

Chang Gung Medical Journal

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REVIEW ARTICLES

1 Neonatal Vitamin-responsive Epileptic Encephalopathies

Sidney M. Gospe, Jr., MD, PhD

Certain rare vitamin-responsive inborn errors of metabolism may present as a neonatal encephalopathy with intractable (anticonvulsant-resistant) seizures. This review discusses our current understanding of three neonatal vitamin-responsive epileptic encephalopathies: pyridoxine-dependent seizures, folinic acid-responsive seizures, and pyridoxal phosphate-dependent seizures. A diagnostic and treatment protocol is proposed.

13 Heme Oxygenase-1 in Cardiovascular Diseases: Molecular Mechanisms and Clinical Perspectives

Chao-Yung Wang, MD; Lee-Young Chau, PhD

Heme oxygenase-1 (HO-1) catalyzes the rate-limiting step in the conversion of heme to iron, carbon monoxide, biliverdin and its subsequent metabolite, bilirubin, in mammalian cells. In addition to its well-defined role in heme catabolism and erythrocyte turnover, HO-1 also has important functions in various disease states, including coronary artery diseases, cardiac hypertrophy, diabetes mellitus, ischemic/reperfusion injury, atherosclerosis and cancer. This review provides an overview on the regulation and function of HO-1 from molecular mechanisms to various clinical perspectives, with particular emphasis on cardiovascular diseases.

ORIGINAL ARTICLES

25 Characterization of the Response of Dendritic Cells and Regulatory T Cells to Tumor Antigens in Patients with Renal Cell Carcinoma

Ming-Mo Hou, MD; John Wen-Cheng Chang, MD; See-Tong Pang, MD, PhD;
Yang-Jen Chiang, MD; Yung-Chi Shen, MS; Shuen-Kuei Liao, PhD; Jia-Juan Hsieh, MS;
Kun-Yun Yeh, MD, PhD; Nai-Jen Chang, MD; Cheng-Keng Chuang, MD, PhD

This study characterized dendritic cells (DCs), regulatory T cells (Tregs) and the immune responses to tumor antigens in renal cell carcinoma (RCC) patients. The RCC-DCs were as efficient as DCs from healthy donors in stimulating the proliferation of allogeneic T cells, but these RCC-DCs were less efficient in stimulating autologous T cells. In RCC patients, Tregs inhibited autologous T cell proliferation rather than allogeneic T cell proliferation in response to TuLy-DCs stimulation. Patients with RCC have less potent anti-tumor immune responses.

36 Comparing Late-onset and Neonatally-diagnosed Congenital Cystic Adenomatoid Malformation of the Lung

Sui-Ling Liao, MD; Shen-Hao Lai, MD; Chuen Hsueh, MD; Kin-Sun Wong, MD

Congenital cystic adenomatoid malformations (CCAM) are rare malformations mostly diagnosed during the neonatal period. Neonates with CCAM often present with respiratory distress. Spontaneous resolution may occur postnatally, and therefore expectant management seems justified. Lower airway infections were the most common clinical manifestations of lesions that persisted to later ages. Due to infrequent regression of the lesion beyond the neonatal period, surgical resection is recommended.

44 Oculopharyngeal Muscular Dystrophy - A Genetically Verified Taiwanese Family

Chia-Ling Huang, MD; Shey-Lin Wu, MD; Szu-Chia Lai, MD; Chin-Song Lu, MD;
Yah-Huei Wu-Chou, PhD

Oculopharyngeal muscular dystrophy (OPMD) is a rare inherited muscular disorder characterized by late-onset, slowly progressive bilateral ptosis, dysphagia, and possible proximal limb weakness. A short polyalanine expansion in the polyadenylate binding-protein nuclear 1 (*PABPN1*) gene is a commonly reported mutation.

Here, we report a large family with 12 affected members whose phenotypic characteristics and genetic information confirmed the diagnosis of OPMD. We suggest that genetic intervention should be undertaken to understand the genetic epidemiology and provide counseling for carriers of OPMD in Taiwan.

51 Clinical Outcome of Signet Ring Cell Carcinoma and Mucinous Adenocarcinoma of the Colon

Jinn-Shiun Chen, MD; Pao-Shiu Hsieh, MD; Jy-Ming Chiang, MD; Chien-Yuh Yeh, MD; Wen-Sy Tsai, MD; Reiping Tang, MD; Chung-Rong Changchien, MD; Ren-Chin Wu, MD

We evaluate the clinicopathologic features and prognosis of colon cancer with different distribution of mucin in pathology. From 1995 to 2003, 45 signet ring cell carcinoma (SCC) patients and 332 non-SCC mucinous adenocarcinoma (MC) patients were identified and compared with those of 2984 consecutive nonmucinous adenocarcinoma (NMC) patients in the same period. The prognosis of SCC is poorer than that of MC and NMC. More locally advanced, less hematogenic spread, almost no early detected, and very poor surgical outcomes was found in SCC. The role of resection for late stage SCC should be carefully evaluated.

