Good control HbA1c, without or less Diabetic chronic complications

Dr. CHOI HONG
Department of Internal Medicine
CHCSJ

BACKGROUND:
• Diabetes mellitus is a worldwide problem. In 2000, according to the World Health Organization, at least 171 million people worldwide suffer from Diabetes. Its incidence is increasing rapidly, and it is estimated that by 2030, this number will almost double. There are more than 40,000 people suffering from Diabetes in Macau. It is very important to prevent or delay Diabetic chronic complications.
• The glycolated hemoglobin (HbA1c) is a very important blood test to determine control of Diabetes. It provides an average blood glucose level during the past 6 to 12 weeks.
• The ideal range for DM patients is generally less than 6.5%.

METHODS:
• I chose 89 DM patients of my consultation (Female: 56 patients, Male: 33 patients) to join the research. They are the following conditions:
  – They are Type II DM patients
  – They suffer from DM over 10 years
  – I follow them in my consultation about 4 or 5 months
  – Their HbA1c is <7% every consultation
  – I check their Fundoscopy and Microalbuminuria from time to time.
• I count the morbidity of DM Nephropathy and DM Retinopathy in the above 89 patients whose HbA1c <7%.

RESULTS:
• Without DM Nephropathy or DM Retinopathy or Both (DM Nephropathy + DM Retinopathy):
  Totally 64 patients: 71.9%, including: Female: 40 patients (71.4%), Male: 24 patients (72.7%)
• With DM Nephropathy or DM Retinopathy or Both (DM Nephropathy + DM Retinopathy):
  Totally 25 patients: 28.1%, including: Female: 14 patients (25.0%), Male: 11 patients (33.3%)
• Only with DM Nephropathy:
  Totally 14 patients: 15.7%, including: Female: 9 patients (16.1%), Male: 5 patients (15.2%)
• Only with DM Retinopathy:
  Totally 8 patients: 9.0%, including: Female: 4 patients (7.1%), Male: 4 patients (12.1%)
• With Both (DM Nephropathy + DM Retinopathy):
  Totally 3 patients: 3.4%, including: Female: 1 patient (1.8%), Male: 2 patients (6.1%)

CONCLUSIONS:
• During the research, even the patients’ HbA1c are <7% but 28.1% of them still have Diabetic chronic complications including DM Nephropathy or DM Retinopathy or Both (DM Nephropathy + DM Retinopathy). Diabetes relates health and social-economic problems in the world. The higher the HbA1c, the higher the risks of developing Diabetic chronic complications. Good management of Diabetes and prevention of its chronic complications are very important.
Postprandial Hypotension
By IP KAR HUNG
Department of Internal Medicine, CHCSJ, Macau

Postprandial hypotension (PPH) is a common but under-recognized condition among older adults. It was first described in 1977 in a patient with severe Parkinson disease, but a later study has shown that the prevalence in institutionalized elders is approximately 25%-38%, although one study conducted in a Dutch hospital reported a prevalence as high as 67%.

It’s Definition
Classically, postprandial hypotension is defined as a decrease in SBP of ≥20 mm Hg or a decrease below 90 mm Hg from ≥100 mm Hg within 2 hours after a meal.

Why is it important?
PPH has been associated with syncope, falls, and even coronary events and stroke. One study found that 23% of hospitalized elderly patients with either syncope or falls experienced PPH. A prospective study of patients with essential HTN demonstrated that 83% of elderly hospitalized patients with postprandial decreases in Bp of 10 mm Hg or more had evidence of lacunae on MRI, as compared with 44% of patients without such decreases. Coronary events and overall mortality also are associated with PPH.

Who is more likely to have PPH?
PPH is common among patients with DM, essential HTN, end-stage renal disease undergoing HD. Patients with Parkinson disease are at particular risk. There are case reports of occurring in patients with Fragile X mutation and Shy-Drager syndrome. It occurs even in the healthy elderly. In a study of 21 community-dwelling elderly adults, the mean postprandial decrease in SBP was 11 mm Hg, and 2 patients (~5%) had a decrease of 20 mm Hg or more.

What precipitate PPH?
Carbohydrate-rich meals predispose patients to more immediate decreases in Bp compared with protein or fat rich meal, possibly secondary to increased amounts of insulin and its corresponding vasodilator effect. Warm meals (50 C) appear to cause a greater decrease in postprandial Bp than meals served cold (5 C). Diuretics, particularly furosemide, have been shown to exacerbate PPH.

Is there difference between meals in a day?
PPH can occur after any meal, but breakfast and lunch appear to be associated with the most pronounced decrease in Bp. It is possibly secondary to baroreflex dysfunction associated with increased premeal SBP.

Pathophysiology
The mechanism has yet to be fully delineated. Splanchnic dilatation after a meal is the most important factor. Sympathetic activity after a meal should increase 2-3 times to prevent PPH. An inadequate postprandial increase in cardiac output can be due to an impairment of baroreflex function and inadequate compensation of the sympathetic NS. An increased release of vasodilatory gastrointestinal peptide-like calcitonin gene-related peptide (CGRP) may play a role. PPH is also related to the rate of glucose entry into the duodenum.

