agenesis of the cerebellar vermis of genetic origin are distinguished from the Dandy-Walker malformation. These include Joubert syndrome, Walker-Warburg syndrome, Meckel-Gruber syndrome, and atypical Dandy-Walker with facial angioma. Joubert syndrome includes panting respirations, abnormal eye movements, facial asymmetry and ataxia, in addition to vermian agenesis. The MRI shows an umbrella shaped fourth ventricle. Walker-Warburg syndrome includes lissencephaly, retinal abnormalities and hydrocephalus. Meckel-Gruber syndrome is characterized by occipital encephalocele, polycystic kidneys, polydactyly and hydrocephalus. Some have congenital muscular dystrophy in addition. The inheritance pattern is autosomal recessive. An MRI with median sagittal cuts is usually required in the diagnosis of partial agenesis. No reliable metabolic marker has been determined but some cases of vermian agenesis are associated with abnormal urinary excretion of succinyl-purines and pipecolic acid. Shunting operations are required when hydrocephalus develops. Operations on the posterior fossa have a high rate of failure. Prognosis depends on the occurrence of other CNS abnormalities. (Bordarier C., Aicardi J. Dandy-Walker syndrome and agenesis of the cerebellar vermis: Diagnostic problems and genetic counselling. Dev Med Child Neurol April 1990; 32:285-294).

> COMMENT. In patients with agenesis of the cerebellar vermis a correct diagnosis is important in therapy, genetic counseling, and prognosis. Cases with complications which are usually autosomal recessive in inheritance and having a poor prognosis must be distinguished from the typical Dandy-Walker syndrome which is often amenable to surgical therapy.

CONGENITAL CONTRACTURAL ARACHNODACTYLY

An infant girl with arachnodactyly and spontaneously resolving contractures who died in cardiac failure is reported from the Paediatric Unit, Northern General Hospital and Department of Ophthalmology, Royal Hallamshire Hospital, Sheffield, England. In addition to the arachnodactyly the infant had dolichostenomelia, iridodonesis, and mitral and tricuspid incompetence. There was no evidence of lens subluxation on slit lamp biomicroscopy. Chromosome studies and urinary homocystine were normal. (Huggon IC et al. Contractural arachnodactyly with mitral regurgitation and iridodonesis. Arch Dis Childhood March 1990; 65:317-319).

> COMMENT. Congenital contractural arachnodactyly has been described as an autosomal dominant syndrome distinct from classical Marfan's syndrome and usually unassociated with serious ocular and cardiovascular complications. This case report questions this distinction and emphasizes the importance of cardiovascular and ophthalmic assessment of patients with contractural arachnodactyly. As an editorial comment from Springfield, Illinois, I cannot omit the frequent reference to President Abraham Lincoln and Marfan's syndrome.