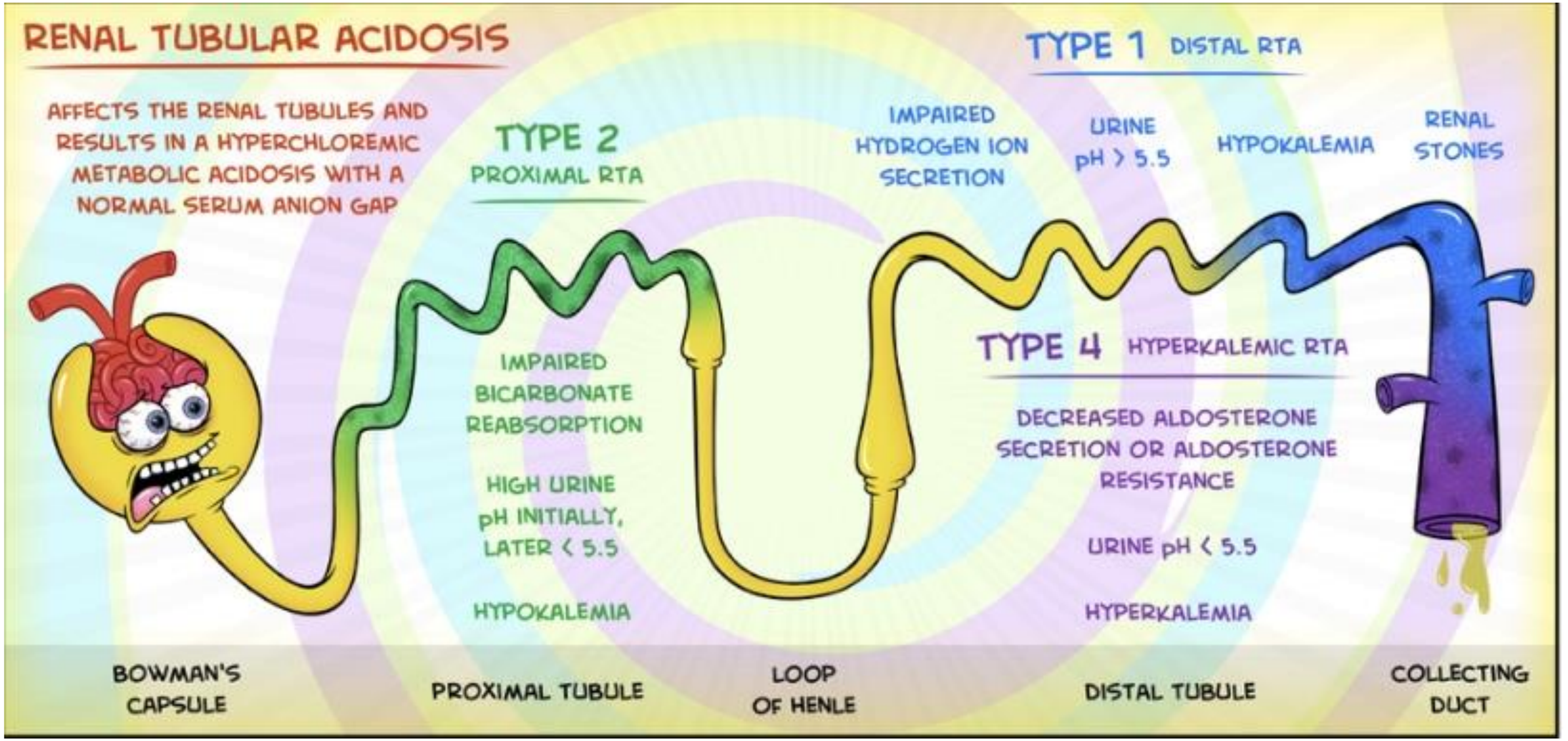
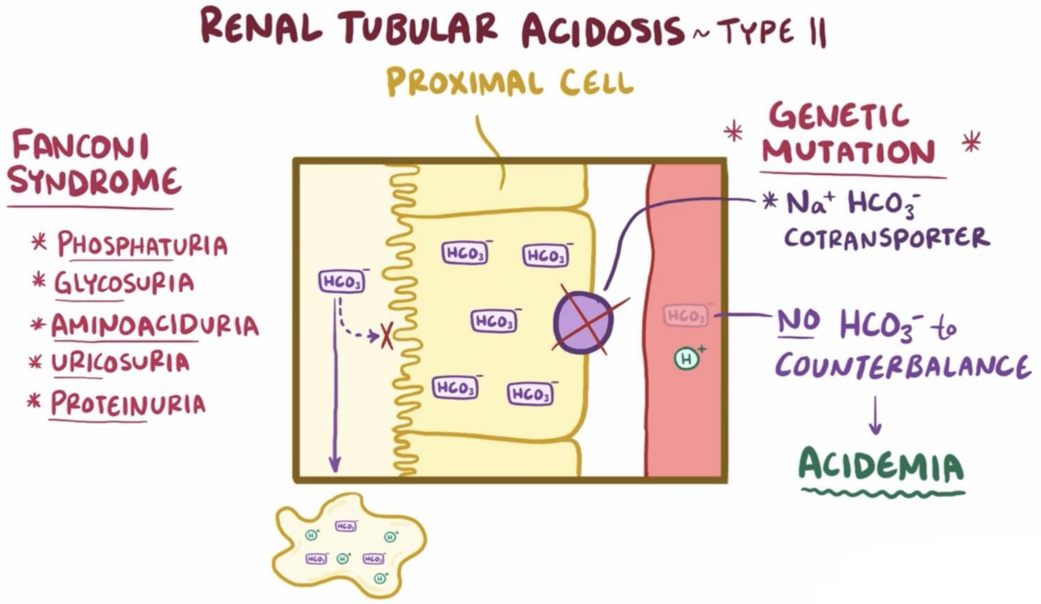