How to Diagnose PPH?
Postprandial hypotension is defined analogously to orthostatic hypotension as a decrease in blood pressure of ≥20 mm Hg. Monitoring of Bp and symptoms should continue for 2 hours after a meal. The maximum decrease in Bp typically occurs within the first 35 minutes to 1 hour after a meal.

Are there treatment for PPH?
Nonpharmacologic Modifications
Drinking 350-480 mL water before a meal can attenuate the decrease in Bp by as much as 20 mm Hg in patients with autonomic failure. Autonomic failure patients should eat 6 small meals of equal caloric content rather than 3 large meals. Maintain a sitting or recumbent position after a meal, unless they are walking, in which case they should once again resume a recumbent position immediately upon stopping.

Pharmacotherapy
Caffeine
Caffeine, an adenosine receptor antagonist, may ameliorate postmeal Bp decreases when coffee or tea is given before a meal, however, data about its effects are inconsistent. It may be worth trying for symptomatic patients, titrating from 60 to 200 mg (approximately 2 cups) before meals.

Alpha-glucosidase Inhibitors
Alpha-glucosidase inhibitors such as acarbose or voglibose have been shown to diminish the decrease in postprandial Bp. Their effects might reflect alterations in circulating vasodilators and in the amounts of gut peptides secreted.

 Guar
Guar gum is derived from the guar bean and acts as a bulking agent. It may prevent PPH by slowing glucose absorption.

 Octreotide
Octreotide has been shown to alleviate symptomatic Bp decreases in elderly patients, hypertensives, and in those with autonomic failure, perhaps by increasing splanchnic and peripheral vascular resistance.

Conclusion
Postprandial hypotension may be an important causative factor in elderly patients with unexplained syncope. It is common in older adults, especially the institutionalized elderly, and is associated with significant morbidity and mortality. Falls, syncope, strokes, transient ischemic attacks, angina, and myocardial infarctions can result, and PPH is an independent predictor of mortality. Prompt diagnosis and treatment with lifestyle adjustments such as drinking water before meals, avoiding diuretics, and eating smaller, more frequent meals might prevent recurrent syncope, further ischemic insults, unnecessary testing, and anxiety.
LEONG Wai I, Su Mei-Fang*  
(Neurology) Internal Medicine Department of C.H.C.S.J.

**Neurocysticercosis:**  
A case report 病例報告

---

- **Abstract** We report a case of Neurocysticercosis, an M/82y who admitted to hospital due to severe impaired cognitive function. Brain CT+MRI showed multiple high density images (within 1cm) scattered in the cortical of bilateral frontal temporal lobes with slightly peripheral edema. Lower limbs x-ray had shown small calcification in the lower leg muscle. Because clinically highly suspected Neurocysticercosis, Praziquantel was prescribed and improved cognition on the fifth day of treatment. Brain CT followed up had shown notable diminished brain lesion. We discussed the differential diagnosis and management.

- **Key words** Neurocysticercosis, Praziquantel

---

**CASE**  
The patient is an M/82y Chinese, married, primary school education, retired tailor, non-smoker and social drinker. He was born in China, stayed in Vietnam for 4 years since his age of 16y, and immigrated to Hong Kong afterwards until 70y of age, when he resided in Macau since then. No history of taking under-cooked pork.

He was healthy and ADL-independent until Nov. 2006 when he appeared dull and could not handle his daily life. He was sent to our hospital March 2007 for management. Clinically, he presented with severe impaired cognitive function (MMSE 8) with full muscle power and negative Babinski sign. His routine blood test, tumor marker, CXR, abdominal ultrasound and CSF analysis were unremarkable.

25/3/2007 brain CT: multiple high density images (within 1cm) scattered in the cortical of bilateral frontal temporal lobes. No remarkable contrast enhancement is noted. But slightly peripheral edema is noted of those high density images. Brain MR: those above images present with slightly peripheral hypo-signal intensity in T1WI, slightly hyper-signal intensity in T2WI. The central of those images is hypo-signal in T1WI, T2WI. Slightly peripheral edema is noted. No sign to suggest cerebral hematomas.

- Lower limbs x-ray had shown multiple small calcification in the lower leg muscle. However, antibodies for cysticercosis (Taenia sodium) was not available in Macau.

Since neurocysticercosis was highly suspected, praziquantel (150mg tid x 2d -> 300mg bid x 8d) was prescribed for 10 days(1)(2). On the day fifth of treatment, the patient had improved cognition (MMSE 15) and he was able to walk by himself. Brain CT on 17/04/2007 had shown notable diminished brain lesion. We discussed the differential diagnosis and management.

**DISCUSSION**  
The patient diagnosis base on the image showed a small, spherical, ring enhancing lesion with a regular margin located at the cortico-medullary junction with slightly peripheral edema(3). The differential diagnosis needs to excluded metastases, cerebral abscesses, subacute infarction or resolving hematoma.

Metastases usually located in corticomedullary junction, can enhance in a ringlike pattern, and often more edema.  
Cerebral abscesses may also located at corticomedullary junction and the thin, regular capsule formation that appears hypointense on T2-weighted images(4). Causes include bacterial, fungal, or granulomatous agents. Parasitic infection, such as Neurocysticercosis infection, can also be considered.

Less likely, a subacute infarction or resolving hematoma of the cortex rarely enhances in a ring patten.

