

palate, congenital malformations of the urinary tract. The authors considered that their findings strengthened the hypothesis that BZD intake during early pregnancy is associated with teratogenicity in man but there is as yet no firm proof of association. Diazepam in early pregnancy should be avoided on the basis of this and other studies which strongly suggest a teratogenic effect of BZD.

## NEURO-OPHTHALMOLOGY

### OCULAR SIGNS OF CHIARI MALFORMATION

Twenty-eight patients (14 females and 14 males aged between four and 34 years) with myelomeningocele and Chiari malformations were examined neuro-ophthalmologically at the Karolinska Institute, Huddinge University Hospital, Huddinge, Sweden. The Chiari malformation, determined with MRI, was type I in three patients and type II in 25. All had hydrocephalus; mild in 12, moderate in 13, and marked in three. Shunt procedures had been performed in 20. Spontaneous or gaze related nystagmus and abnormal optokinetic nystagmus were the most common disturbances of ocular motility. Horizontal gaze paresis occurred in 14 patients and vertical gaze limitations in nine, all in upward gaze. Horizontal nystagmus occurred in 17 whereas vertical nystagmus was uncommon and downbeat nystagmus was not observed. Strabismus occurred in 11 patients and esotropia was more common than exotropia. No signs of optic atrophy or other changes in the visual pathways were found. (Lennerstrand G, Gallo JE. Neuro-ophthalmological evaluation of patients with myelomeningocele and Chiari malformations. Dev Med Child Neur May 1990; 32:415-422).

COMMENT. Downbeat nystagmus has been considered almost pathognomonic of the Chiari malformation but was absent in the cases in this study. A girl aged ten referred to me recently because of migraine headaches had downbeat nystagmus and a refractive error; the MRI was negative for Chiari malformation and brain stem auditory evoked responses were normal. Lennerstrand G et al have used the MRI to determine the correlation between disturbances of ocular motility and the degree of hydrocephalus, tactile plate deformity, and dislocation of the cerebellum and medulla oblongata in 28 patients with myelomeningocele. (Dev Med Child Neur May 1990; 32:423-431). All patients had Chiari malformations. Strabismus and spontaneous nystagmus were related to the degree of hydrocephalus and to the amount of lower brain stem deformities. Convergence defects correlated with upper brain stem deformities.

### OPTIC NERVE HYPOPLASIA

The pathology, clinical features, and disorders associated with optic nerve hypoplasia in children are reviewed from the Tennent Institute of Ophthalmology, Weston Infirmary, Glasgow, Scotland. A total of 100 references is provided. Histologically a reduced number of optic nerve fibers can be demonstrated in a smaller than normal optic nerve. An overgrowth of retinal pigment epithelium surrounding