CONDITION

Distal renal tubular acidosis

Distal renal tubular acidosis is a condition in which the kidneys cannot remove enough acid from the blood.

WHAT IT IS
Impaired acid removal by the kidneys

BODY SYSTEM
Kidneys and urinary system

MAIN SYMPTOMS
Tiredness, muscle weakness, kidney stones, poor growth

WHAT CAUSES IT
Inherited changes, autoimmune conditions, medicines, kidney disorders

HOW IT IS DIAGNOSED
Blood bicarbonate and urine acidification tests

MAIN TREATMENT
Alkali replacement and treatment of the cause

CAN IT BE CURED?
Sometimes - acquired causes may reverse

IS IT AN EMERGENCY?
Sometimes - paralysis, fainting, or abnormal heartbeat

LONG-TERM OUTLOOK
Usually manageable with lifelong treatment and monitoring

Overview

Distal renal tubular acidosis, shortened to distal RTA or dRTA, affects how the kidneys control the body's acid balance.

Normal body processes continually produce acids. The kidneys usually remove this acid through urine while returning bicarbonate, an alkaline substance, to the blood. In distal RTA, the final part of the kidney tubules cannot make urine acidic enough. Acid remains in the blood, causing metabolic acidosis , which means the blood has too much acid or too little bicarbonate.

Distal RTA is also called type 1 RTA. It differs from type 2 RTA, which involves bicarbonate loss earlier in the kidney tubules, and type 4 RTA, which is usually associated with high potassium.

Long-term acid imbalance can affect muscles, bones, growth, and kidney health. Calcium

deposits and kidney stones may develop, and potassium can fall too low. Distal RTA may be inherited or caused by another condition or medicine. Treatment usually replaces alkali with citrate or bicarbonate and addresses the cause when possible.

Symptoms

Symptoms depend on age, the severity of the imbalance, and whether complications have developed. Mild or incomplete distal RTA may be found during tests for kidney stones or calcium deposits.

Adults may have:

Low potassium can cause constipation, marked weakness, or an abnormal heartbeat. A sudden severe fall can rarely make it difficult to move the arms or legs.

Babies and children may have poor feeding, vomiting, slow weight gain, or delayed growth. Ongoing acidosis can impair bone mineralisation and cause rickets, in which growing bones become softer and weaker.

Some inherited forms are associated with hearing loss beginning in infancy, childhood, or later life. Not everyone with inherited distal RTA develops it.

Symptoms alone cannot confirm distal RTA because tiredness, weakness, vomiting, and stones have many possible causes.

- Tiredness or reduced stamina
- Muscle weakness, cramps, or aching
- Frequent or painful kidney stones
- Pain in the side or back
- Increased thirst or urination
- Bone pain or fractures in advanced cases

Causes and risk factors

Inherited distal RTA results from changes in genes needed for acid transport in the kidney tubules. It may be inherited from one or both parents, depending on the gene involved.

Inherited forms often appear in infancy or childhood but may be recognised later. A family history of distal RTA, unexplained stones, kidney calcium deposits, poor childhood growth, or early hearing loss may increase suspicion.

Acquired distal RTA develops when another condition or exposure disrupts acid handling. Causes include:

A medicine associated with distal RTA does not always cause it. Do not stop prescribed medicine without medical advice, because sudden withdrawal may cause harm.

Sometimes no clear cause is found.

- Autoimmune conditions, especially Sjögren syndrome and sometimes lupus
- Certain medicines, including amphotericin B and lithium
- Chronic disorders affecting the kidney tubules
- Urinary tract obstruction
- Some disorders involving abnormal calcium levels or blood proteins

Diagnosis

Diagnosis begins with blood and urine tests. Blood results may show low bicarbonate, increased chloride, and sometimes low potassium. This pattern is called normal anion gap metabolic acidosis . The anion gap is a calculation used to distinguish causes of excess acid.

Doctors must exclude other causes. Diarrhoea, for example, can produce a similar pattern by removing bicarbonate from the body.

Urine tests assess whether the kidneys are responding properly. When blood is acidic, healthy kidneys should produce acidic urine and remove acid as ammonium. Urine that remains inappropriately alkaline supports distal RTA, but urine pH alone cannot confirm it. Infection, recent meals, medicines, and other factors can also raise urine pH.

Urine measurements may include pH, electrolytes, calcium, citrate, and estimates of ammonium removal. If results are unclear, a supervised acidification test may check whether the kidneys can lower urine pH. These tests require clinical monitoring and are not home tests.

Further assessment may include:

In incomplete distal RTA , blood bicarbonate is usually normal, but the kidneys still cannot fully acidify urine. This may contribute to recurrent stones or calcium deposits and often requires specialist testing.

- Kidney imaging to look for stones or calcium deposits
- Kidney function tests
- Tests for autoimmune or other underlying conditions

- Hearing tests when an inherited form is suspected
- Genetic testing for early-onset, familial, or unexplained cases

Treatment and management

Treatment aims to correct acidosis, maintain a safe potassium level, protect the bones and kidneys, and support children's growth.

The main treatment is oral alkali replacement. This provides bicarbonate directly or citrate, which the body converts into bicarbonate. Potassium-containing alkali may help when potassium is low. The medicine and dose depend on age, blood results, kidney function, stone risk, and other health needs.

Repeat blood and urine tests guide treatment. Too little alkali leaves acidosis untreated, while too much can cause another electrolyte imbalance. Use products as prescribed rather than adjusting them according to symptoms alone.

Treating an underlying condition may improve acquired distal RTA. A suspected medicine may be changed or stopped under medical supervision. Inherited forms usually require long-term or lifelong alkali treatment.