Under the clinical and imaging study, highly suspected the patient suffered from neurocysticercosis. So began the treatment of Praziquantel. Because his old age, we modificated the therapy dose to 150mg tid x 2d -> 300mg bid x 8d. His cognitive function (from 8→15) and Brain CT improved after treatment and the diagnosis was clinically confirmed.

In summary, we report a case (first report in Macau) of Neurocysticercosis in a man who admitted to hospital due to severe impaired cognitive function. Brain CT+MRI showed multiple high density images (within 1cm) scattered in the cortical of bilateral frontal temporal lobes with slightly peripheral edema. Lower limbs x-ray had shown small calcification in the lower leg muscle. Because clinically highly suspected Neurocysticercosis, Praziquantel was prescribed and improved cognition on the day fifth of treatment.

**REFERENCES**  
兩種聚乙二醇干擾素聯合利巴韋林治療1b及6a亞型慢性丙型肝炎的療效

李德明
澳門仁伯爵綜合醫院

摘要
慢性丙型肝炎在澳門的發生率約為1.2%，雖然遠遠低於乙型肝炎，但其危害性卻不容忽視。而且其治療難度較大，副作用多，對治療方案的個體化要求較高。因此我們回顧總結了仁伯爵醫院171例慢性丙型肝炎患者的臨床資料，對兩種聚乙二醇干擾素聯合利巴偉林在治療1b及6a亞型慢性丙型肝炎的療效方面進行了比較，希望能為治療藥物選擇方面提供一些參考。
Early Detection of Central Airway Lung Cancer among Smokers with Silicosis

Lo Iek Long, Chan Hong Tou, Mok Tin Hou, Iam Lap Fong, Maria Teresa, Cheong Tak Hong, Lam Bing*

Department of Respiratory Medicine, CHCSJ, Macao SAR, China
* Respiratory Medicine Centre, Hong Kong Sanatorium & Hospital, Hong Kong SAR, China

**Introduction:** Smokers with silicosis are at increased risk of lung cancer. We aim at evaluating the feasibility of using Autofluorescence Imaging (AFI) after sputum examination for the early detection of large airway lung cancer and factors associated with the presence of cancerous and pre-cancerous lesions among smokers with silicosis.

**Methods:** Subjects were recruited from the Pneumoconiosis Clinic if they fulfilled the following criteria: 1) ≥ 40 years old, 2) ≥ 20 pack-years smoking history, and 3) confirmed diagnosis of silicosis. Sputum specimens were collected for cytology / cytometry examination and Autofluorescence Bronchoscopy would be performed in subjects with abnormal sputum result.

**Results:**
- 48 subjects were recruited during the study period. The mean age and smoking history were 63 ± 10 years and 51 ± 30 pack-years respectively. Other clinical characteristics of this cohort were shown in Table 1.
- Intraepithelial lung cancers and pre-neoplastic lesions (squamous metaplasia or above) were detected in 2 (4.2%) and 14 (29.2%) subjects respectively.
- The proportion of current smoker (75.0% vs 40.6%, P=0.03) and asbestos exposure (37.5% vs 9.4%, p=0.04) were significantly higher in subjects harboring these lesions compared with those without (Table 2).

**Conclusions:** Sputum examination followed by AFI might be a useful way in identifying cancerous / precancerous lesions among smokers with silicosis. Current smoker and asbestos exposure were associated with the presence of these lesions.

**Key words:** Lung cancer; silicosis; Autofluorescence Bronchoscopy; early detection

<table>
<thead>
<tr>
<th>Characteristics</th>
<th>N</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age, years (range)</td>
<td>63 ± 10 (48-82)</td>
</tr>
<tr>
<td>Current smoker (%)</td>
<td>25 (52.1)</td>
</tr>
<tr>
<td>Pack-years</td>
<td>51 ± 30</td>
</tr>
<tr>
<td>Exposure to asbestos (%)</td>
<td>9 (18.8)</td>
</tr>
<tr>
<td>Spirometric result</td>
<td></td>
</tr>
<tr>
<td>FEV1, % predicted</td>
<td>81 ± 22</td>
</tr>
<tr>
<td>FVC, % predicted</td>
<td>98 ± 16</td>
</tr>
<tr>
<td>FEV1/FVC ratio, %</td>
<td>63 ± 13</td>
</tr>
<tr>
<td>Airflow limitation (%)</td>
<td>32 (66.7)</td>
</tr>
<tr>
<td>Sputum cytology abnormality</td>
<td></td>
</tr>
<tr>
<td>Atypical cells (%)</td>
<td>37 (77.1)</td>
</tr>
<tr>
<td>Squamous metaplasia or above (n=16)</td>
<td></td>
</tr>
<tr>
<td>Normal to hyperplasia (n=32)</td>
<td></td>
</tr>
</tbody>
</table>

Data are expressed as Mean ± SD and number (%) if not specified; FEV1: forced expiratory volume in one second; FVC: forced vital capacity; Airflow limitation was defined as a FEV1 to FVC ratio of less than 70%

