CONGENITAL RUBELLA AND CNS DEFECTS

The University Department of Pediatrics and Child Health, Leeds, and the Department of Microbiology, Hospital for Sick Children, London, collaborated in a study of the time relations between maternal rubella infection in pregnancy and the presence and type of defects in 422 children with confirmed congenital rubella registered in the National Congenital Rubella Surveillance Program at their institutions.

In 106 cases with maternal rubella confirmed by a 4-fold rise in hemagglutination inhibition (HI) titer and/or detection of rubella-specific lgM, 66 (62%) had defects, 44 (67%) were exposed to rubella between the 3rd and 12th weeks of pregnancy and the remaining 22 (33%) between the 13th and 17th weeks. Multiple defects involving the CNS, heart, eye, and hearing were recorded only in children exposed early and up to the 12th week, deafness was also found in some exposed up to the 17th week, and no defects occurred in 20 children exposed after this time. Deafness was the most common defect, reported in 62 (58%) of 106 children; it was sensorineural in 55 (89% of the 62) and mixed conductive and nerve deafness in 7 (11%). Of 9 children with CNS defects, 6 had microcephaly, 2 cerebral palsy, and 1 mental retardation.

Among 316 cases with confirmed congenital rubella but without laboratory confirmation of maternal infection, 265 (84%) had defects. Deafness occurred in 75%, eye disorders in 30%, CNS defects in 25% and the heart was affected in 20%.

School placements for 148 affected children are as follows: 62 (42%) schools for the deaf, 33 (22%) partial hearing units in normal schools, 3 educationally subnormal, 3 blind, and 40 (27%) attend normal schools. Three-quarters of children with confirmed congenital rubella attend special schools. (Munro ND, Jones G et al. Temporal relations between maternal rubella and congenital defects. Lancet 1987; 2:201-204).

COMMENT: These data demonstrate that the risk of fetal damage from maternal rubella is very small with exposure later than the 16th week of pregnancy. Before that time the risk of congenital defects is high. The incidence of defects in this study (62%) is considerably higher than that reported previously (20%) by Miller E et al. (Lancet 1982; 2:781). The increased incidence is explained by the high proportion (65%) of infants exposed up to the 16th week of pregnancy as contrasted to 38% in the earlier study. A small head circumference (<10th percentile) occurring in 18% of 102 affected infants was an additional abnormality in the Miller study but was not included among the reported rubella defects.

## PERINATAL DISORDERS

INTRACRANIAL HEMORRHAGE IN TERM INFANTS

The authors identified 12 cases of symptomatic intracranial hemorrhage (ICH) among 23,000 infants born at the Beth Israel and Brigham and Women's Hospitals in Boston, MA, in 1982 and 1983. The incidence of ICH was 5.2/10,000 live births or 5.9/10,000 term appropriate-for-gestation age infants (>-2,500g and >-37 wks gestation). Delivery was spontaneous vaginal in 8, forceps in 3, and cesarean section in 1. Pitocin was used in 5 cases; the indications were induction for postdate in 3 and prolonged second state in 2. Mean Apgar scores at 1 and 5 mins were 5 (range 2-9)

and 7 (1-10). Neonatal seizures were the most common presenting symptom, occurring in 7 (60%) infants. Most infants were symptomatic by the second day. Eleven survived the neonatal period. None developed hydrocephalus. The diagnosis of ICH was confirmed by CAT scan. In most cases the hemorrhage was in the subarachnoid space and the CSF obtained by lumbar puncture contained blood. Obstetrical risk factors were: 1) prolonged second stage of labor (>2 hrs), 2) very rapid labor with rapid descent, and 3) Pitocin and forceps delivery. CAT scan or ultrasound were considered essential for diagnosis of ICH. (Sachs BP, Acker D et al. The incidence of symptomatic intracranial hemorrhage in term appropriate-for-gestation-age infants. Clinical Pediatrics 1987; 26:355).

COMMENT: Primary subarachnoid hemorrhage (SAH) was the most common form of hemorrhage in a previous study of 22 term newborn infants seen in a 5 year period with intracranial hemorrhage on CT examination (Fenichel GM et al. Arch Neurol 1984;41:30). All infants with diffuse SAH had seizures on the first day of life. Five of 10 infants with SAH were born after prolonged labors, traumatic deliveries, and intrauterine asphyxia; three had intrauterine asphyxia. Five of 8 infants with intraventicular hemorrhage (IVH) had difficult deliveries with trauma and asphyxia. In 3 infants with IVH and 3 with intracerebral hemorrhage (ICH) the cause was unexplained, i.e. in almost one-third of the total group. The occurrence of intracranial hemorrhage does not preclude a good outcome. Development was normal or near normal at follow-up in one-half the infants who suffered a SAH and in 3 of 7 with IVH. Seizures in the first week of life are an important sign of SAH, IVH and ICH, occurring in almost all cases.

PROGNOSIS OF NEONATAL ASPHYXIA

Of 86 asphyxiated full-term neonates with CNS complications admitted to the Intensive Care Unit at the Nagoya City University Hospital, Japan, during a 10 year period (1972-81), 63 (73%) survived the neonatal period. The CNS complications were hypoxic-ischemic encephalalopathy (58), intracranial hemorrhage (27) and brain infarction (1). Of 54 survivors who were followed for 3 to 13 years, 8 (15%) had major handicaps such as cerebral palsy, epilepsy and mental retardation and 5 (9%) had minor CNS abnormalities. Factors predictive of long-term CNS sequelae were: 1) absent Moro-reflex over 6-days, and 2) abnormal neurological signs on discharge.

Each neonate had a 1 min Apgar score of 6 or less, neurological abnormalities including stupor or lethargy, hypotonia, abnormal respirations, jitteriness or seizures, and one or more abnormal findings on laboratory tests such as spinal tap, echoencephalograpy, cerebral angiography, EEG, and CAT scan. (Ogawa IT, Kanayama M, Wada Y. Long-term prognosis of asphyxiated full-term neonates with CNS complications. Brain Dev 1987; 9:48-53).

COMMENT: Another method of assessment of neurologic outcome in asphyxiated term infants is by use of serial CK-BB, or creatine kinase brain-type isoenzyme measurement (Walsh P. J Pediat 1982; 101:988). Serum CK-BB activity, when measured in cord blood and at 6-12 hours of life, correlates with neurologic outcome after severe asphyxia, comparing favorably with CT scanning as a prognostic factor. The mean of elevations in CK-BB in asphyxiated was 4-fold the values obtained in control infants. Normal CK-BB activity was a predictor of good neurologic outcome.