NEPHROLOGY - I

RENAL TUBULAR ACIDOSIS - APPROACH AND MANAGEMENT

***Mohamed Azarudeen J**
****Mehul A Shah**

Abstract: *Renal tubular acidosis is a group of inherited and acquired tubular disorders presenting as hyperchloremic normal anion gap metabolic acidosis in the presence of normal kidney function. The four phenotypes include: Type 1- distal renal tubular acidosis, Type 2- proximal renal tubular acidosis (isolated or renal Fanconi syndrome), Type 3- combined proximal and distal tubular dysfunction and Type 4- aldosterone deficiency / resistance. Clinical presentation includes polyuria, polydipsia, failure to thrive and occasionally, refractory rickets and evaluation for renal tubular functions and genetic studies are essential for making the diagnosis. Management includes supplementation of alkali, potassium, phosphorus and active vitamin D3 and specific therapy for identified etiology. Early diagnosis and management improve long term outcomes.*

Keywords: *RTA, Renal tubular acidosis, Renal Fanconi syndrome, Normal anion gap metabolic acidosis, Renal rickets.*

* Department of Pediatrics
Velammal Medical College Hospital and Research Institute,
Madurai.

** Consultant Pediatric Nephrologist,
Little Stars & She Hospital and Apollo Health City,
Hyderabad.

email: mehulashah@hotmail.com

Points to Remember

- ***RTA is characterised by hyperchloremic normal anion gap metabolic acidosis.***
- ***Presenting features include failure to thrive, polyuria and polydipsia.***
- ***Proximal RTA is characterised by impaired proximal tubule bicarbonate reabsorption and distal RTA due to impaired distal hydrogen secretion.***
- ***The complete normalization of plasma bicarbonate levels with alkali supplementation suggests a diagnosis of dRTA.***
- ***Early diagnosis, adequate alkali supplementation and regular long-term follow-up with monitoring of growth, bony deformities and renal function is required.***

References

1. Bockenhauer D, Lopez-Garcia SC, Walsh SB. Renal tubular acidosis. In: Pediatric Nephrology. Emma F, Goldstein SL, Bagga A, Bates CM, Shroff R (eds). Eighth Edition. Springer International Publishing, Cham. 2022; pp 903-928.
2. Palmer BF, Kelepouris E, Clegg DJ. Renal Tubular Acidosis and Management Strategies: A Narrative Review. Adv Ther. 2021 Feb;38(2):949-968. doi: 10.1007/s12325-020-01587-5.
3. Quigley R, Wolf MT. Renal tubular acidosis in children. In: Pediatric Nephrology. Emma F, Goldstein SL, Bagga A, Bates CM, Shroff R (eds). Seventh Edition Springer- Berlin Heidelberg. 2020; pp1273-306.
4. Soleimani M, Rastegar A. Pathophysiology of Renal Tubular Acidosis: Core Curriculum 2016. Am J Kidney Dis. 2016 Sep;68(3):488-98. doi: 10.1053/j.ajkd.2016.03.422.
5. Finer G, Landau D. Clinical Approach to Proximal Renal Tubular Acidosis in Children. Adv Chronic Kidney Dis. 2018 Jul;25(4):351-357. doi: 10.1053/j.ackd.2018.05.006.
6. Giglio S, Montini G, Trepiccione F, Gambaro G, Emma F. Distal renal tubular acidosis: a systematic approach from

Cite as: Mohamed Azarudeen J, Mehul A Shah, Renal tubular acidosis - Approach and management, Indian J Pract Pediatr. 2024; 26(3):226-235.

diagnosis to treatment. J Nephrol. 2021 Dec;34(6): 2073-2083. doi: 10.1007/s40620-021-01032-y.