**Table 2 - Comparison of clinical characteristics between the two groups**

<table>
<thead>
<tr>
<th>Variables</th>
<th>Squamous metaplasia or above (n=16)</th>
<th>Normal to hyperplasia (n=32)</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age, years</td>
<td>62.3 ± 11.6</td>
<td>64.1 ± 9.7</td>
<td>0.56</td>
</tr>
<tr>
<td>Old tuberculosis, n (%)</td>
<td>3 (18.8)</td>
<td>15 (46.9)</td>
<td>0.06</td>
</tr>
<tr>
<td>Other malignancy, n (%)</td>
<td>1 (6.3)</td>
<td>0 (0.0)</td>
<td>0.33</td>
</tr>
<tr>
<td>Current smoker, n (%)</td>
<td>12 (75.0)</td>
<td>13 (40.6)</td>
<td>0.03</td>
</tr>
<tr>
<td>Smoking, pack-years</td>
<td>54.7 ± 42.4</td>
<td>49.2 ± 22.3</td>
<td>0.56</td>
</tr>
<tr>
<td>Exposure to asbestos, n (%)</td>
<td>6 (37.5)</td>
<td>3 (9.4)</td>
<td>0.04</td>
</tr>
<tr>
<td>Profusion score, n (%)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>1 (including 0/1, 1/0, 1/1)</td>
<td>9 (56.3)</td>
<td>20 (62.5)</td>
<td>0.46</td>
</tr>
<tr>
<td>2 (including 1/2, 2/1, 2/2)</td>
<td>7 (43.8)</td>
<td>10 (31.3)</td>
<td></td>
</tr>
<tr>
<td>3 (including 2/3, 3/2, 3/3)</td>
<td>0 (0.0)</td>
<td>2 (6.3)</td>
<td></td>
</tr>
<tr>
<td>FEV1, % predicted</td>
<td>77.9 ± 23.4</td>
<td>82.1 ± 21.1</td>
<td>0.54</td>
</tr>
<tr>
<td>FVC, % predicted</td>
<td>87.6 ± 16.7</td>
<td>97.6 ± 15.6</td>
<td>0.99</td>
</tr>
<tr>
<td>FEV1/FVC ratio, %</td>
<td>61.8 ± 13.9</td>
<td>64.1 ± 11.7</td>
<td>0.54</td>
</tr>
<tr>
<td>Airflow limitation, n (%)</td>
<td>11 (68.8)</td>
<td>21 (65.6)</td>
<td>0.83</td>
</tr>
</tbody>
</table>

Data are expressed as Mean ± SD and number (%) if not specified; FEV1: forced expiratory volume in one second; FVC: forced vital capacity; Airflow limitation was defined as a FEV1 to FVC ratio of less than 70%
Fever of Unknown Origin
Dr. Pun Weng Hong
Department of Internal Medicine
CHCSJ

Case Report
Mr. H; 52 yrs / M
- Electricity technician
- Good past health
- Denied recent travel/ wide animal contact/ insect bite/ toxic substance contact/ raw food intake history
- Asked for transferred from private hospital after 2 weeks of treatment
- Denied recent travel/ wide animal contact/ insect bite/ toxic substance exposure

Mr. H, 52 yrs / M

Good past health

Arrived at our ER on 28/Jun with fever and only signs of hepatosplenomegaly. Only history of fever with no other symptoms.

Further deterioration:
- Liver function: significant increase in bilirubin, AST/ALT, and alkaline phosphatase
- Cardiac echo: rupture of the anterior mitral valve leaflet
- Skin biopsy: non-specific change
- Liver biopsy: granulomatous inflammation
- 16/July PET CT scan showed diffuse liver uptake with increased metabolic activity
- Echocardiography showed no vegetation, normal structure and function

Further workup:
- HAV/HEV/CMV recent infection: no evidence
- HIV/VDRL: -ve, urine Legionella Pneumophila Ag: -ve
- CMV: pp65: 1 cell per 2x10^5 WBC
- Mantoux test: -ve
- Widal test: -ve
- HIV/VDRL: -ve, urine Legionella Pneumophila Ag: -ve

Diagnosis:
- Acute Q fever confirmed by vaccination
- Isolation of C. burnetii by culture
- C. burnetii IgG >=1:800 (while phase II IgG elevated as well)
- Isolation of C. burnetii DNA +ve via amplification by PCR assay

Probable case of Acute Q fever
- Clinically compatible case
- Isolated prozone reaction with more high grade fever to 40C with chills
- Progressive normocytic anemia and hypoalbuminemia, persistent coagulopathy and progressive liver dysfunction
- Bone marrow biopsy showed few histiocytosis and increased amount of megakaryocytosis together with hypercellularity, suggests reactive changes

Further workup:
- Skin biopsy over R trunk rash: non-specific change
- Isolated prolongation of APTT, anti-cardiolipin +ve
- Liver biopsy on 7/Aug: granulomatous inflammation
- Tryptophan and fever response
- Repeated Coxiella burnetii shown +ve (as followed)

Provisional Diagnosis:
- Acute Q fever complicated with hepatitis, endocarditis, lupus anti-coagulant +ve and indocinocytosis
- Treatment planned as doxycycline for 3 yrs with hydroxychloroquine 1.5yrs

Follow up in clinic with good condition, normal LFT/clothing/inflammation markers, negative in anti-cardiolipin, good control in cardiac echo follow up.