Regular monitoring may include:

People who form stones may be advised to drink enough fluid to keep urine dilute. Dietary changes should be based on stone-test results and overall health. Diet cannot replace prescribed alkali.

Severe acidosis or a dangerous potassium abnormality may require hospital treatment, with heart and blood-test monitoring while electrolytes are corrected.

- Bicarbonate, potassium, chloride, and kidney function
- Urine calcium, citrate, and other stone-risk measurements
- Kidney imaging when stones or calcium deposits are a concern
- Height, weight, and bone development in children
- Hearing assessment for relevant inherited forms

Complications

Untreated or undertreated distal RTA can cause calcium phosphate kidney stones. Calcium may also collect throughout kidney tissue, known as nephrocalcinosis . This differs from a

single stone and may remain visible on scans after blood chemistry improves.

Repeated stones, nephrocalcinosis, or an underlying kidney disorder may reduce kidney function over time. Chronic kidney disease can occur but is not inevitable.

Long-term acidosis removes minerals from bone and impairs bone formation. Children may develop rickets, delayed growth, or short stature. Adults may develop osteomalacia, meaning soft or weakened bones, and have a greater fracture risk.

Severely low potassium can affect the muscles used for breathing or disrupt heart rhythm. These complications need urgent treatment.

Consistent correction of acidosis reduces many risks. Established hearing loss or kidney calcification may not reverse, so monitoring remains important even when blood results are controlled.

Prevention

Inherited distal RTA cannot currently be prevented. Genetic counselling can explain inheritance and testing options for relatives or future pregnancies.

Not every acquired case is preventable. When a medicine that may affect kidney tubules is necessary, planned blood and kidney monitoring may detect changes earlier. Do not avoid or stop an important medicine solely because distal RTA is listed as a possible adverse effect.

For people with distal RTA, taking treatment consistently and attending follow-up can help prevent acidosis, stones, bone problems, and growth difficulties. Adequate fluid intake may reduce stone risk unless fluids have been restricted for another medical reason.

Living with it and what to expect

Most people can continue school, work, exercise, and family life when acid and potassium levels are controlled. Management may involve medicine several times a day, repeat blood and urine tests, and periodic kidney scans.

Weakness and poor appetite often improve after the imbalance is corrected. Children may grow better, although catch-up growth depends partly on how early treatment starts.

Inherited distal RTA is usually lifelong. Treatment controls the chemical imbalance but does not remove the genetic cause. Alkali needs may change with growth, illness, pregnancy, kidney function, diet, or other medicines.

Acquired distal RTA may improve if its cause is controlled or a responsible medicine can be

withdrawn. Persistent tubular damage may still require long-term treatment.

Stones can recur if urine chemistry is not fully corrected. Hearing loss associated with inherited distal RTA may progress separately and require specialist support.

Vomiting, diarrhoea, or difficulty taking medicine can disrupt acid and potassium control. Have a plan for illness and request advice if prescribed alkali cannot be kept down.

When to get medical help

Attend routine follow-up for blood tests, prescription reviews, growth checks, hearing monitoring, and kidney imaging. Seek an earlier appointment if tiredness or muscle symptoms return, stones become more frequent, or treatment is difficult to take.

Ask for prompt medical advice if you have persistent vomiting or diarrhoea, cannot keep alkali treatment down, or notice a clear reduction in urination.

Flank pain with fever, chills, or painful urination needs urgent assessment because it may indicate a urinary infection or an obstructed, infected kidney.

Seek emergency help for:

These symptoms can result from a major acid or potassium disturbance or another serious condition.

- Sudden severe weakness or inability to move normally
- Fainting, severe palpitations, or chest pain
- New confusion, a seizure, or unusual difficulty staying awake
- Severe breathing difficulty

Common questions

Is distal RTA the same as kidney failure?

No. Distal RTA is a problem with acid removal by kidney tubules. The kidneys may still filter waste at a near-normal rate. However, stones, calcium deposits, or an underlying kidney condition can reduce function over time.

Can distal renal tubular acidosis be cured?

Inherited distal RTA cannot currently be cured, but treatment usually controls the imbalance. Acquired distal RTA may improve if its cause is removed or treated, although some people need long-term alkali replacement.

What should I eat if I have distal RTA?

There is no single distal RTA diet. Enough fluid may help prevent stones, while some people need advice about sodium, potassium, calcium, or citrate. Restrictive or "alkaline" diets do not replace prescribed treatment. Advice should reflect blood results, kidney function, and stone type.

Will a child with distal RTA grow normally?

Correcting acidosis supports growth and bone development. Many children grow better after treatment starts, but final height depends on the cause, age at diagnosis, treatment consistency, nutrition, and other conditions.

Can I exercise with distal RTA?

Exercise is usually possible when bicarbonate and potassium are controlled. Stay hydrated, especially during heat or prolonged activity. Stop and seek assessment for unusual weakness, severe cramps, faintness, chest symptoms, or palpitations.

Do my relatives need genetic testing?

Testing may be useful when distal RTA began early, affects multiple relatives, or occurs with hearing loss. A genetics service can explain who may benefit and what results could mean.

Sources

- National Institute of Diabetes and Digestive and Kidney Diseases: Renal Tubular Acidosis
- Merck Manual Professional Edition: Renal Tubular Acidosis
- GeneReviews: Hereditary Distal Renal Tubular Acidosis
- ERKNet and ESPN: Distal renal tubular acidosis clinical practice points, Nephrology Dialysis Transplantation
- Palmer BF, Kelepouris E, Clegg DJ. Renal Tubular Acidosis and Management Strategies: A Narrative Review. Advances in Therapy, 2021