Urinary cyclic adenosine 3',5'-monophosphate response in McCune-Albright syndrome: Clinical evidence for altered renal adenylate cyclase activity

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The recent finding of an activating mutation in the Gs ? protein, the protein that couples receptors to stimulation of adenylate cyclase, from endocrine and nonendocrine tissues of patients with McCune-Albright syndrome (MAS) suggests that alterations in adenylate cyclase activity may account for the clinical abnormalities in these patients. Many patients with MAS have hypophosphatemia. This may result from the presence of the activating Gs ? mutation in proximal renal tubules or the elaboration of a phosphaturic factor from fibrous dysplasia. We, therefore, sought to characterize renal cAMP generation and phosphate handling in MAS patients. Intravenous infusion of PTH is a classic clinical test used to evaluate hormonal responsiveness of renal proximal tubule adenylate cyclase and examine PTH-dependent phosphate clearance. We performed PTH infusion in 6 MAS patients, 10 normal subjects, and 6 patients with pseudohypoparathyroidism (PHP). The basal urinary cAMP (UcAMP) level in the MAS