184 D.R. BROWN & J.T. MacDONALD. Children's Health Center & Hospital, Univ of Minnesota, Minneapolis.

Antidiuretic Hormone Changes with Acute Increased Intracranial pressure (ICP).

Clinical SIADH and DI are seen with changes of ICP in acute encephalopathies. Changes in serum ADH (RIS) were correlated with direct measurements of ICP with a Richmond screw transducer, in 9 patients with Reye's syndrome. Serum ADH measured by method of Skowsky, with bentonite extraction to remove vasopressin from specimen. Extracts were resuspended in phosphate buffer & measured by RIA, utilizing ^{125}I-AVP (2500 PM), 100 ul. (Normal ADH $\bar{x}$ + 2 SD = -.1 to 3.54 uIU/ml). All patients were in stage III or IV & had clinical & laboratory ADH alterations. ICP changes corresponded with serum ADH changes. Patients in stage I & II showed no clinical or laboratory ADH changes. SIADH is seen initially with serum ADH & ICP measurement rising together. Sustained increased ICP leads to DI, decreased serum ADH & a poor prognosis. Increased ICP may be associated with direct hypothalamic effects producing changes in ADH & appropriate correlated clinical syndrome.

	FIRST 72 HOURS ICP MONITORING	AFTER RECOVERY
$ADH/\{Na^+\}_S \times 100$	8 - 67	6 - 11
Mean	15	7
$\{ADH\}_S$	4.2 - 10.0 uIU/ml	0.5 - 2.2 uIU/ml

185 G.J. Bruining, B.M. de Jongh, Sophia Children's Hospital, Erasmus University Rotterdam and Dept. of Immunohematology University Medical Center Leiden, The Netherlands. Relation of HLA with insulin dependent diabetes (IDM) and multiple sclerosis (MS) in one family.

MS is associated with an increase, and IDM with a decrease of the frequency of HLA-DR2 in most Caucasoid populations. Thus, HLA-DR2 is conferring susceptibility to MS and resistance to IDM in unrelated individuals.

We could study the relationship of HLA with IDM and MS in one family with seven children. One sib had MS and three had IDM. We typed the family for the HLA-A, B, C and DR antigens, the polymorphisms of BF, C2 and GLO and screened the sera for various autoimmune antibodies.

Of the three sibs with IDM, two inherited the parental haplotypes b/d(DR4/DR4), and one a/d(DR5/DR4). The latter sib also had trisomy 21. The sib with MS inherited haplotypes b/c(DR4/Dr2).

The segregation of the HLA haplotypes in this family confirmed that:

1-DR4 is conferring susceptibility to IDM

2-DR2 is conferring resistance to IDM but susceptibility to MS.

186 L.CAVALLO, M.ALTOMARE, P.PALMIERI, D.LICCI, F.CARNIMEO and F.MASTRO (Intr.by G.Chiumello)

Institute of Child Health, University of Bari, Italy.

Endocrine function in anencephalic newborn infants.

A combined LHRH and TRH test was performed, followed by an IvGTT 1 hour later, in 2 female and 2 male anencephalic infants, 5 hours old delivered after a 38-47 weeks uncomplicated pregnancy. No hypothalamo-hypophiseal structures were found in any case macroscopically. The presence of some functioning hypophiseal cells could not be excluded.

Test	TRH			LHRH		IvGTT			
Hormone	TSH	T_4	T_3	FSH	LH-HCG	Cortisol	hGH	BG	Insulin
	µU/ml	µg/dl	ng/dl	µU/ml		ng/ml	ng/ml	mg/dl	µU/ml
1.Basal	4.3	8.8	31.0	<1.5	33.3	48.5	0.9	58.0	4.6
Peak	80.0	8.8	182.0	<1.5	33.3	79.5	1.2	299.0	17.1
2.Basal	14.7	13.5	113.0	2.7	41.5	48.2	5.3	0	9.5
Peak	>40.0	13.5	113.0	2.7	41.5	48.2	6.3	162.5	13.2
3.Basal	6.6	8.6	82.0	-	-	59.4	2.5	10.0	35.7
Peak	85.9	13.5	510.0	-	-	139.8	3.7	230.0	200.0
4.Basal	4.3	8.1	93.0	3.1	5.4	133.0	<1.5	64.0	10.0
Peak	>40.0	13.0	144.7	3.8	5.7	133.0	1.7	380.0	39.5

A.adenohypophiseal tissue in anencephaly is able to synthetize TSH constantly, hGH and ACTH inconstantly, if specifically stimulated. B. serum hGH and TSH increase present after delivery is hypothalamic dependent. C. thyroid and adrenals are able to synthetize hormones. D. β pancreatic function is not markedly impaired in anencephaly.

