

¹To be performed at initial presentation in all girls suspected of having MAS, regardless of clinical symptoms. ²Gonadotropins should be suppressed in MAS-associated PP, unless autonomous estrogen production has induced central PP (typically occurs when bone age reaches approximately 11 years). ³Estrogen production in MAS-associated PP is intermittent, and undetectable levels do not rule out disease. ⁴Ovarian cysts are suggestive of MAS-associated PP, however absence of cysts does not rule out disease. ⁵In girls presenting with isolated peripheral PP, the differential also includes estrogen-producing tumors. Work-up for additional features of MAS may establish the diagnosis. ⁶Unlike other features of MAS, autonomous ovarian activity may present at any time during infancy or childhood. Girls should continue to be monitored clinically for signs of peripheral PP, however routine labwork and imaging is not recommended. ⁷Patients may rarely present with intermittent ovarian activity with only subtle signs of estrogenization (mild intermittent breast development without vaginal bleeding), which may not be appreciated by families and practitioners. ⁸Hyperthyroidism and GH excess may present with bone age advancement.

References

Collins MT, Singer FR, Eugster E. McCune-Albright syndrome and the extraskeletal manifestations of fibrous dysplasia. 2012. Orphanet J Rare Dis. Foster CM, Feuillan P, Padmanabhan V, Pescovitz Oh, Beitins IZ, Comite F, Shawker TH, Loriaux DI, Cutler GB Jr. Ovarian function in girls with McCune-Albright syndrome. Pediatr Res. 1986.

Legend

SH = follicle stimulating hormone; GH = growth hormone; H&P = history & physical exam; MAS = McCune-Albright syndrome; PP = precocious puberty; US = ultrasound