COMMENT. Duration of disease and age are important in reviewing the symptoms and serum enzymes at time of diagnosis of JDM. Children with longer disease duration tend to have fewer diagnostic findings of JDM, and their muscle enzymes are often normal. Older children have more symptoms of arthritis and dysphagia compared to children age 6 years and under. Systemic vasculopathy, with decreased food absorption, and dysphagia may account for the lower percentile height and weight of children with JMD. Early diagnosis and prompt institution of treatment are recommended.

Dysphagia in facioscapulohumeral muscular dystrophy (FSHD) was demonstrated in 7 of 8 adults, most having weakness of tongue and jaw muscles. (Wohlgemuth M et al. **Neurology** 2006;66:1926-1928). This finding is contrary to the original description of FSHD by Landouzy and Dejeurine in 1885, emphasizing the integrity of the muscles of the tongue and jaw. Dysphagia appears to occur only in severe cases of FSHD, and does not involve muscles of the pharynx.

SEIZURE DISORDERS

HYPOGLYCEMIA AND SEIZURES IN LARGE FULL-TERM NEONATES

The risk of hypoglycemia-associated seizures in large-for-gestational-age (LGA) full-term neonates was assessed by analysis of data from the Netherlands Perinatal Registry at Wilhelmina Children's Hospital, Utrecht. Hypoglycemia, defined as a plasma glucose below 2.5 mmol/l, was recorded from 1997-2002 in 1513 of 9318 (16.2%) of LGA full-term neonates without maternal diabetes; 20 (1.3%) of these had seizures, and hypoglycemia was the single cause of seizures in 6. Perinatal asphyxia, birth trauma and infections may have contributed to the hypoglycemia and seizures in the remaining 14. Hyperinsulinism was demonstrated in one LGA neonate with seizures and maternal diabetes, and in none without diabetes. The number with abnormal neurodevelopment was not recorded. The safe lower level of plasma glucose in healthy LGA neonates needs to be established. (Groenendaal F, Elferink-Stinkens PM, & the Netherlands Perinatal Registry. Hypoglycaemia and seizures in large-for-gestational-age (LGA) full-term neonates. **Acta Paediatrica** July 2006;95:874-876). (Respond: Floris Groenendaal, Department of Neonatology, Room KE 04.123.1, Wilhelmina Children's Hospital/University Medical Center, Lundlaan 6, 3584 EA Utrecht, the Netherlands).

COMMENT. Hypoglycemic seizures may occur in LGA full-term neonates even in the absence of risk factors, such as maternal diabetes, perinatal asphyxia, trauma or infections, but the incidence is low. A wide variation in definition of hypoglycemia is quoted in textbooks, ranging from a glucose level of <1 to <2.5 mmol/l for term babies of appropriate weight. An even greater range is used by practicing pediatricians (Koh THHG et al. Neonatal hypoglycemia – the controversy regarding definition **Arch Dis Child** 1988;63:1386-1398). The safe blood glucose concentration varies with many factors, including rate of fall of blood glucose. (Etheridge JE Jr. Hypoglycemia and the central

nervous system. In Pediatric Neurology, Ed. Millichap JG. **Ped Clin N Amer** 1967;14:865-880).

RATE OF DEVELOPMENT OF INTRACTABLE EPILEPSY

The time taken to develop intractability of epilepsy was determined prospectively in a cohort of 613 children followed in the Connecticut Study for a median of 9.7 years and reported by researchers from Northern Illinois University, DeKalb, IL; University of California Los Angeles, CA; Yale Medical School, New Haven, CT; and Albert Einstein College of Medicine, Bronx, NY. Intractability was defined in two ways: 1) 2 drugs failed, 1 seizure/month (average) for 18 months (stringent definition), and 2) failure of 2 drugs. Delayed intractability was defined as 3 or more years after epilepsy diagnosis. Intractability (stringent form) developed in 83 children (13.8%), and the 2-drug definition of intractability was met by 142 (23.2%). Intractability was delayed in 26 (31.3%) children meeting stringent and in 39 (27.5%) meeting the 2-drug definition. Intractability varied with the epilepsy syndrome and was delayed more often in focal than catastrophic (including encephalopathic) epilepsies (stringent: 46.2 vs 14.3%, p=0.003; 2-drug: 40.3 vs 2.2%, p<0.0001). Early remission preceded delayed intractability in 65.4 to 74.3% of cases. After developing intractability, 20.5% entered remission, and 13.3% were seizure-free at last follow-up. Referral to surgery may be delayed for 20 or more years, because of this interim period of remission. (Berg AT, Vickrey BG, Testa FM, et al. How long does it take for epilepsy to become intractable? A prospective investigation. **Ann Neurol** June 2006;60:73-79). (Respond: Anne T Berg PhD, Department of Biology, NIU, DeKalb, IL 60115).

COMMENT. In the earlier Dutch study of 453 children with newly diagnosed epilepsy followed prospectively for 5 years (Arts WFM et al. **Brain** 2004;127:1774-1784), significant variables for the worst outcome group included a symptomatic or cryptogenic etiology, early age at onset, and a history of febrile seizures. Of 108 (24%) patients with a terminal remission (TS) of <1 year, 27 had intractable seizures at 5 years. Of patients receiving 2 or more AEDs, almost 60% had a TR5 >1 year. AEDs were successfully withdrawn in 227 (59%). The course of epilepsy was constantly favorable in 51%, steadily poor in 17%, remitting after intractability in 25%, and deteriorating in 6% (**Ped Neur Briefs** August 2004;18:57-59).

VALPROATE-INDUCED HYPERAMMONEMIC ENCEPHALOPATHY

Valproate-induced hyperammonemic encephalopathy (VHE), predisposing causes, clinical, laboratory, and EEG findings, and therapy are reviewed from the Hospital del Mar, Barcelona, Spain. Urea cycle enzyme deficiency, especially ornithine transcarbamylase (OTC), is an inherited cause of hyperammonemia and a risk factor for developing VHE in patients taking VPA. Screening tests are recommended in patients with a known family history of OTC deficiency, and in patients who develop unexplained episodes of confusion, lethargy, and vomiting, and/or increased frequency of seizures, while on VPA therapy. Blood ammonia level, renal and liver function, and urinary orotic acid excretion should be tested. Blood VPA levels are within therapeutic ranges in most cases of VHE, and the dose of VPA and the height of the ammonia level are not related to VHE severity. Organic acidemias