Fig. 4. Differential diagnosis of the causes of metabolic acidosis. GFR glomerular filtration rate

Types of RTA


RENAL TUBULAR ACIDOSIS
AFFECTS THE RENAL TUBULES AND RESULTS IN A HYPERCHLOREMIC METABOLIC ACIDOSIS WITH A NORMAL SERUM ANION GAP
TYPE 2
PROXIMAL RTA
IMPAIRED BICARBONATE REABSORPTION
HIGH URINE pH INITIALLY, LATER < 5.5
HYPOKALEMIA
TYPE 1 DISTAL RTA
IMPAIRED HYDROGEN ION SECRETION
URINE pH > 5.5
HYPOKALEMIA
RENAL STONES
TYPE 4 HYPERKALEMIC RTA
DECREASED ALDOSTERONE SECRETION OR ALDOSTERONE RESISTANCE
URINE pH < 5.5
HYPERKALEMIA
BOWMAN'S CAPSULE
PROXIMAL TUBULE
LOOP OF HENLE
DISTAL TUBULE
COLLECTING DUCT

Proximal RTA

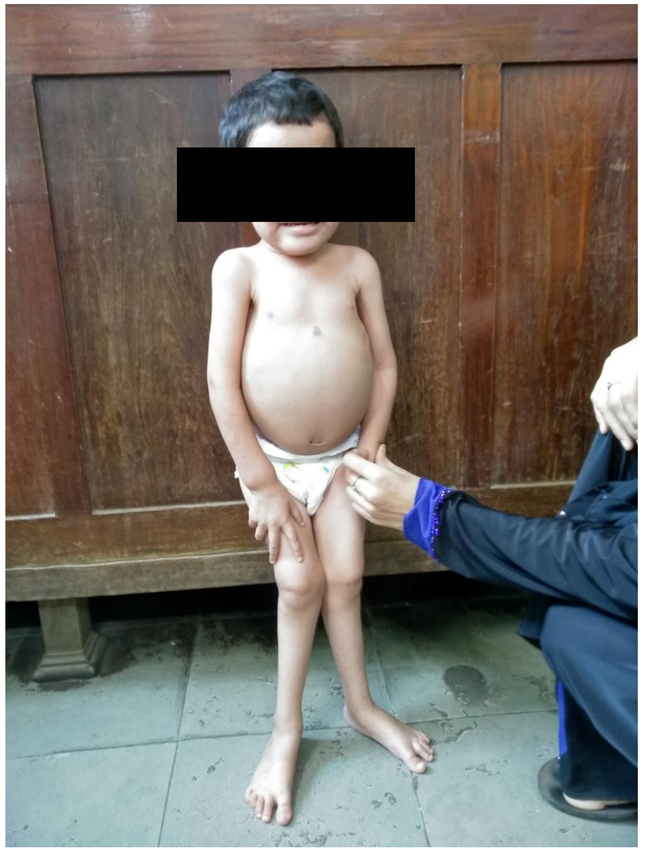


- Inability to reabsorb bicarbonate at PCT
- Distal acidification mechanism are intact
- Isolated HCO3 Defect or
- Generalised PCT dysfunction(Fanconi syndrome) (more common)

* Genetic(Cystinosis, Lowe syndrome, tyrosinemia)
* Acquired (Acetazolamide, ifosamide, Vitamin D deficiency)

