Pituitary gigantism and central precocious puberty presenting with prognathism in a pediatric patient

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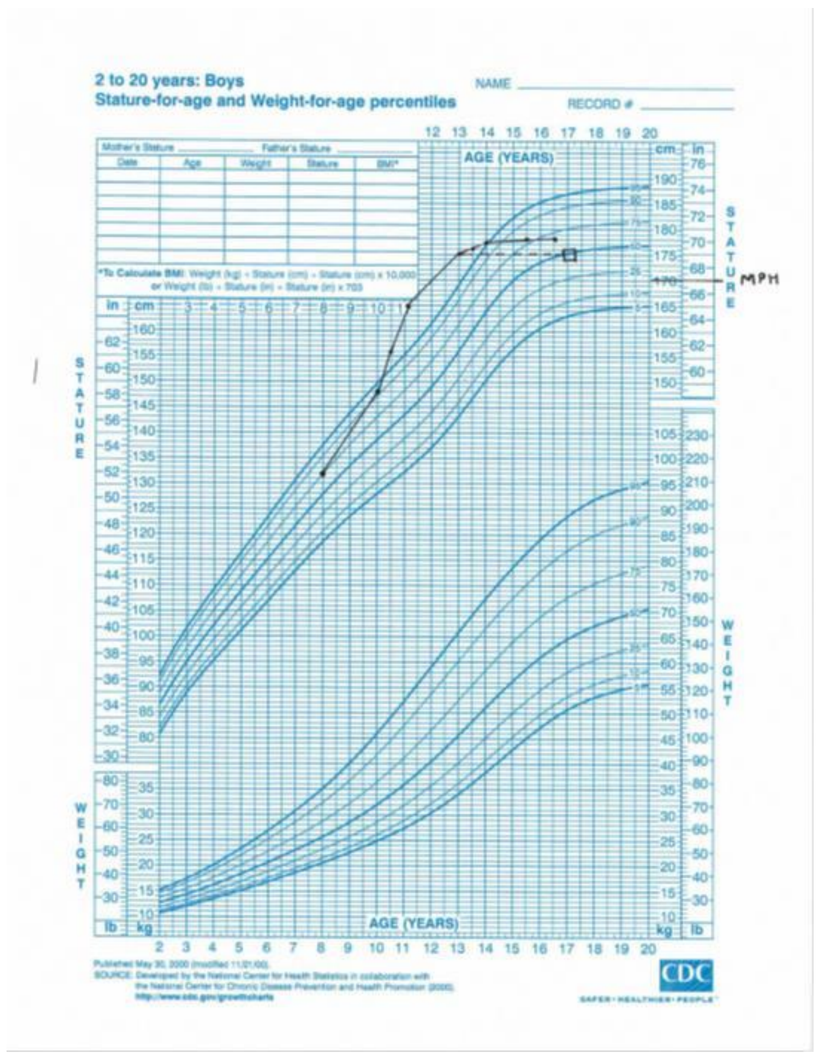
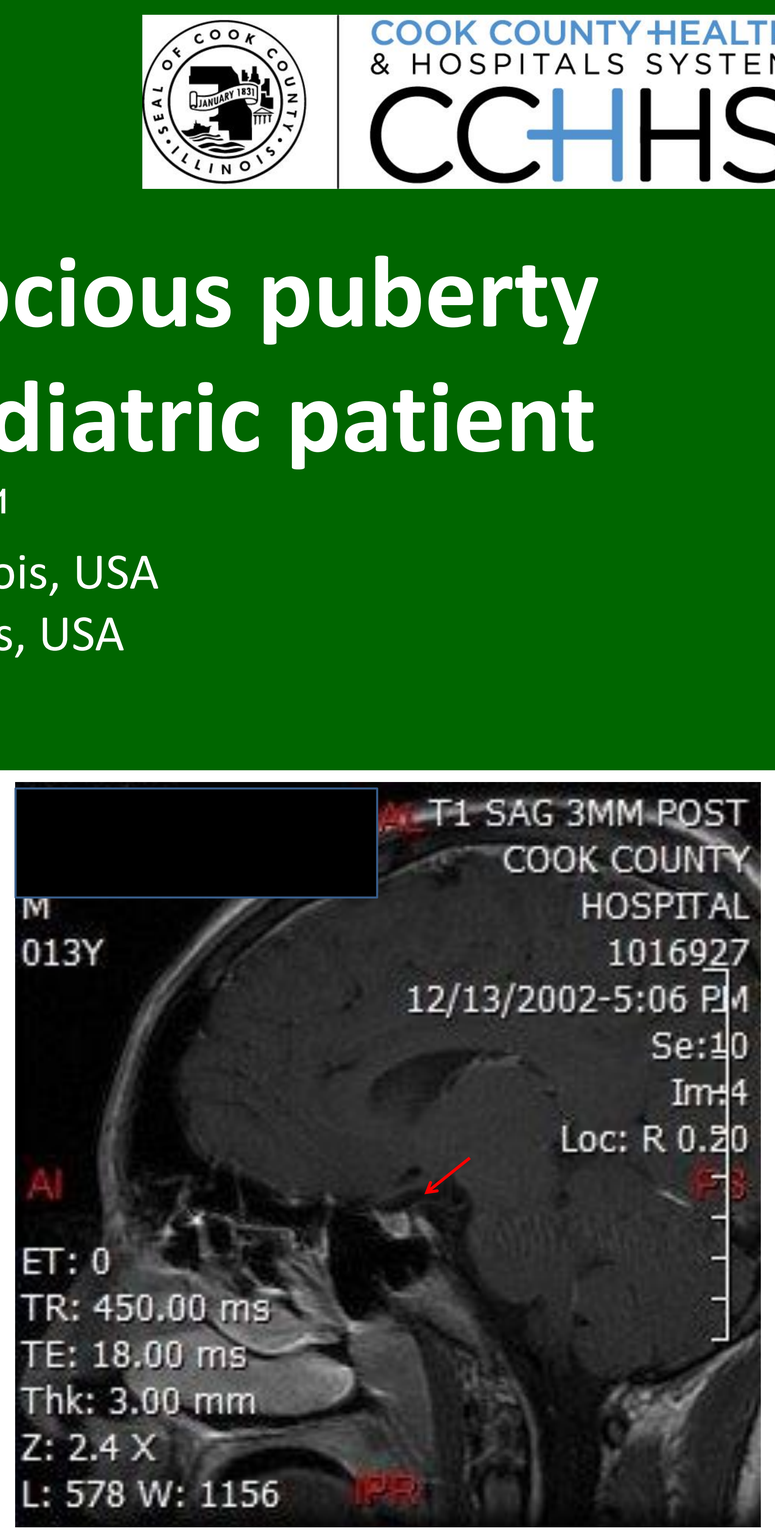
Case Report:

