PEDIATRIC NEUROLOGY BRIEFS

A MONTHLY JOURNAL REVIEW

J. GORDON MILLICHAP, M.D., F.R.C.P., EDITOR

Vol. 2, No. 3 March 1988

CONGENITAL MALFORMATIONS

DIABETES AND FETAL MALFORMATIONS

Diabetic and healthy control pregnant women were followed in a multicenter collaborative study coordinated by the Epidemiological Branch, National Institute of Child Health and Human Development, Bethesda, MD. Major malformations, including ananencephaly, arhinencephaly and holoprosencephaly, microcephaly, meningomyelocele and hydrocephalus, were detected in 4.9% of diabetic women who entered the study early compared to 9% in late-entry diabetic subjects (P=.032) and 2.1% in controls (P=.027). Mean blood glucose and glycosylated hemoglobin levels during organogenesis were not significantly higher in women whose infants were malformed, and hypoglycemia was not more common in the same group. Hyperglycemia during organogenesis was not correlated with malformation. The authors conclude that not all malformation can be prevented by good glycemic control but the lower incidence among women studied within 21 days of conception (early-entry group) as compared with the late-entry group justifies good metabolic control around time of conception. (Millis JL et al. Lack of relation of increased malformation rates in infants of diabetic mothers to glycemic control during organogenesis. N Engl J Med March 17 1988;318:671-6.

COMMENT. Previous studies have shown low malformation rates in diabetic women who achieved excellent periconceptional glycemic control. The present study suggests that poor glycemic control explains some but not all diabetes associated malformations. Metabolic factors other than glycemic control may be relevant according to animal studies. Genetic factors may also be involved and female offspring are more susceptible. Congenital optic nerve hypoplasia, not encountered in this study, has been reported in women whose mothers had diabetes mellitus. (Nelson M et al. Arch Neurol 1986;43:20).

ACROCALLOSAL SYNDROME (SCHINZEL SYNDROME)

The acrocallosal syndrome, first described by Schinzel (Helv Paediatr Acta 1979;34:141) and characterized by dysmorphic features, macrocephaly, polydactyly, mental retardation, and agenesis of the corpus callosum, is

PEDIATRIC NEUROLOGY BRIEFS ©1988 covers selected articles from the world literature and is published monthly. Subscription requests ($25 US or £15 UK) may be sent to: Pediatric Neurology Briefs - J. Gordon Millichap, MD, FRCP - Editor, P.O. Box 11391, Chicago, IL 60611, USA, or P.O. Box 1605, London W8 7JD, UK. The Editor is Professor of Neurology and Pediatrics at Northwestern University Medical School, Chicago, Illinois, USA.