Clinical features

- Growth retardation, failure to thrive
- Polyuria, polydipsia
- Night time awakening for water
- Recurrent episodes of fever, dehydration, vomitting
- Craving of salt
- Irritability, constipation
- Rickets, bone pain



On evaluation

- Ask for **positive family history of mental retardation, ESRD, failure to thrive, infant deaths or miscarriage**
- **Past History: recurrent diarrhoeal illness, episodes of fever**
- **Drug history**
- **On examination::**

Anthropometry

Dysmorphic features

Assessment of volume status

Investigations

Basic

- CBC
- Venous blood gas
- Serum electrolytes with Chloride
- Serum calcium, phosphorus, alkaline phosphatase
- BUN, serum creatinine
- Urine routine
- Urine calcium : creatinine ratio

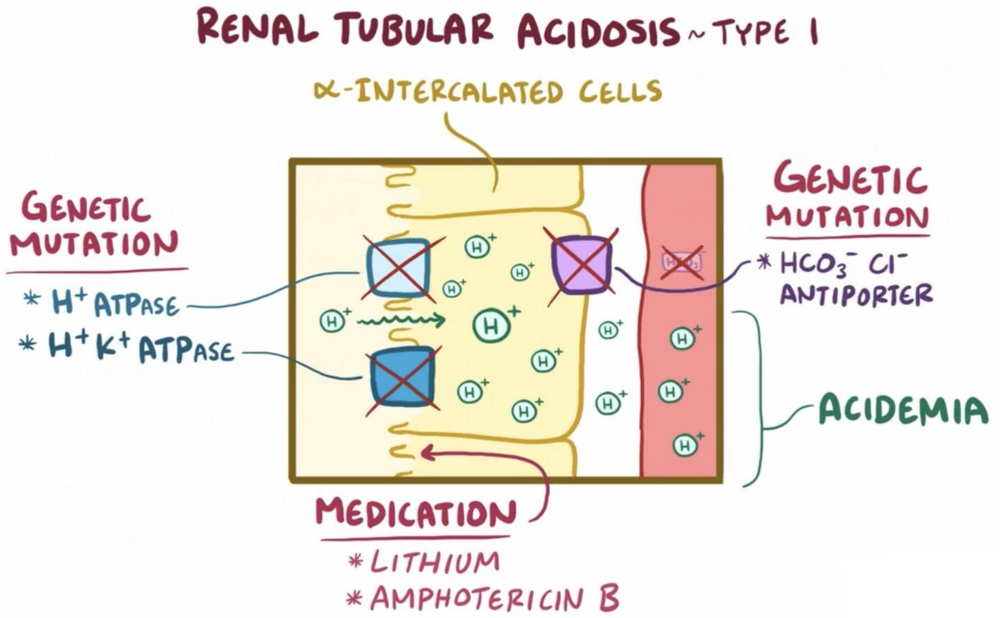
Specific investigations

- Urine pH
- Urine Anion gap(Urine Na+K)— Urine Cl-
- Fractional excretion of HCO3- (>15%)
- Tubular reabsorption of phosphorus
- (Urine-blood) pCO2