58 Clinical Assessment of Patients with Cervicogenic Headache: A Preliminary Study

Jia-Pei Hong, MD; Cheng-Hsiu Lai, PhD; Yin-Chou Lin, MD; Shih-Wei Chou, MD, PhD

Traditionally, the diagnostic criteria of cervicogenic headache (CEH) are mainly based on subjective symptoms, thus rendering its differential diagnosis difficult. We recruited 22 patients with subjectively diagnosed CEH and 14 healthy subjects as controls. All received plain cervical radiograms including lateral flexion, neutral, and extension views. The degree of localized kinking was measured to define the level of cervical malalignment. Results showed that localized kinking in the lower cervical spine was significantly different between the study and control groups. On the questionnaire, nasal symptoms were among the most common clinical manifestations. The concept of "cervicogenic cephalic syndrome" is discussed.

67 Hand-assisted Laparoscopic Splenectomy - Preliminary Experience in Southern Taiwan

Wei-Feng Lee, MD; Shih-Chung Wu, MD; Chee-Chien Yong, MD; Chao-Long Chen, MD; Chih-Chi Wang, MD

We report our preliminary experience with hand-assisted laparoscopic splenectomy in patients with splenomegaly. Although the large splenic size made the technique difficult, the procedure was performed successfully in all 5 cases. The benefits of laparoscopic surgery were retained, and there was no mortality and morbidity.

73 The Role of Endoscopic Ultrasonography Examination for Evaluation and Surveillance of Gastric Subepithelial Masses

Cheng-Hung Chien, MD; Rong-Nan Chien, MD; Cho-Li Yen, MD; Kua-Meen Fang, MD; Ching-Jung Liu, MD; Chih Lang Lin, MD; Jia-Jang Chang, MD; Li-Wei Chen, MD; Tsung-Shih Lee, MD; Shuo-Wei Chen, MD; Ching-Chih Hu, MD; Liang-Che Chang, MD

We used endoscopic ultrasonography (EUS) to evaluate gastric subepithelial masses and their malignant potential. Pathological findings confirmed that 73.3% of larger tumors (> 30 mm) were accurately diagnosed. Most small gastric subepithelial masses (< 30 mm) did not exhibit changes during follow-up. If the tumor size increases or tumor ultrasonographic features suggest malignant possibility during EUS surveillance, surgical resection should be considered.

82 Mental Health of Parents Having Children with Physical Disabilities

Jen-Wen Hung, MD; Yee-Hwa Wu, OTR; Yi-Chien Chiang, RN, PhD; Wen-Chi Wu, OTR; Chao-Hsing Yeh, RN, PhD

This study aims to investigate the mental health of parents of children with physical disabilities and to elucidate the factors affecting parental mental health. This study was a cross-sectional study in which we recruited parents who had a physically disabled child. Parental mental health was evaluated as the main outcome measure using the 12-item version of the Chinese Health Questionnaire (CHQ-12). Of the 91 parents, 43.98% were categorized as cases on the CHQ. The most significant predictor of overall parental mental health was parental distress.

92 Cross-talk between Bradykinin and Epidermal Growth Factor in Regulating IL-6 Production in Human Airway Smooth Muscle Cells

Po-Hao Feng, MD; Te-Chih Hsiung, MD; Han-Pin Kuo, MD, PhD; Chien-Da Huang, MD

Bradykinin (BK) induces interleukin (IL)-6 expression in airway smooth muscle (ASM) cells by involving extracellular signal-regulated kinase 1/2 (ERK1/2) signaling pathway. Our aim is to study whether there is cross-talk between BK and epidermal growth factor (EGF) in regulation of IL-6 gene expression. The data show that BK and EGF act in concert to regulate the expression of IL-6 in ASM cells possibly via transcriptional mechanisms involving epidermal growth factor receptor (EGFR)-associated key signaling molecules.

CASE REPORTS

100 Erlotinib-associated Near-fatal Interstitial Pneumonitis in A Patient with Relapsed Lung Adenocarcinoma

Chun-Liang Chou, MD; How-Wen Ko, MD; Chih-Wei Wang, MD; Chih-Teng Yu, MD; Han-Pin Kuo, MD, PhD; Chien-Da Huang, MD

Erlotinib (Tarceva®) is a human epidermal growth factor receptor (EGFR) tyrosine kinase inhibitor used for treatment of locally advanced or metastatic non-small cell lung cancer (NSCLC) after failure of at least one prior chemotherapy regimen. We deem this to be the first case report in the English language of a histologically-confirmed case of near-fatal interstitial pneumonitis associated with erlotinib in East Asian patients. This case suggests that the outcome of erlotinib-associated pulmonary toxicity with acute respiratory failure may be favorable with adequate early management.

106 Intramuscular Lorazepam in Catatonia in Patients with Acute Renal Failure: A Report of Two Cases

Chao-En Huang, MD; Tiao-Lai Huang, MD

We describe two renal-insufficient patients with catatonia who responded well to intramuscular lorazepam in order to relieve catatonic symptoms. Case 1 involved a patient with end-stage renal disease and severe pneumonia related respiratory failure. He responded well and the catatonia was relieved with intramuscular lorazepam (total dose, 4 mg). Case 2 involved a patient with alcoholic liver cirrhosis and rhabdomyolysis-related acute renal failure. He showed a marked improvement and the catatonia was relieved with intramuscular lorazepam (2 mg). In conclusion, intramuscular lorazepam may play an important role in the treatment of catatonia associated with renal insufficiency.

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