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Medullary sponge kidney presenting with hypokalaemic paralysis; JArticle Postgraduate Medical Journal 1984 Apr;60(702):303-4 Vol: 60 No.(702) 1984_.303-4pp

Abstract :Medullary sponge kidney associated with a defect in urine acidification is rare and usually of no clinical significance. We report a case presenting as hypokalaemic paralysis due to associated congenital distal renal tubular acidosis.