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Migeon et al (1968) described an entity of adrenal unresponsiveness to ACTH characterized by hypoglycemia,hyperpigmentation normal tolerance to salt deprivation and no elevation of plasma cortisol with ACTH. The possibility of X-linked and autosomal forms suggests heterogeneity in this rare condition. Two affected brothers, offspring of first-cousin parents are presented. The first case died at the age of three years of "unknown cause" with a history of intermittent hypoglycemia precipitated by the stress of infections. Post-mortem examination revealed marked hyperpigmentation, cerebral edema, small foci of calcifications scattered in the arteriolar wall. The adrenals were hypoplastic showing degeneration of the zona fasciculata and reticularis, with preservation of the zona glomerulosa. The second son started with similar signs and symptoms at the age of two years and also showed hyperpigmentation when he was admitted to the Pediatric Service of the University of Brasilia, six months later. Laboratory investigation revealed 28mg% glucose, 136mEq/l sodium, 4,1mEq/l potassium,calcium, phosphorus, uric acid, alkaline phosphatase, colesterol, triglycerides were within normal range. Endocrine testing showed normal HGH, PRL, T3, T4, TSH responses. The cortisol levels were undetectable even after six hours IV ACTH administration. The patient was put in a regular diet with prednisone 2,5mg twice daily and is quite well on this regimen.

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PROLACTINOMA IN PEDIATRIC AGE: A LONGITUDINAL FOLLOW-UP
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Approximately 10-20% of the prolactinomas are recognized before age 20. However, there are few reports on their medium and long term follow-up. Thirteen patients (8♀, 9♂) aged 14,2 - 20 yr. ($\bar{X}$ 16,5 yr) with prolactinoma were followed for 9 m-12 yr. Headaches, impairment of pubertal development, galactorrhea and visual field defects were the most common clinical findings. The responses of LH and FSH to LH-RH among patients varied from normal, exaggerated or absent. THe TRH (TSH) test was normal in 8/9 cases and in 1♀ a primary concomitant hypothyroidism was reported. Serum PRL was elevated in all patients ranging 77-1150 ng/ml ($\bar{X}$ 378±285). PRL response after TRH was inadequate ($\bar{X}$ Δ MAX:25%). Radiologically, the patients were classified (Vezina and Sutton) in G1: n=3 (♀); G2: n=5 (4♀, 1♂); G3: n=3 (1♀, 2♂) and G4: n=2 (♂). Eight patients (4♀, 4♂) were initially treated with surgery, 7 of them (3♀, 4♂) with a persistent hyperprolactinemia required Bromocriptine (BEC) after surgery. In 5 cases (4♀, 1♂) BEC constituted the primary treatment, but 1♀ with failure to BEC therapy needed transsphenoidal adenomectomy. Follow-up was possible in 11 patients. Among those who underwent surgery, 1♀ recovered gonadal function spontaneously and 5 cases (3♀, 2♂) required complementary medical treatment. Among those treated initially with BEC 1♀ recovered menses spontaneously, 1♀ needed progesterone therapy, 1♀ did not achieve normal PRL values after 9 m of treatment, the adolescent who required surgery had normal PRL levels without menses and the ♂ recovered gonadal function. In conclusion: - Basal PRL levels seem to be the most relevant element in hte diagnosis of prolactinomas. - Clinical trials would suggest an increase in tumoral growth velocity in males. - In pediatric patients, BEC appears as a useful resource for primary or complementary treatment of prolactinomas.