

Ultrasound vs MRI for detecting intracranial hemorrhage in preterm neonates.

Investigators at Johns Hopkins Hospital studied 12 premature neonates with a mean gestational age of 32 weeks, comparing US and MRI for detection of grade I-III germinal matrix hemorrhage (GMH) and PVH. US had high sensitivity (100%) and specificity (93%) in detecting grade III GMH but poor sensitivity (0%) in detection of intraventricular hemorrhage (grade II GMH). US is first line of imaging for brain injury in the evaluation of premature neonates with suspected intracranial hemorrhage, but usefulness of MRI and susceptibility-weighted imaging for predicting long-term neurological outcome remains to be determined [1].

References.

1. Intrapiromkul J, et al. *J Neuroradiol.* 2013 May;40(2):81-8.

AUTOIMMUNE DISORDERS

RITUXIMAB IN AUTOIMMUNE CNS DISEASE

Investigators at University of Sydney, Australia, and 14 international centers assessed the utility and safety of rituximab in 144 children (median age 8 years, range 0.7-17; 103 female) with autoimmune and inflammatory disorders of the CNS. These included NMDAR encephalitis in 39 patients, opsoclonus myoclonus ataxia syndrome in 32, neuromyelitis optica spectrum disorder in 20, lupus erythematosus in 18, and other neuroinflammatory disorders in 35. A standardized questionnaire and Rankin Scale were used for a retrospective evaluation of treatment outcome. Infusion adverse events occurred in 18/144 (12.5%), including anaphylaxis in 3, and infection in 11 (7.6%), 2 of whom died. Benefit was reported in 125 (87%) patients, greater in patients treated early. The off-label use of rituximab should be restricted to disorders having significant morbidity and mortality. (Dale RC, et al. Utility and safety of rituximab in pediatric autoimmune and inflammatory CNS disease. *Neurology* 2014 Jul 8;83(2):142-50).

COMMENTARY. Suggested guidelines for rituximab treatment in children with neuroimmunologic conditions are listed in an editorial [1]. Originally approved by the FDA in 1997 for the treatment of B-cell non-Hodgkin lymphoma and later for rheumatoid arthritis, more recently it has been used in a variety of autoimmune disorders including multiple sclerosis.

References.

1. Chitnis T, Waubant E. *Neurology.* 2014 Jul 8;83(2):111-2.

ENCEPHALITIS / ENCEPHALOPATHY

DYSPLASTIC NEURONS IN OVARIAN TERATOMAS IN NMDAR ENCEPHALITIS

Investigators at University of Toronto, Canada, report detection of atypical (dysplastic) neuronal elements in 4 of 5 teratomas resected from cases with NMDAR

encephalitis but not in 39 controls. These atypical neurons resembled gangliogliomas (n=3) or ganglioneuroblastoma (n=1). Abnormal neuroglial elements were closely related to immune infiltrates resected from 4 of 4 cases. Abnormal neurons within teratomas distinguish cases with NMDAR encephalitis from controls and may promote the development of autoimmunity. (Day GS, et al. Abnormal neurons in teratomas in NMDAR encephalitis. *JAMA Neurology* 2014 Jun;71(6):717-24).

COMMENTARY. The authors propose that pathological examination of teratomas removed from patients with NMDAR encephalitis should first focus on areas containing CNS tissue and second on neurons closely approximated by inflammatory infiltrates. The colocalization of dysplastic CNS neurons and inflammatory infiltrates support an autoimmune cause for the clinical encephalitic presentation [1].

Broca's aphasia: a new phenotype of anti NMDAR encephalitis.

Investigators at Le Kremlin-Bicetre and centers in Paris, France, report a novel case of anti NMDAR encephalitis in a 4-year-old girl who presented with partial seizures that evolved to sudden and isolated Broca's aphasia and subsequently resolved. Anti NMDAR antibodies were positive in CSF and serum [2].

References.

1. Day GS, et al. *JAMA Neurol.* 2014 Jun;71(6):717-24.
2. Deiva K, et al. *Eur J Paediatr Neurol.* 2014 Jun 11. [Epub ahead of print]

ROTAVIRUS-ASSOCIATED ENCEPHALOPATHY AND SUD

Investigators at Fujita University School of Medicine, Toyoake, Aichi, Japan, performed a nation-wide survey to determine the incidence and clinical features of rotavirus-associated encephalitis/encephalopathy (RV-AE) and sudden unexpected death (SUD) in Japan. Of questionnaires sent to hospitals, 963 (70.5%) were returned, reporting 58 cases of RV-AE diagnosed by immunochromatography between 2009 and 2011 and 7 cases of SUD in the same period. Neurological sequelae occurred in 15/58 (25.9%) and fatal outcomes in 7/58 (12.1%); 36/58 (62.1%) had no sequelae. In patients with RV-AE, CSF pleocytosis was observed in 9/40 (22.5%) patients and protein levels were elevated in only 4/40 (10%). Serum sodium was normal in 40%, elevated in 17.8%, and low in 42.2%. CT showed brain edema in 9/34 (26.5%), and MRI was abnormal in 64.7% patients. Elevated lactic dehydrogenase (>500 IU/L) or acidemia (pH<7.15) was an indication of a poor prognosis. Annual incidence of RV-AE was 44 and SUD 4.9 cases. (Kawamura Y, et al. Nationwide survey of rotavirus-associated encephalopathy and sudden unexpected death in Japan. *Brain Dev* 2014 Aug;36(7):601-7).

COMMENTARY. In Japan, the annual number of cases of RV-AE was estimated at 41.1 and of SUD 5.0. In the US, a fatality rate of 1 death per 1616 rotavirus hospitalizations (0.06%) is cited in a 2007 report [1]. Since rotavirus vaccines became available in the US in 2006, ED visits and hospitalizations for RV related complications have decreased by 50,000 cases in children younger than 5 years [2].