●1315 SYNDROMIC AND ISOLATED CRYPTOPHTHALMOS. I. T. Thomas, V. Felix, L. Sanchez de Leon, R. A. Hernandez, M. C. Jones and J. L. Frias. University of Florida College of Medicine, J. Hillis Miller Health Center, Department of Pediatrics, Gainesville, Florida, Residencia Sanitaria, Toledo, Spain, and University of California, Department of Pediatrics, San Diego, California.

Cryptophthalmos syndrome is an autosomal recessive pattern of malformation characterized by cryptophthalmos, syndactyly, laryngeal atresia, abnormal genitalia and renal agenesis. We have found 72 cases of the syndrome and 40 of isolated cryptophthalmos among 112 cases reported in the literature since Zehender's first description in 1872. The isolated cases, which have not been previously analyzed, contain three families showing vertical transmission, while the syndromic ones suggest autosomal recessive inheritance. We report three stillborn siblings pairs who presented with cryptophthalmos, bilateral renal agenesis, oligohydramnios sequence, genital abnormalities and syndactyly. Four had laryngeal stenosis or atresia and two siblings had aplasia or hypoplasia of the thymus. Review of the literature and our own experience illustrate the high frequency of renal agenesis in Cryptophthalmos syndrome and the existence of the disorder without cryptophthalmos. Abnormalties similar to those found in this syndrome, some of which may involve defects in programmed cell death, have been observed in two vitamin A deprived animal models. The Cryptophthalmos syndrome should be considered more frequently in the differential diagnosis of multiple congenital malformations, especially when associated with renal agenesis and even in the absence of cryptophthalmos.

1316 CEREBRAL METABOLIC ALTERATIONS IN RATS FOLLOWING ALCOHOL EXPOSURE: R.Vingan,D.Dow-Edwards, and E.Riley Lab of Cerebral Metabolism, Dept. Neurosurgery, SUNY Downstate Med.Ctr., Dept. Psych., SUNY Albany. (Spon. AK Brown)

In order to define the cerebral metabolic correlates occurring with prenatal alcohol exposure in rats we chose the autoradiographic ^{14}C deoxyglucose method for measuring in vivo glucose utilization rates in brain (Sokoloff et al., 1977). Isocaloric liquid diets containing either 35% or 0% ethanol derived calories (EDC's) were fed (pair-feeding) to pregnant Long Evans rats during day 6 to 20 of gestation. The offspring were tested with Shuttle box avoidance techniques on day 90 and were subjected to the deoxyglucose procedures at 105 days of age. ^{14}C labeled deoxyglucose was administered as an intravneous pulse (125 uCi/kg). Timed arterial samples were taken for determination of the time course of the arterial plasma concentrations of (^{14}C) deoxyglucose and glucose. At 45 minutes the animals were sacrificed, the brains removed, frozen and later sectioned for autoradiography. Areas of interest were analyzed densitometrically to determine the local concentrations of ^{14}C in the tissues. From the local tissue ^{14}C concentrations and the time courses of the plasma (^{14}C) deoxyglucose and glucose concentrations, local rates of glucose incorporation were calculated by the operational equation of the method. Preliminary analysis suggests that alterations in glucose metabolism occur in the posterior pituitary, supraoptic nucleus of the hypothalamus, ventral caudal nucleus of the lateral lemniscus, median raphe nucleus and sensory-motor cortex.

NEONATOLOGY

1317 THE PERINATAL/NEONATAL PROFILE OF SUBARACHNOID HEMORRHAGE (SAH). D.H. Adamkin, J.W. Fleischaker, L.N. Cook, R.J. Shott. University of Louisville, Division of Neonatal Medicine. Spon. B.F. Andrews.

One-hundred and four neonates with SAH diagnosed by CT scan within the first week of life admitted to the NICU over a 34 month period were studied. Mean birthweight and gestational age were 3110gm (range 1659-5290) and 38.7 weeks (31-42) respectively.

Abnormal fetal heart tracings preceded delivery in 29/104 (27.8%) and resulted in 22 emergency c-sections. Mean one and five minute Apgar scores were 5.4 and 7.1 respectively. One-third had an Apgar of ≤3 at one minute and one-half of these neonates still were ≤3 at five minutes.

Ninety-three (89%) were symptomatic with over one-third exhibiting more than one symptom. These included apnea and bradycardia (42%), seizures (36%), hypotonia-lethargy (30%), cyanosis-dusky spells (20%). Onset of symptoms occurred within 24 hours of birth in 2/3 while the remainder became symptomatic between 24 and 48 hours.

Follow-up CT scans performed in 28 infants showed a normal or resolving scan in all but one who developed posthemorrhagic hydrocephalus and brain atrophy. The other infants followed during the first year showed no clinical evidence of hydrocephalus.

Apneic and/or bradycardic spells, seizures, lethargy and unexplained cyanotic episodes occurring in term infants within 48 hours of birth associated with a history of fetal distress or perinatal asphyxia are characteristic of SAH. Follow-up CT scans and clinical evaluation demonstrate rapid resolution without PHH in most infants.

1318 A PROPOSED CLASSIFICATION FOR SUBARACHNOID HEMORRHAGE (SAH). D.H. Adamkin, J.W. Fleischaker, L. Shearer, L.N. Cook, S.A. Wilkerson. University of Louisville, Division of Neonatal Medicine. Spon. B.F. Andrews.

