

†1666 EFFECTS OF UNILATERAL HEMISPHERE LESIONS ON LANGUAGE AND INTELLECTUAL DEVELOPMENT. Dorothy M. Aram (Spon. by R.J. Martin) CWRU Dept. Ped., Cleve., Ohio.

The relationship between age and degree of hemisphere specialization vs. plasticity for higher cognitive functions remains controversial. The language and intellectual development of 8 left (L) and 8 right (R) hemisphere lesioned subjects (S's) thus were compared to controls (C's) matched by age, sex, race, and severity of congenital heart disorder. All S's were developing normally prior to sustaining CT scan verified lesions between 2 mos. and 6 yrs. (M: L=14.9 mos.; R=17.2 mos.) and were tested on a battery of intelligence and language measures > one year post onset (M age at test: L=4.3 yrs.; R=3.9 yrs.). Full range IQ's (M IQ: L=98.8; R=91.1) were below C's for both L (p=.02) and R S's (p=.01), yet most functioned within the normal range.

Semantic comprehension was depressed in both L (p=.04) and R S's (p=.04), yet semantic production was depressed only in L (p=.02) and not R S's. Syntactic abilities were significantly impaired for L but not R S's as assessed by the measures summarized below.

Syntax Measures	LS/LC	RS/RC
Developmental Sentence Score	p=.03	NS
Northwestern Syntax Screening Test	p=.01	NS
No. Grammatical Markers Used	p=.05	NS
% Complex Sentences Correct	p=.02	NS

These findings reveal that early L and R lesions impair IQ and semantics somewhat, but only L lesions significantly impair syntactic development. The study provides the first objective evidence, based on acquired L and R lesions, which demonstrates the L hemisphere's early specialization for syntactic abilities.

1667 DIFFERENTIAL SYMPTOM ONSET AND EVOLUTION IN CLINICAL SUBGROUPS OF TOURETTE SYNDROME.

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Having recently reported a high incidence of migraine among Tourette Syndrome [TS] patients and their families, and having found a greater prevalence of associated symptoms (e.g., somnambulism, night-terrors) and motion sickness in this clinically-identified subgroup, we investigated the possibility of a differential onset and evolution of tic symptoms depending upon the presence or absence of migraine in the patient and/or his or her immediate family. With a sample of 36 Tourette Syndrome children ranging in age from 5 to 19 years (x=11.9 years), we compared the 24 patients with either migraine (n=12) or a family history of migraine (n=12) (i.e., the "TS/migraine" group) with 12 patients having neither migraine nor a family history of migraine (i.e., the "TS/non-migraine" group). Significant differences were found: The TS/migraine group had onset of motor tics at an average age of 7.5 years, in contrast to 8.3 years in the TS/non-migraine group. Vocal tics emerged at an average age of 7.4 years in the TS/migraine group, compared with 10.2 years in the TS/non-migraine group. Compulsive acts such as touching objects, self, or others, were noted at an average age of 6.6 years in the TS/migraine group and 9.8 years in the TS/non-migraine group. Among TS/migraine patients, the majority of symptoms (75.8%) were present before age 8, in contrast to the TS/non-migraine patients in whom the majority of symptoms (82.9%) appeared after age 8.

1668 ABNORMAL SUCKING BEHAVIOR IN CNS-DAMAGED PREMATURE INFANTS. Murray A. Braun, Marjorie M. Palmer (spon. by N. Paul Rosman), Boston Univ. School of Med.

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Sucking behavior in growing premature infants has received little attention. Non-nutritive (NNS) and nutritive (NS) sucking was measured by suckometer 30 minutes prior to routine feedings, in 11 premature, AGA infants at 5-12 weeks of age, mean corrected age (C.A.) = 39-40 weeks. Six infants had intraventricular hemorrhage (IVH) by cranial ultrasound and computerized tomography. Non-IVH (n=5) infants included 2 with methadone abstinence syndrome, 2 with apnea/bradycardia treated with methylxanthines, and 1 with hydrancephaly. Sucking rate (sucks/sec) was measured during continuous (or the longest) sucking bursts.

	sucking rate (NNS)	sucking rate (NS)
IVH group (n=5)	1.37 sucks/sec	1.25 sucks/sec
hydrancephaly (n=1)	1.68 " "	1.63 " "
non IVH group (n=4)	1.88 " "	1.35 " "

The data indicates that in NNS, sucking rate is slower in infants with IVH and hydrancephaly versus those without IVH. The decrease in rate from NNS to NS is less in the IVH group and hydrancephaly vs. the non-IVH group. Thus, premature infants with CNS structural damage may show deficient adaptive sucking behavior.