187 C.DACOU-VOUTETAKIS, A.MANOLAKI*, S.ANTONIADIS*, A.MINAKIDIS*, CH.KATERELOS*. A' Dept.of Pediatr. Athens University. Hypopituitary dwarfism with abnormal sella turcica (ST) in a father and son.

Familial cases of panhypopituitary dwarfism (PD) are rare and the type of inheritance has been autosomal recessive or sex linked. PD associated with hypoplastic pituitary and abnormal ST, as an isolated defect, has rarely been reported in siblings. Our report refers to a unique occurence of pituitary dwarfism with small pituitary fossa and abnormal sphenoidal bone in a father and son (Father: GH↓ son: ↓ GH, ACTH, TSH and pitressin). The TRH test on the son points to a primary pituitary defect. The mother's pituitary function was normal. This unusual maldevelopment which involes organs of different embryologic derivation (adenohypophysis on the father, adenohypophysis and neurohypophysis on the son) is difficult to interpret. In conjuction with the observation that in holoprocencephaly or other analogous CNS maldevelopment with pituitary hypoplasia, the pituitary fossa is normal, one may speculate that the primary developmental defect is dysplasia of the sphenoidal bone and especially of the pituitary fossa with secondary pituitary dysplasia, probably inherited as dominant trait, although heterozygocity in the mother cannot be excluded.

188 C.DACOU-VOUTETAKIS, M.PASVOURI*, S.BENETOS*, A.PAPAIOANNOU*. A' Dept.of Pediatr., Athens University

The association of congenital hypothyroidism (CH) and congenital heart disease (CHD). A unique developmental abnormality.

The concurrence of different teratogenetic events may not be fortuitus and their recording may lead to the pathogenetic mechanisms involved. For the past 6 years we have encountered the association of CHD and CH in 4 nonrelated families as follows: Family I: 2 brothers one with CHD (transposition of the great vessels), CH, and microcephaly, and the other a normally developing boy with only CH. Family II: 2 brothers; one with CHD and the other with CH. Family III: 2 brothers; one with CHD and CH who died of cardiac defect in infancy, and the other with CHD (tetralogy). Family IV : Identical twin girls; one with CH and the other with CHD (patent ductus arteriosus). In our children with CH the incidence of CHD is 10% which is higher than expected. However our material may be selected since complicated cases are more apt to be referred. These observations may be interpreted to indicate that the association of CHD and CH represents a unique teratogenetic complex probably genetically distinct from simple CH which is usually non familial.

189 D. EVAIN*, M. DONNADIEU*, M. ROGER, J.C. JOB, Hôpital Saint-Vincent de Paul, Paris, France.

Simultaneous study of somatotropic and corticotropic pituitary secretions by ornithine infusion test.

Stimulation of GH response to aminoacids infusion is well established. The ornithine chloride infusion (12 g/m2 within 30 min) induces an elevated peak of GH at a faithful time of 45 minutes. This test performed in 50 children with constitutional short stature showed the following results :

	T0	30 min	45 min
GH (ng/ml) (n = 50)	3.9 ± 1.0	10.5 ± 1.1	15.3 ± 1.2
Cortisol (ng/ml) (n = 50)	99 ± 5	160 ± 8	202 ± 8
ACTH (pg/ml) (n = 7)	29 ± 12	111 ± 18	98 ± 18

In pituitary dwarfs no elevation of GH and cortisol was observed.

In conclusion these data show that GH and cortisol levels mesured at zero and 45 min of ornithine infusion test allow to study simultaneously somatotropic and corticotropic pituitary functions.