

CDCs Emerging Infections Program showed a statistically significant association between nonadjuvant 2009 influenza A (H1N1) influenza vaccines and GBS. (Tokars JJ et al. *Pharmacoepidemiol Drug Saf* 2012 May;21(5):546-52). The RR varied between 2.1 and 3.0; attributable risks were 1.5 – 2.8 per million doses administered. In the 1976-1977 US epidemic, an unusually high rate of GBS followed the inactivated “swine” influenza A (H1N1) vaccination program. The variable risk of GBS with different H1N1 vaccines is an annual concern.

Increased vaccine-attributable risk for febrile convulsions following influenza vaccine. (Kelly H, Carcione D, Dowse GK, Effler P. *Pediatr Infect Dis J* 2012 Jul;31(7):792). Analysis of vaccine-attributable risk (VAR) was confined to children 6-59 months of age presenting to 9 Perth, Australia hospitals for management of a febrile convulsion in the 49 days between March 8, 2010, when the influenza vaccination program commenced and April 24, 2010, 48 hours after vaccinations for children were suspended. Ninety-nine children presented with febrile convulsions, 39 within 72 hours of receiving an influenza vaccine and 60 who had not been vaccinated. Of the 39 who had a febrile convulsion after influenza vaccination, 38 had received the 2010 vaccine, Fluvax. The total number receiving Fluvax in Perth metropolitan area was 11, 963. The 49-day risk of ED presentation for febrile convulsion following Fluvax was 38/11,963 or 32/10,000. The risk in those not receiving Fluvax was 7/10,000. The VAR as risk difference is 25/10,000. For the corresponding period in 2009 the VAR is estimated as zero. Further use of Fluvax was suspended in Australia.

MUSCLE DISORDERS

MYOTONIC DISORDERS IN AN EMG LAB OVER 12 YEARS

Researchers at the EMG Laboratory, Boston Children’s Hospital, MA assessed the spectrum of disorders associated with electrophysiologic myotonia in a pediatric electromyography laboratory. Of 2234 patients tested from 2000-2011 and screened retrospectively, 11 patients had myotonic discharges alone; 8 exhibited both myotonic discharges and myopathic motor unit potentials; and 54 demonstrated myopathic motor unit potentials alone. The diagnoses of patients with myotonic discharges alone included myotonia congenita (Thomsen disease) in 3, paramyotonia congenita in 2, congenital myopathy in 1, and Pompe disease (acid maltase deficiency) in 1; one mimicking SMA type 1 in infancy had Prader-Willi syndrome. The diagnoses of patients with both myotonic discharges and myopathic motor unit potentials included congenital myopathy in 2 and non-Pompe glycogen storage diseases in 3. The presence of myopathic potentials helps in the differential diagnosis. (Shah DU, Darras BT, Markowitz JA, Jones HR Jr, Kang PB. The spectrum of myotonic and myopathic disorders in a pediatric electromyography laboratory over 12 years. *Pediatr Neurol* 2012 Aug;47(2):97-100). (Respond: Dr Kang. E-mail: peter.kang@childrens.harvard.edu).

COMMENT. Channelopathies (myotonia congenita and paramyotonia congenita), glycogen storage diseases (primarily Pompe disease), and congenital myopathies are most commonly associated with EMG myotonic discharges in childhood.