Q fever
- Described in 1935 by E.H. Derrick, termed "query fever"
- Isolated by Burnet and Freeman in 1937
- Pathogen isolated from ticks in Montana in 1938 and officially named Coxiella burnetii in 1948
- During the 2007-2010 outbreak in the Netherlands persons living within 5 km of dairy goat farms (abortion waves in goats confirmed as primary source of human infection)

Coxiella burnetii
- Gram-negative cocobacillus
- Intracellularly replicates in macrophages and monocytes
- Resistant to heat, low or high PH, 0.5% sodium hypochlorite, UV irradiation, and environmental conditions, such as desiccation, extreme temperatures and sunlight due to resistance to a spore-like stage, survive for 7-10 mths on wool at 15-20C, >1mth on fresh meat in cold storage, for 40 mths in skim milk at room temperature
- 2 antigenic forms: phase I/II
- Infected by inhalation of aerosols or contaminated dusts
- Acute infection: fever, rigors, myalgia, malaise, retrobulbar headache, fatigue, night sweats, dyspnea, confusion, nausea, diarrhea, abd. pain, non productive cough or chest pain. Severe disease include acute hepatitis, atypical pneumonia and meningooencephalitis. Lab. data found liver enzyme elevation, leukocytosis and thrombocytopenia
- Chronic infection: infection persists >6 months, complicated with fatal endocarditis, osteomyelitis, osteoarthritis and pneumonitis

Case Classification:
- Acute Q fever confirmed clinically compatible case AND paired sera with >= fourfold change in C burnetii IgG phase II by IFA (3-6weeks apart); or C. burnetii DNA +ve via amplification by PCR assay; or Isolation of C. burnetii by culture
- Probable case of Acute Q fever clinically compatible case for acute disease who not meet any of above lab criteria but has a single IFA IgG titer >= 1:128
- Chronic Q fever confirmed clinically compatible case
- Acute Q fever confirmed by vaccination
- Isolation of C. burnetii by culture

Probable case of Chronic Q fever
- Clinically compatible case for chronic disease who not meet any of the above lab criteria but has a single IFA IgG titer >= 1:128 or <1:800 by IFA
- Demonstrated of C. burnetii Ag in clinical specimen by IHC; or Isolation of C. burnetii by culture

Probable case of Chronic Q fever
- Clinically compatible case for chronic disease who not meet any of the above lab criteria but has a single IFA IgG titer >= 1:128 or <1:800 by IFA

Note: samples from suspected chronic patients should be evaluated for IgG titers to both phase I and II Ag.

Treatment:
- Most cases of acute Q fever will resolve without antibiotic treatment
- Acute infection: Tetracycline 500mg q6h po x 5-7 days
- Doxycycline 100mg q12h po x 5-7 days
- Or combination of erythromycin/rifampin

Chronic infection: esp. involving endocarditis, requires extended treatment
Motor neuron disease (MND)—a case report  WAN Chun, LEONG Wai I (Neurology) Internal Medicine Department of C.H.C.S.J.

• 57 years old female patient, Chinese, Macau resident
• Presented face and left arm numbness for half year since August of 2008. Accompanied with gradually upper limbs atrophy, insomnia and constipation.
• Past history of post operation of colon polyps and goiter
• Denied she’d special family history
• Brain CT scan on 16/03/2009 showed no significant finding
• EMG of Canton hospital indicated extensive neurogenic lesion
• Motor neuron disease (MND) was diagnosed
• Riluzole (50mg P.O. Bid) was initially prescribed by Neurologist of Canton hospital since May of 2009
• Progressive dysphagia occurred since March of 2009 and the patient was referred to Gastroenterology outpatient clinic to follow up
• The above patient was admitted in K.W. hospital on 24/10/2009 for shortness of breath and ABG indicated respiratory failure type II. She was subsequently intubated with mechanical ventilation. Then transferred to ICU of C.H.C.S.J. and received tracheostomy on 04/11/2009 due to intolerance to withdraw MV. Finally admitted in Neurology ward for supportive care including Bipap and physiotherapy etc.

MND, is also known as Amyotrophic lateral sclerosis (ALS), Charcot's disease or Lou Gehrig’s disease, is a disease of unknown cause characterized by slowly progressive degeneration of upper motor neurons (UMNs) and lower motor neurons (LMNs). They are a direct consequence of muscle degeneration via destruction of the anterior horn cell. There is no specific test for MND. Investigations are carried out to exclude other diseases. MND is clinical diagnosis and eventually fatal because of respiratory muscle weakness. Aspiration pneumonia, respiratory failure and other complications of immobility contribute to morbidity. Early diagnosis and associated management demonstrate very helpful to the patients’ life quality and their families, even the MND is irremediable.

