degree of disability. Thoracic scoliosis and horseshoe kidney that may be recognized during fetal life are associated with sensory levels in the thoracic region and are predictive of a poor prognosis. The author points out that the reliable discrimination between mild and severe cases of spina bifida in early pregnancy is not yet possible and parents should be informed of the likelihood of prolonged dependency into adulthood.

ANENCEPHALY

The Medical Task Force on anencephaly coordinated by Stumpf DA (Northwestern University Medical School, Chicago) presents a consensus statement limited to medical issues of organizations of physicians caring for fetuses and infants with anencephaly. The statement was approved by the AAP, AAN, ACOG, ANA, and CNS. Anencephaly is defined as a congenital absence of a major portion of the brain, skull, and scalp with its genesis in the first month of gestation. The primary abnormality is failure of cranial neurulation, the embryologic process that separates the precursors of the forebrain from amniotic fluid. Anencephaly does not mean the complete absence of the head or brain. Craniofacial anomalies are associated and up to 1/3 have defects of the non-neural organs that could preclude their use for transplantation. The maternal serum a-fetoprotein level is elevated in 90% of cases, and elevated a-fetoprotein levels in amniotic fluid and the presence of acetylcholinesterase on electrophoresis occur in virtually all cases. Ultrasonography is also reliable in the prenatal diagnosis of anencephaly. The postnatal diagnosis requires the following criteria: 1) Absence of a large portion of the skull, 2) absence of scalp over skull defect, 3) exposed hemorrhagic fibrotic tissue, 4) absence of recognizable cerebral hemispheres. The cause is usually not known and a polygenic or multifactorial etiology is suggested. Chromosome abnormalities and mechanical factors are recognized associations. Hyperthermia and deficiencies of folate, zinc and copper in the mother have been invoked. In recent years, 80-90% are aborted, 7-10% are stillborn, and 3-11% are live born. Most live born anencephalic infants have died within the first days after birth and survival beyond one week occurred in 0-9% in three series. Two months was the longest survival confirmed with accepted diagnostic criteria. The estimated incidence of anencephaly in the U.S. is 0.3-7/1000 births and the incidence of live born infants with anencephaly would be less than 100 per year. Anencephalic infants have no functioning cerebral cortex and are permanently unconscious. Brain stem functions are present in varying degrees and the diagnosis of brain stem death depends on the disappearance of previously existing brain stem functions, including loss over an observation period of at least 48 hours of measurable cranial nerve function and spontaneous movements, and a positive apnea test. Confounding factors such as drugs, hypothermia, or hypotension should be excluded. The use of organs from infants with anencephaly for transplantation is also discussed. (Stumpf DA et al. The infant with anencephaly. N Engl J Med March 8, 1990; 322:669-674).

COMMENT. This comprehensive report on the infant with anencephaly provides medical information of importance in the analysis of social, legal, and ethical issues concerning transplantation of organs from anencephalic infants.

CONGENITAL NEUROLOGIC MALFORMATIONS

A 17-year survey of major congenital neurologic malformations among infants born in U.S. Army Hospitals worldwide from January 1, 1971 through December 31, 1987 is presented from the Neonatology Services, Walter Reed Army Medical Center, Washington, DC and Travis Grant USAF Medical Center, Travis Air Force Base, CA. From a population of 763 364 live-born and stillborn infants, 275 had anencephaly (0.36 per 1000 total births), 526 had spina bifida (0.69 per 1000), 112 had encephaloceles (0.15 per 1000), and 370 had hydrocephalus (0.48 per 1000 total births). The incidence of CNS defects among stillborn infants was 24 times greater than among live-born infants. There was a female preponderance of infants with anencephaly, spina bifida and encephalocele and a male predominance for hydrocephalus. Black infants were less likely than white infants to have spina bifida. Other congenital anomalies were associated in 20% of infants with anencephaly, 40% with encephaloceles, 37% with hydrocephalus, and 22% with spina bifida. (Wiswell TE et al. Major congenital neurologic malformations. A 17-year survey. AJDC Jan 1990; 144:61-67).

COMMENT. The racial background of the patient population in this study closely resembled that of the United States as a whole and the results may reflect those of the U.S. In the past 20 years, declines in the frequencies of anencephaly and spina bifida have been noted in many countries, particularly in the British Isles. In the present study the incidence of neural tube defects decreased only among white female infants and no etiological factor could be implicated.

CEREBELLAR VERMIS AGENESIS

The syndromes of vermal agenesis are reviewed from the Department of Pediatrics, Hopital des Enfants Malades, Paris, France. These include the Dandy-Walker syndrome and other complicated cases associated with multiple abnormalities. The Dandy-Walker syndrome consists of three abnormalities of development: 1) Partial or complete agenesis of the vermis of the cerebellum; 2) cystic formation in the posterior fossa communicating with the fourth ventricle; and 3) hydrocephalus. Enlargement of the posterior fossa and elevation of the torcular and lateral sinuses are sometimes included among the diagnostic criteria. Associated abnormalities include agenesis of the corpus callosum (7-15% of patients), occipital encephalocele (18%), cleft lip and palate, cardiac malformations, urinary tract abnormalities, and minor facial dysmorphisms. The prognosis is guarded, 75% having borderline IQ or lower, and a mortality rate of 27% in some series. Various chromosomal abnormalities have been demonstrated in a few patients but their significance is unclear. There is a 1-2% chance of recurrence in the same family. Syndromes of