Treatment

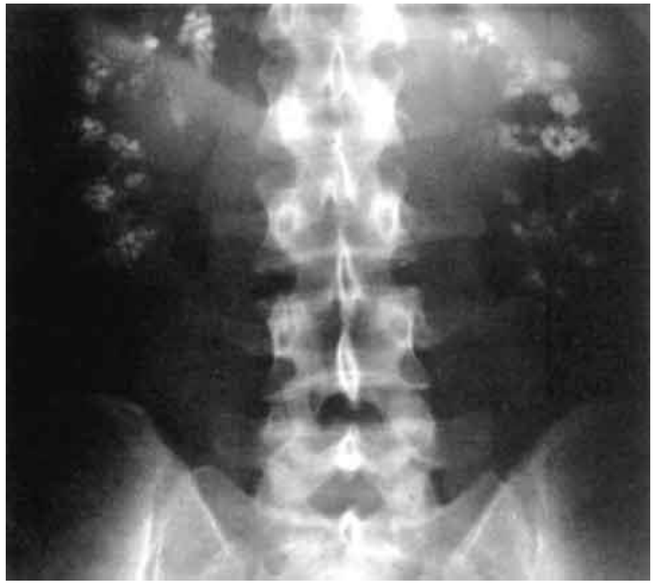
- May require up to 20meq/kg of alkali to compensate
- Phosphorus supplements
- Vitamin D supplements
- Thiazide diuretic
- Specific treatment for cystinosis, GSD diet, etc..

Distal RTA(type 1 RTA)



- Distal acidification defect
- Most common
- Genetic or acquired
- Acquired may be secondary to hyperparathyroidism, vitamin D intoxication, drugs like amphotericin B etc..

Clinical features and evaluation



- Similar to proximal RTA plus history of stones
- Hypercalciuria, nephrolithiasis on USG KUB
- No evidence of phosphorus or bicarbonate wasting
- Urine pH cannot be <5.3

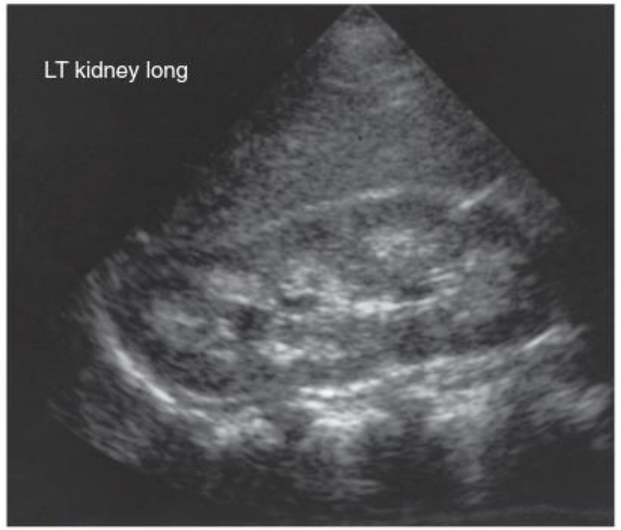


Fig. 547.2 Ultrasound examination of a child with distal RTA, demonstrating medullary nephrocalcinosis.