上述內容介紹了仁伯爵綜合醫院 神經內科收治之本地一例運動神經元病患。運動神經元疾病又稱肌萎縮性脊髄側索硬化，Charcot病或Lou Gehrig病；是一種病因尚未明確的漸進性運動神經退化症，好發於40歲至50歲的中年人，大約每10萬人中就有5個人可能患病。男女罹患的比例約為1.5比1。當上運動神經元發生病變時，會產生肌肉僵直、反射増強；而下運動神經元病患，則以肌肉萎縮的症狀為主，患者最終會死於吸入性肺炎及呼吸衰竭等併發症。這種疾病目前仍未有特異性診斷方法，而且治療以支持療法為主；雖然患者的平均存活時間只有三年左右，但及早診斷和相關處理對患者之生活質量，以及他(她)們的家人仍有一定的幫助。
Henoch Schölein purpura

Wong Sio Mui, Chan Chio Peng

CHCSJ

Case Summary

- 33 years old
- Female
- History:
  - Normal delivery, first baby at 5/2006
  - Allergy to Borfizin and tetracycline antibiotic
  - Subclinical hypothyroidism
  - GPT 56, GGT 54

- Her skin lesion getting worse with arthritis and general edema after delivery at 5/2006, even with prednisolone 30mg/day
- Admitted again at 26/12/2006 due to clinical symptom getting worse:
  - Blood test no special finding
  - Skin biopsy done again showed compatible with LE, ASK, CREST, LAM, SLE, and SLE-like features
  - Increased the Prednisolone 30mg + 25mg/day
  - Consulted Rheumatologist and suggested add low-dose aspirin and NSID
- The patient’s clinical symptom improved and steroid tapered down the steroid dose till stop, and only keep cholecalciferol

- 1st admission during 28/1-2/2005
  - Both lower limbs recurrent purpura for 4 months after delivery. 1st baby: w. abdominal pain
  - Her clinical symptom improved after steroid treatment, but recur under after stopped the steroid
  - Blood analysis:
    - HEP: coagulation time: RPT: LFT: N
    - Ferrum and hepatitis marker: N
    - Immune marker: ANA, ANCA, 50 pattern and CH50: N
    - 24-hour urine: 0.75g 24h
    - Thyroid function: TSH mild elevated, others normal
    - Skin biopsy: consistent with Loevskydetaidic vasculitis

- 3rd pregnancy at 3/2010
- Her skin rashes appeared again
- Resumed Prednisolone 25mg/day since 06/05/2010
- Consulted Rheumatologist in HBL, add HCQ 200mg/day and adjust the dose of Prednisolone
- Now, the patient on pregnancy 36 weeks
- Keep taking low dose steroid. HCQ treatment and her skin lesion control acceptable without renal involvement.

Henoch Schölein purpura

Henoch-Schönlein purpura (HSP) is the most common form of systemic vasculitis in children. 90% of cases occur in the pediatric age group. In contrast to many other forms of systemic vasculitis, HSP is self-limited in the great majority of cases.

Clinical manifestations

- Palpable purpura in all patients without thrombocytopenia and coagulopathy. It is symmetrical distribution and located in periorificial-dependent areas
- Arthralgia/arthritis (3-4 patients) – usually transient or migratory, typically oligoarticular and non-deforming.
- Abdominal pain (about 2 patients) – Continuous or episodic bloating and intussusception is the most common complications.
- Renal disease (21 to 54 %) – isolated hematuria and/or proteinuria without renal function abnormality, or with renal insufficiency.

These clinical manifestations may develop over the course of days to weeks and may vary in presentation.

ACR criteria for the diagnosis

- Palpable purpura
- Age at onset <20 years
- Acute abdominal pain
- Involving skin, upper respiratory, or gastrointestinal tracts

Epidemiology and Pathogenesis

- Primarily between the ages of 3 and 15 years. The annual incidence is 10-20 per 100,000 in children. 17 years of age with a peak incidence in children between 4 to 6 years of age. Approximately 10% cases occur in adults
- Male-to-female ratio of 1.2 to 1.8
- Less frequently seen in black compared to white or Asian
- Primarily in the fall, winter, and spring but ready in the summer
- An immune-mediated vasculitis associated with immunoglobulin A (IgA) deposition.

The underlying cause remains unknown. It is thought that result of an immunemediated vasculitis disorder, that is often triggered by infections or immunizations.

Management

- Most patients with Henoch-Schönlein purpura can be managed conservatively as outpatients during the acute illness.
- Symptomatic relief of joint and abdominal manifestations.
- Targeted treatment to decreases the risk of gastrointestinal and renal complications.
- Although many different therapeutic agents, no therapy has been shown to be beneficial in a controlled trial.
- Patients with renal disease may be hypertensive and require antihypertensive therapy.

Hospitalization

- Inability to maintain adequate hydration with oral intake
- Severe abdominal pain
- Significant gastrointestinal bleeding
- Changes in mental status
- Severe joint involvement limiting ambulation or self-care
- Renal insufficiency, hypertension, or nephrotic syndrome

The outcome of children is generally excellent. In the absence of significant renal disease, typically resolves within one month. In two-thirds of children, there are no recurrent episodes.

The long-term morbidity in patients is a result of renal disease. The risk of renal disease is increased in adults.

Prognosis and Follow Up

- The disease in children is usually self-limited. Follow all patients with renal involvement at the office for at least 3 months after the last episode.
- No difference in clinical and laboratory findings between patients with or without renal involvement.
- Usually follow all patients with arthritis and blood pressure monitoring for the first one to two months after presentation.

Figure 3. A: Palpable purpura on the lower legs. B: Skin test reaction with Mischke-Schuchardt and Mantoux test in the subcutaneous tissue.

Figure 4. A: Palpable purpura on the lower legs. B: Skin test reaction with Mischke-Schuchardt and Mantoux test in the subcutaneous tissue.