†1669 ADRENOLEUKODYSTROPHY (ALD) WITH TRANSIENT AMAUROSIS WITHOUT CLINICAL DEGENERATION, A DISEASE VARIANT OR THIRD ALLELE. Gary G. Carpenter, Leonard J. Graziani, Jefferson Medical College, Phila., Pa.; Hugo W. Moser, John F. Kennedy Institute, Baltimore, Md.; Herbert H. Schaumberg, Albert Einstein College of Medicine, New York, N.Y. (Spon. by Herbert C. Mansmann, Jr.)

A 12 year old boy who presented with rapid onset of blindness and underlying Addison's disease recovered vision during maintenance with hydrocortisone and fludrocortisone acetate. At age 21 he remains clinically and neurologically well with the exceptions of red central scotomata and bilateral flexor plantar responses. Fibroblast assay revealed increased levels of saturated very long chain fatty acids and plasma hexacosanoate (C26 fatty acid) represented 0.048% total fatty acid with a C26/C22 ratio of 0.065, characteristics of ALD hemizygoty.

ALD has been differentiated from Adrenomyeloneuropathy (AMN), the latter involving the older male with slowly progressive spastic paraparesis, distal symmetrical polyneuropathy and variable hypogonadism; both associate with Addison's disease. Whether this patient made the transition from one (ALD) to the other (AMN) or whether he now shows the new pattern of a third disease with adrenal energy, deferred central nervous system decay and excessive collection of saturated very long chain fatty acids, remains questionable. Hypothetical recovery from ALD seems least likely, a possibility that might result from a third allele diminishing accumulated levels of very long chain fatty acids.

1670 EFFECT OF MATERNAL HYPOXIA ON FETAL RAT BRAINS Rebecca Cooper and Robert C. Vanucci

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Cerebral metabolic and neuropathologic changes in fetal rats were examined during and following maternal hypoxia. Term pregnant rats were tracheostomized or intubated, paralyzed, and artificially ventilated with 40%, 15% or 10% O₂-balance N₂. Maternal acid-base and cardiovascular status was monitored. Dams then were subjected to C-section or allowed to recover and deliver spontaneously. Dams exposed to 15% O₂ for 60 minutes were hypoxic (paO₂=38mmHg) and acidotic (pHa=7.22mmHg). MABP fell by 25%. In fetal brain, lactate (L) and pyruvate (P) increased, but disproportionately such that the L/P ratio increased 10 fold. Brain glucose and glycogen decreased. Both ATP and P-Creatine decreased, indicating a disruption of the energy state of the brain.

In resuscitated dams, fetal survival at delivery was 86%. Sequential measurements of postnatal growth and development of surviving pups were the same as offspring of control dams. No histologic alterations of hypoxia-ischemia were found in brains of pups sacrificed at 30 days; hypomyelination of ascending and descending spinal cord pathways was seen in a few animals. In dams exposed to 10% O₂ for 30 minutes, systemic hypoxia was more severe (paO₂=33mmHg), but pHa and MABP were comparable to levels obtained in dams exposed to 15% O₂ for 60 minutes. Changes in glycolytic intermediates and high energy metabolites of fetal brains as well as functional parameters were comparable to fetuses of dams exposed to 15% O₂ for 60 minutes.

The findings indicate that although maternal hypoxia leads to major alterations in the energy status of the fetal brain, this does not necessarily culminate in overt brain damage.

1671 INFANTILE APNEA AND SEIZURES: A POSSIBLE ROLE FOR CAFFEINE (C) STIMULATION. J.M. Davis, T. Turmen, J.V. Aranda, Dept Pediatrics, McGill University - Montreal

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A diagnosis of "Near-miss Sudden Infant Death Syndrome (SIDS)" is usually made after a thorough medical evaluation fails to reveal the cause of an apneic episode. "High risk" infants are placed on apnea monitors or are treated with respiratory stimulants such as C or theophylline. We have treated over twenty infants in the past three years with C for infantile apnea. All infants had normal diagnostic evaluations, including electroencephalograms (EEG), before starting treatment. Two infants experienced short, generalized, tonic-clonic seizures soon after being treated with C (10 mg/kg). Repeat metabolic studies were normal and C levels were within the therapeutic range (8-20 mg/l). The seizures were controlled with anticonvulsants. Repeat EEG's in both patients revealed epileptiform discharges. Anticonvulsant therapy was continued with complete resolution of the apnea and no further seizures. Follow-up EEG's are now normal and both patients are off all medications. Experience in these two infants suggests: 1) There is a subgroup of patients diagnosed as "Near-miss SIDS" who have apnea associated with seizures. 2) Seizure threshold may be lowered by central nervous system stimulants such as C and may produce convulsions in infants with latent seizure disorders not detected by routine EEG. 3) It is unlikely that C given in therapeutic doses directly causes seizures, since a group of premature infants treated for neonatal apnea showed only transient jitteriness at levels up to 84 mg/l. 4) C may be of value in the diagnosis of infantile apnea caused by seizures.