Treatment

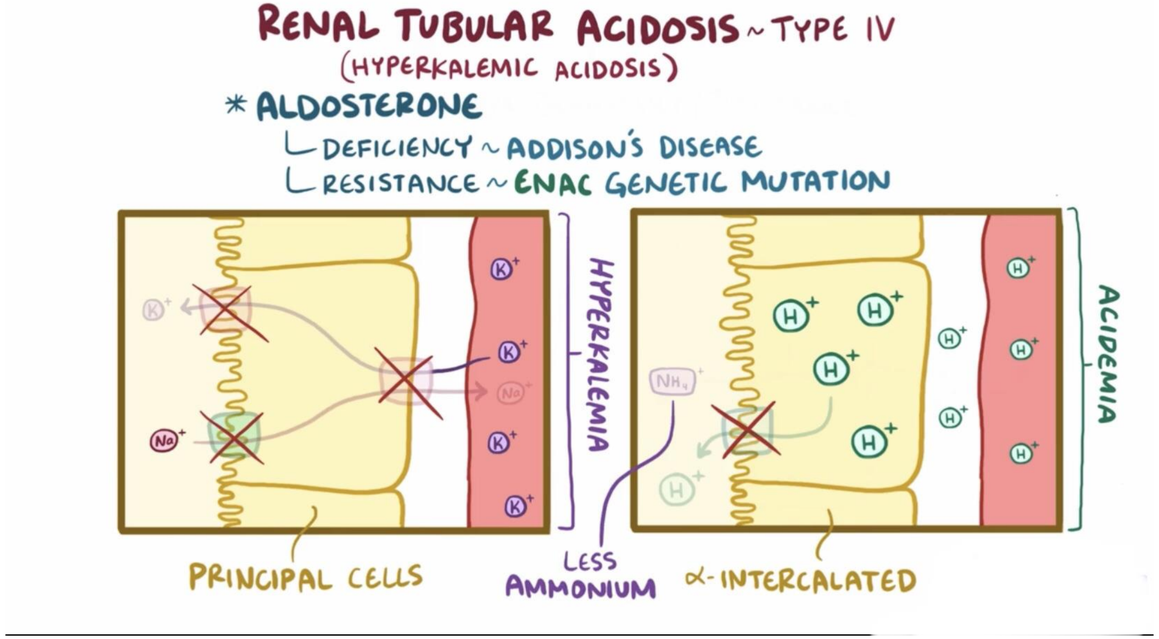
- Alkali as 2-4 meq per kg per day
- SOS potassium supplements
- Vitamin D supplements
- Withdrawal of causative agent if any

Type 4 RTA

Aldosterone mediated

1. Either due to reduced production of aldosterone(eg. Addison disease)
2. Or due to Aldosterone resistance eg.

- Pseudohypoaldosternism
- Obstructive uropathy, immediately due to acute pyelonephritis
- Drugs(Spironolactone, amiloride, tacrolimus)



Type 4 RTA

- Hyperkalemic HCMA
- Urine pH alkaline or acidic
- Elevated urine sodium with low potassium levels

★Treatment

- K binding agents
- Fludrocortisone

Finding	Proximal RTA	Classic distal RTA	Hyperkalemic RTAs
Serum [K^+]	↓	↓	↑
Urine pH with severe acidosis	<5.5	>5.5	<5.5 (aldosterone deficiency) >5.5 (voltage gradient defect)
Urine acidification	N	↓	↓
NH_4^+ excretion	N	↓	↓
U_{AG}	+(or −)	+	+
Fanconi syndrome	Yes	No	No
U-B pCO_2	Normal	↓	↓
Hypercalciuria	No	Yes	No
Nephrocalcinosis/lithiasis	No	Yes	No
Citrate excretion	N	↓	N
Bone lesions	Yes	Yes	No
Renal insufficiency	No	No	Yes
Response to alkali therapy	Less	Good	Variable

N normal, ↑ increase, ↓ decrease

Type 3 RTA(Guibaud Vainsel syndrome)

- Mutation in CA II enzyme leading to both proximal and distal RTA feature
- Autosomal recessive
- Osteopetrosis, cerebral calcification, development delay, nephrocalcinosis
- Treatment- bone marrow transplant, alkali supplements

Take home message

- One needs to rule out diarrhoeal illness before evaluating for RTA
- Proximal RTA is associated with Fanconi syndrome
- Family history must be enquired in every patient
- Renal stones and hypercalciuria points towards distal RTA
- One needs to rule out hyperkalemic RTA in patient with obstructive uropathy with persistent hyperkalemia
- Rickets resistant to Vitamin D therapy needs evaluation to rule out RTA


THANK
YOU
(...
JALPA
7/12/20