Figure 5. A: Henoch Schnollein purpura with lesions on the lower legs. B: Bone biopsy results with H&E and PAS staining showing granulocytes in the walls of small arteries and/or venules.
WILSON'S DISEASE (WD)

Introduction:
Wilson's disease (WD) is an autosomal recessive disorder of hepatic copper metabolism, first described as progressive hepatocellular degeneration by Wilson in 1912. The condition is characterized by excessive deposition of copper in the liver, brain, and other tissues. The worldwide prevalence has been reported to be approximately 1 in 30,000, most patients present between the ages of five and 40, peaking at ages 10-15 years. Diagnosis is based on at least two of the following: detection of KP rings on slit-lamp examination, typical neurological symptoms, and/or low serum ceruloplasmin (Cp) concentration (<0.2 g/L).

Case report:
A 18-year-old student boy started to have the symptoms of both hard tumor about one year ago. He didn't seek medical care till he has slurred speech and unstable gait for 6 months. The above symptoms worsened progressively. On May 2007, he consulted local hospital for assessment. With brain CT showed abnormal signal in basal ganglia, he was transferred to our hospital for further investigation with suspect Wilson's disease. In physical examination, he has jaundice, tremulousness, and mild hepatosplenomegaly. His family history was unremarkable. Further examination showed that the Kayser-Fleischer rings (KF) was found by slit-lamp examination by ophthalmologist. Serum ceruloplasmin level was 0.03 g/L (normal range: 0.22-0.56 g/L) and the 24-hour urine copper was 1.39 mg (normal range: 0.5 mg). The abdominal ultrasound confirmed the absence of kidney stones, no masses or occupying lesions. Brain MRI showed abnormal signal in basal ganglia (Figure 2). Subsequently, liver biopsy compatible with chronic liver disease with moderate to severe fibrosis. Dopa stain for tyrosine hydroxylase was performed in liver biopsy tissue. Histological color showed Kupffer cell hyperplasia in peri-portal hepatocytes (Figure 3).

Discussion:
Copper is an important component of several enzymes and is an essential cofactor for heme synthesis. Copper concentration is highest in the liver, accounting for approximately 80% of total body copper. In normal individuals, copper is absorbed from the gut into the circulation, undergoes complexation with serum proteins, and is transported to the liver. Copper is secreted into the bile and excreted into the GI tract. Only a small portion (1.5%) of dietary copper is absorbed, and this is sufficient to meet the body's needs. Copper balance is maintained by the regulation of copper absorption, excretion, and tissue utilization. Copper absorption is influenced by dietary factors, such as the intake of copper, zinc, and iron. Dietary copper excess or deficiency can lead to copper-related diseases.

Wilson's disease is one of the inherited copper storage disorders, characterized by the accumulation of copper in the liver, brain, and other tissues. The excess copper results in the formation of copper chelates, which can cause injury to these organs. The clinical manifestations of Wilson's disease vary widely and can include neurological symptoms, such as tremors and dystonia, as well as liver disease, such as cirrhosis and hepatic encephalopathy.

Mrs. Wilson disease requires lifelong therapy. Initial treatment for symptomatic patients usually includes a chelating agent, either penicillamine or trientine. Penicillamine is still the 'gold standard' for therapy. Oral zinc can prevent copper absorption. Patients should avoid foods containing high concentrations of copper, including shellfish, nuts, chocolate, mushrooms, and organ meats. Patients should also avoid alcohol, which can exacerbate liver disease.

References:

Figure 1: Kayser-Fleischer rings are found in the cornea of the eyes as a result of excess copper deposition.

Figure 2: Liver biopsy showing chronic liver disease.

Figure 3: Dopa stain for tyrosine hydroxylase in liver biopsy tissue.
Objective  Retrospectively analyze the clinical utility of Continuous Renal Replacement Therapy (CRRT) in the Critical Care Unit of the Internal Medicine Department.

Method  During the period of Jun 2004 to Dec 2007, CRRT were performed on 42 cases of critically ill patients in our department. General data including age, gender, primary underlying disease, comorbidity, and laboratory examinations pre- and post-treatment were collected and analyzed.

Result  25 out of 42 patients had been survival from the acute phase of the illness. The survival rate was 59.62%. 17 patients died during the acute phase. The death rate was 40.48%. The mean age of the dead group is 74.2 ±18.5 yrs, which was obviously older than that of the survival group (64.7±18.7 yrs), but there was no statistical significance (P=0.497).

Conclusion  CRRT might provide an effective organ support to the critically ill patients. CRRT might be of help to increase the survival rate of the critically ill patients in the internal medical department.
Objective Retrospectively study the thrombolytic effect of urokinase in the treatment of thrombosed permanent catheter of dialysis patients.

Methods Diluted Urokinase (5000iu/ml) was instilled in thrombosed catheter for 30-60min, observed the change of arterial lumen blood flow and venous pressure in venous lumen during dialysis before and after the thrombolytic treatment. Side effects of the treatment were also recorded.

Results of 20 cases with 61 times and 70 lumens of thrombolytic treatments, 78.33% arterial lumen restored sufficient blood flow and 90% venous lumen pressure dropped to normal range to ensure an effective and fluent dialysis. The overall successful rate of thrombolytic treatment was 80%. No complication was recorded.