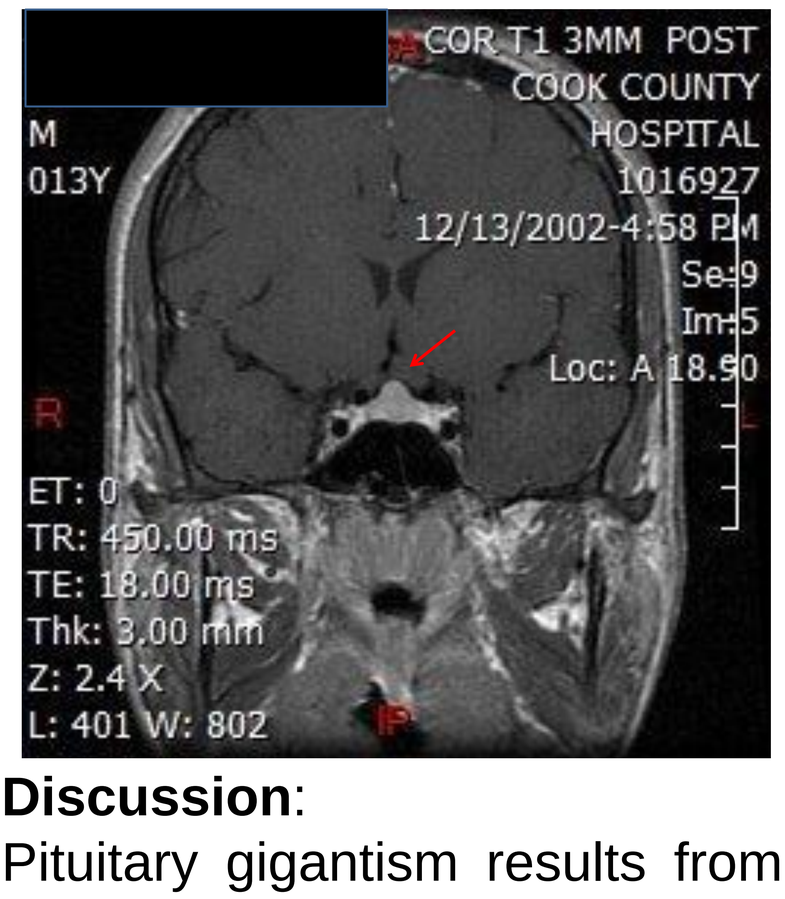
A 13-year old male presented to a dental office for evaluation of prognathism. After evaluation, his dentist referred him to pediatric endocrinology. No significant past medical history. He denied any signs or symptoms associated with any hormonal deficit or excess, as well as headaches or visual disturbances. Denied excessive growth of hands or feet. Parents reported appearance of pubic and axillary hair, mild acne, and significant growth over the last few years. On exam, he was found to be above the 95th centile for height (mid-parental height around the 20th centile). Genitalia and pubic hair were both were Tanner stage IV. Previous growth records revealed a growth acceleration, starting at around 8 years of age, and that he had crossed height percentiles from the 75th to above the 95th from age 8 to age 11 years old. Bone age was read at 17 years, at a chronological age of 13 years (>3SD above the mean). Hormonal evaluation revealed growth hormone (GH) levels of 7.3 ng/mL (0-5.0) and IGF-1 levels of 725 ng/mL (upper limit of normal for his Tanner stage). Thyroid function tests, morning cortisol and prolactin were normal. FSH, LH and testosterone were late pubertal.

GH suppression test was abnormal, and confirmed the suspected diagnosis of GH excess.



MRI with contrast revealed a pituitary macroadenoma. The patient had a transsphenoidal resection of the macroadenoma. On follow up he reported, no further mandible growth, his growth was normal as were all his pituitary hormones. He did not need any hormonal replacement. Post-surgical MRI showed no evidence of residual mass. He was referred for jaw reconstruction.





Discussion:

Pituitary gigantism results from persistent increased secretion of GH and it occurs before fusion of the epiphyseal growth plates, hence it will have an effect on stature. Clinical manifestations may include increased growth velocity and occasionally growth of hands and feet, frontal bossing and prognathism. Our patient had a component of central early puberty as well, related to his macroadenoma, which resulted in premature epiphyseal closure.

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Syndromes : Mechanisms and Management

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