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Responsiveness of normal and tumoral human adrenal cells to ACTH and prostaglandin E_1 (PGE_1) : Role of the cAMP-protein kinase system.

Isolated cells were prepared from normal human adrenal and adrenocortical secreting tumors, the latter were : A) sensitive to ACTH and PGE_1 ; B) ACTH insensitive, PGE_1 sensitive ; C) ACTH sensitive, PGE_1 insensitive ; D) ACTH and PGE_1 insensitive. *In vitro* production of cortisol (F) and cAMP and activation of cAMP-dependent protein kinase (PK) were measured. After 1 h incubation of normal adrenal and tumor A with increasing amounts of ACTH and PGE_1, PK activation and F production showed a good correlation but cAMP and F production did not at low hormonal concentration. ACTH was unable to stimulate PK and F of tumor B but it responded to PGE_1 and dibutyryl cAMP (DbcAMP). Tumor C responded to ACTH and DbcAMP but did not to PGE_1. Tumor D only responded to DbcAMP. The results implicated that the hormonal activation of PK and stimulation of steroidogenesis require very small changes of cAMP levels. The lack of such increase in tumors with abnormal membrane receptors would explain the insensitivity of these tumors to ACTH and PGE_1.

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Thyroid function in early infancy in relation to gestational age at birth.

We studied thyroid function longitudinally from 5 days to 6 weeks of age in 43 full-term (group A) and 54 premature infants with gestational ages at birth of 8 months (group B, 29 subjects) and 7 months (group C, 25 subjects). Formal consent was obtained from the parents. On day 5, serum levels of TSH and T_4 were within the normal range in all patients. Serum T_4, T_3 and free T_4 were significantly lower in group C than in groups A and B. TBG was similar in the three groups. In premature infants as compared to full-term infants, the response to TRH (50 µU IV) was markedly increased for TSH and decreased for T_3. The group differences progressively disappeared during the first 4 weeks of life. However, in group C, TRH-induced TSH responses remained higher than in groups A and B. The data reported evidence a transient state of compensated hypothyroidism in premature infants in our country.

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Effect of hGH therapy on the head circumference in hypopituitary children.

Head circumference was measured before and during hGH therapy in 14 children with IGHD (mean CA 6:10 and mean BA 1:6 yrs) and in 21 children with MPHD (mean CA 12:5 and mean BA 7:4 yrs). In both groups there was a retardation in the growth of the neurocranium, with a mean SD - 2.7 in IGHD and of -1.3 in MPHD. In IGHD, initiation of hGH therapy before the age 5 and BA of 3 led to a catch-up in cranial growth with normalization of the head size (m SD: -1.4). In older children the induction of head growth was similar to that achieved by the long bones but without catch-up. In 3 adults with hereditary IGHD without therapy, the head circumference was below normal (-2.3, -3.2 and -4.4 SD). In view of the possible influence of GH on the development of the brain and of the role played by the brain upon the growth of the cranium, the importance of early diagnosis and initiation of therapy in infants and young children with a deficiency of GH is stressed.

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Pituitary function in 8 patients with familial pituitary dwarfism.

Pituitary function was studied in four pairs of familial pituitary dwarfs (two sisters 28 and 20 years old, two brothers 15 and 11 years old, one sister aged 19 and her brother aged 6 and one sister aged 9 6/12 and her brother 3 6/12), by performing l-dopa test, TRH-test ,LHRH-test, sulpiride-test and by measuring basal concentrations of ACTH and Cortisol (F). It was found that all patients had abnormal responses of GH to l-dopa test, and of prolactin to sulpiride-test but there was an intra-and interfamilial variability as regards the response of TSH to TRH, of FSH and LH to LHRH and the basal levels of ACTH and F.

Read by title

75 T.STOICA*, M.COCULESCU*, X.LOGIN*, M.OPRESCU* V.DIMITRIU* (Intr.by C.Dacou-Voutetakis). Department of Endocrinology ,Faculty of Pediatrics,Bucharest, Romania. Congenital aqueductal stenosis (CAS) and empty sella syndrome(ESS) with two opposite patterns of gonadotropin secretion.

Both hydrocephalia interna and ESS may be associated with endocrine disturbances and enlarged sella turcica which erroneously suggest a pituitary tumour.Two pubertal girls were followed three years.Both presented ESS and CAS diagnosed by fractionated pneumotomoencephalography. A progressive increase of the pituitary fossa followed a brisk intracranial hypertension.One patient had spaniomenorrhea and a low level of serum RIA gonadotropins with a high response to LRH showing secondary pituitary defficiency.The other girl had accelerated puberty and serum RIA gonadotropins in the range of normal adult woman but a low level of plasma cortisol and a reduced response to metyrapone.It shows that the intracranial hypertension and chronic hydrocephalia interna may produce ESS and opposite effects on gonadotropins secretion.

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Growth pattern in constitutional short stature.

In a retrospective study of 263 short children 149 (56.7%) had constitutional short stature. Exaggerated growth deceleration first became apparent between 3 and 6 mos of age, was greatest in the first 2yrs of life, and resulted in these children falling more than two standard deviations below the mean for height by three years of age. After three years of age, the growth rate of children with constitutional short stature was the same as that of normal children and they ran parallel to the growth curve, below the third percentile. The importance of serial measurements is emphasized and recommendations are made for the evaluation of short stature or abnormal growth deceleration in the first three years of life. Awareness of this typical pattern of growth in healthy children may aid the pediatrician in his evaluation of short stature.