Conclusion Intracatheter thrombolysis treatment with Urokinase is highly effective and safe in restoring the patency of occluded permanent catheter among hemodialysis patients.
Objective To investigate the clinical value of using histamine bronchoprovocation test in the diagnosis of cough-variant asthma (CVA).

Methods In a group of 122 patients with chronic cough, bronchoprovocation test were performed. Comparisons were made between air hyperresponsiveness and history, clinical features in the diagnosis of asthma regarding sensitivity, specificity, Youden’s index.

Results AHR has a higher detection rate for cough-variant asthma compare to the clinical features. For diagnosing asthma, AHR has a sensitivity of 90.8%, Specificity of 91.2%, Youden’s index of 82.0%.

Conclusion Histamine bronchoprovocation test is a useful methods for diagnosing CVA. It has advantages over history, and clinical features.
Apical ballooning syndrome (also named Tako-tsubo syndrome, or stress induced cardiomyopathy) is an increasingly recognized clinical syndrome. The clinical characteristics of this syndrome have been described as follows: acute onset of chest pain with reversible LV apical wall motion abnormalities (ballooning), ECG changes (ST-elevation), minimal myocardial enzyme release, and no significant stenosis of the coronary artery on angiography. The clinical manifestations can mimic an acute coronary syndrome. The common etiologic feature of this syndrome is usually after a sudden emotional or physical stress.

We report the two cases of a 70-year-old female and 37-year-old male, who arrived to the emergency department with chest pain for 1 to 3 hours, ECG showed ST-segment elevation in V1—V5 and cardiac markers (CK, CK-MB, TNT) were elevated, echocardiographic features of hypokinesis at left ventricular apical with lower ejection fraction (E40%), Coronary angiography showed coronary arteries without remarkable stenosis lesions; left ventriculography showed apical ballooning changes. The follow-up performed with echocardiography (1 month later) showed complete recovery wall motion with EF 60%.
The significance of using fibroscan in the diagnosis of liver stiffness in chronic hepatitis B

余漢濠 徐義祥 梁棋 徐威傑 鄭展宏 黃宇鑫
朱雪茵 王文棟 詹德娟
鏡湖醫院內科

• **Objective** To investigate the clinical usage of fibroscan among Chronic Hepatitis B patients.

• **Methods** Fibroscan was performed in 364 Chronic Hepatitis B patients for measuring liver stiffness. HBeAg, HBV DNA, ALT, AST, AFP, GGT, ALB and PLT levels were also detected at the same time. The comparison between liver stiffness and biochemical indexes would be measured.

**Results** There was statistical significance (p<0.05) between liver stiffness among male patients, age group ≤35 and >55, ALT levels within 1-2 ULN and 2-5xULN compared with ≤0.5xULN, HBV DNA levels and all biochemical indexes. The liver fibrosis level in HBeAg among F2 and F4 level had statistical significance (p<0.05).

• **Conclusions** Fibroscan can be used as a non-invasive diagnosis method for monitoring the progression of liver stiffness in chronic hepatitis B.
OBJECTIVES: This study was to compare orthodromic and antidromic technique in sensory conduction study.

METHODS: In 19 healthy volunteers aged between 12 and 70 years, the orthodromic and the antidromic sensory conduction for 66 normal upper limb nerves have been studied. Parameters studied include latency, amplitude and sensory conduction.

RESULTS: The amplitude of the SNAP was higher in antidromic stimulation than in orthodromic. The amplitude of the SNAP of the median nerve (wrist), median nerve (elbow) and ulnar nerve using antidromic stimulation were 80.42±33.17, 37.7±19.68, 80.61±33.98, respectively. In controls, using orthodromic stimulation were 28.33±11.31, 4.41±2.43, 20.75±10.11, (p<0.001). There was no significant difference between orthodromic and antidromic stimulation for latency and sensory conduction velocity, (p>0.05).

CONCLUSIONS: We recommend the use of the antidromic technique because of its higher amplitudes, ease of use and high efficiency.

<table>
<thead>
<tr>
<th>表 1 順向法與逆向法 SNAP 潛伏期比較</th>
</tr>
</thead>
<tbody>
<tr>
<td>正中神經（腕部）</td>
</tr>
<tr>
<td>$ar{X}$</td>
</tr>
<tr>
<td>順向法</td>
</tr>
<tr>
<td>逆向法</td>
</tr>
<tr>
<td>*P&lt;0.05</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>表 2 順向法與逆向法 SNAP 波幅比較</th>
</tr>
</thead>
<tbody>
<tr>
<td>正中神經（腕部）</td>
</tr>
<tr>
<td>$ar{X}$</td>
</tr>
<tr>
<td>順向法</td>
</tr>
<tr>
<td>逆向法</td>
</tr>
<tr>
<td>*P&lt;0.001</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>表 3 順向法與逆向法感覺傳導速度比較</th>
</tr>
</thead>
<tbody>
<tr>
<td>正中神經（腕部）</td>
</tr>
<tr>
<td>$ar{X}$</td>
</tr>
<tr>
<td>順向法</td>
</tr>
<tr>
<td>逆向法</td>
</tr>
<tr>
<td>*P&lt;0.05</td>
</tr>
</tbody>
</table>