7. Wagner CA, Devuyst O, Bourgeois S, Mohebbi N. Regulated acid-base transport in the collecting duct. Pflugers Arch. 2009 May;458(1):137-56. doi: 10.1007/s00424-009-0657-z.
8. Foreman JW. Fanconi Syndrome. Pediatr Clin North Am. 2019 Feb; 66(1):159-167. doi: 10.1016/j.pcl.2018.09.002.
9. Trepiccione F, Walsh SB, Ariceta G, Boyer O, Emma F, Camilla R, et al. Distal renal tubular acidosis: ERKNet/ESPN clinical practice points. Nephrol Dial Transplant. 2021 Aug 27; 36(9):1585-1596. doi: 10.1093/ndt/gfab171.
10. Alexander RT, Law L, Gil-Peña H, Greenbaum LA, Santos F. Hereditary Distal Renal Tubular Acidosis. In: Adam MP, Feldman J, Mirzaa GM, Pagon RA, Wallace SE, Amemiya A, editors. GeneReviews® [Internet]. Seattle (WA): University of Washington, Seattle. 2019; pp 1993-2024.
11. Dawman L, Tiewsoh K, Barman P, Pratyusha K, Chaakchhuak L, Sharawat IK. Phenotype and Genotype Profile of Children with Primary Distal Renal Tubular Acidosis: A 10-Year Experience from a North Indian Teaching Institute. J Pediatr Genet. 2021 Mar 3;11(3):221-226. doi: 10.1055/s-0041-1724114.
12. Tajima T, Morikawa S, Nakamura A. Clinical features and molecular basis of pseudohypoaldosteronism type 1. Clin Pediatr Endocrinol. 2017;26(3):109-117. doi: 10.1297/cpe.26.109. Epub 2017 Jul 27.
13. Ellison DH. Pseudohypoaldosteronism Type II. In: Adam MP, Feldman J, Mirzaa GM, et al., editors. GeneReviews® [Internet]. Seattle (WA): University of Washington, Seattle. 2011 [Updated 2017]; pp 1993-2024.
14. Bökenkamp A, Ludwig M. The oculocerebrorenal syndrome of Lowe: an update. Pediatr Nephrol. 2016 Dec; 31(12):2201-2212. doi:10.1007/s00467-016-3343-3.
15. Jin YY, Huang LM, Quan XF, Mao JH. Dent disease: classification, heterogeneity and diagnosis. World J Pediatr. 2021 Feb;17(1):52-57. doi: 10.1007/s12519-020-00357-1.
16. Bagga A, Sinha A. Renal Tubular Acidosis. Indian J Pediatr. 2020 Sep; 87(9):733-744. doi: 10.1007/s12098-020-03318-8.
17. Fuster DG, Moe OW. Incomplete Distal Renal Tubular Acidosis and Kidney Stones. Adv Chronic Kidney Dis. 2018 Jul;25(4):366-374. doi: 10.1053/j.ackd.2018.05. 007.
18. Uduman J, Yee J. Pseudo-Renal Tubular Acidosis: Conditions Mimicking Renal Tubular Acidosis. Adv Chronic Kidney Dis. 2018 Jul; 25(4):358-365. doi: 10.1053/j.ackd.2018.05.001.
19. Noone D, Langlois V. Laboratory evaluation of renal disease in childhood. Pediatric kidney disease. 2023 Apr 7:37-68
20. Goodyer P, Phadke K. Evaluation of renal disease. In : Manual of Pediatric Nephrology. Berlin, Heidelberg: Springer Berlin Heidelberg, 2013 pp1-64.
21. Alexander RT, Bockenhauer D. Renal Tubular Acidosis. In: Schaefer F, Greenbaum LA, editors. Pediatric Kidney Disease [Internet]. Cham: Springer International Publishing; 2023. p. 1071–93.
22. Kiran BV, Barman H, Iyengar A. Clinical profile and outcome of renal tubular disorders in children: A single center experience. Indian J Nephrol. 2014 Nov; 24(6): 362-6. doi: 10.4103/0971-4065.133002.
23. Küçükali GK, Çetinkaya S, Tunç G, Oðuz MM, Çelik N, Akkaþ KY et al. Clinical Management in Systemic Type Pseudohypoaldosteronism Due to SCNN1B Variant and Literature Review J Clin Res Pediatr Endocrinol. 2021; 13(4):446-51.