One-Hundred and four neonates with SAH diagnosed by CT scan within 7 days of life were graded into 3 groups on the basis of theri CT exam: Grade 1 (mild)- visualization of the falx without brain edema; Grade 2 (mod)- visible pooling of blood below the tentorium in the post fossa; Grade 3 (severe)- diffuse blood in subarachnoid spaces of the brain convexities. The following clinical characteristics were noted:

	Grade 1	Grade 2	Grade 3
n	40	47	17
Mean BW(g)/GA(wks)	3363/39.4	3063/39.1	2644*/36.1**
Fetal Distress	8/40 (20%)	13/47 (28%)	8/17 (47%)*
Emergency c-sect	6/11 (55%)	8/13 (60%)	7/7 (100%)
Apgars 1'/5'	5.9/7.4	5.3/7.2	4.7/6.0
Apgar ≤3 at 1'	9/37 (24%)	16/46 (35%)	4/17 (24%)
Apgar ≤3 at 5'	4/35 (11%)	7/45 (16%)	4/17 (24%)
Onset of SX's <24°	19/40 (48%)	34/47 (72%)	16/17 (94%)***
Seizures	13/40 (33%)	20/47 (43%)	6/17 (35%)

(*p .05: Gp. 2 vs3;**Gps. 1 & 2 vs 3;***Gp. 1vs Gp. 2 & Gp. 3)

Severe (grade 3) SAH was more likely in the smaller more preterm neonates. Grade 3 SAH was more frequently preceded by fetal distress (abnormal fetal heart rate tracings) and required emergency c-section. Earlier onset of symptoms directly correlated with increasing severity of hemorrhage. Finally, seizures, a prominent symptom in all neonates with SAH was not correlated with increasing severity.

1319 BUFFY COAT TRANSFUSION: AN EFFECTIVE ALTERNATIVE IN SEVERELY ILL, GRANULOCYTE-DEPLETED NEWBORNS. Charles Addo-Yobo, Gloria Valencia, Scott T. Miller, Sreedhar P. Rao, Audrey K. Brown. SUNY-Downstate Medical Center-Kings County Hospital, Bklyn, N.Y.

Granulocyte transfusion may be life-saving in newborns with bacterial sepsis, neutropenia and severe depletion of the bone marrow mature neutrophil storage pool (NSP). Since a buffy coat is more readily available than granulocytes obtained by leukapheresis, we studied the efficacy of fresh (<24hrs), irradiated buffy coat transfusion in this high risk group. Bone marrow aspiration was done on 16 newborns with clinical sepsis and an absolute neutrophil count (ANC) <2000/mm^3. Four were found to have severe depletion of NSP, i.e. (metas + bands + segs) <7% of nucleated cells, and were given 10cc/kg of a buffy coat preparation. Three of these infants received 0.5-1x 10^9 neutrophils/mm^3/kg. Two had an increase to normal ANC within 12 hours and recovered from clinical sepsis. The rise in ANC continued and was far in excess of that predicted based on the number of granulocytes transfused. The third infant had a transient rise to 2350 neutrophils/mm^3 6 hours post-transfusion, but subsequently remains neutropenic for 6+ weeks. A fourth infant whose buffy coat provided only 0.06 x 10^9 neutrophils/mm^3/kg received a second transfusion from the same unit 12 hours later, recovered fully from neutropenia, and survived. Twelve infants with NSP >7% were not transfused; one of these had an NSP of 8.5% and expired. The remainder survived but remained neutropenic 4-16 days. Fresh buffy coat preparations appear to accelerate recovery from neutropenia and may improve survival in clinically septic NSP-depleted newborns.

1320 ELEVATION IN SYSTEMIC BLOOD PRESSURE IN THE NEONATE FOLLOWING ABDOMINAL PALPATION. Raymond D. Adelman, Robert A. Sinkin and Barry L. Phillips, University of California, Davis, Department of Pediatrics, and Children's Hospital Medical Center, Oakland, California.

Systemic blood pressure in the neonate may be affected by such variables as crying, postnatal age, and birth weight. This study demonstrates that routine abdominal examination in the ill neonate significantly elevates systolic and diastolic blood pressure.

Blood pressure was monitored on a Corometrics NeoTrak Monitor during palpation of the liver, kidney and bladder of 19 ill infants in a neonatal intensive care unit. Baseline and recovery values of 2 minutes duration were obtained prior to and following palpation. 17 of 19 infants were on mechanical ventilators. Mean birthweight, gestational age and chronological age at the time of study were 1684 grams, 31.8 weeks, and 43 hours respectively. Systolic and diastolic blood pressure rose significantly with palpation. No changes in PO_2 or PCO_2 values were noted.

	Baseline	Liver	Baseline	Kidney	Baseline	Bladder
Syst. BP(mmHg)	54±2	62±3†	54±2	67±2†	53±3	64±2†
Diast.BP(mmHg)	32±1	36±2†	32±1	39±2†	31±2	38±1†

In summary, systolic and diastolic blood pressure increase by up to 24% during abdominal palpation in the ill neonate. Whether abdominal palpation increases the risk of intraventricular hemorrhage in susceptible low birthweight infants remains to be determined.

† p <.001