台灣腎臟醫學會
中部地方會

【第 217 次腎臟學學術討論會】

節目表及摘要

- 第 217 次腎臟學學術討論會
  時　間：民國 100 年 6 月 12 日上午 8:30 – 12:00
  地　點：中國醫藥大學立夫教學大樓國際會議廳

- 第 69 次透析人員繼續教育課程
  【中部場次】
  時　間：民國 100 年 6 月 12 日上午 9:00—12:00
  地　點：中國醫藥大學立夫教學大樓 102~105 教室
  【南部場次】
  時　間：民國 100 年 6 月 5 日上午 9:00—12:00
  地　點：高雄榮民總醫院 第一、二會議室

台 灣 腎 臟 醫 學 會
中 部 地 方 會

時間: 民國 100 年 6 月 12 日(星期日)

地點: 中國醫藥大學立夫教學大樓國際會議廳

♣ 第 217 次腎臟學學術討論會 ♣

【病例討論 I】　主持人：張志宗

08:30 － 08:48　1. Nephronophthisis in Ellis-van Creveld syndrome: a pediatric case report

顏廷芳[1] 文美卿[2] 陳呈旭[3] 賀昊中[4] 傅令嫻[1]

台中榮民總醫院兒童醫學部小兒腎臟科[1] 台中榮民總醫院病理部[2]

台中榮民總醫院內科部腎臟科[3] 台中榮民總醫院外科部泌尿外科[4]

08:48 － 09:06　2. Small bowel perforation: a late and fatal complication of encapsulating peritoneal sclerosis

郭韋宏　陳德全　吳建興　楊智超　許國泰　李建德

高雄長庚紀念醫院　內科部　腎臟科

09:06 － 09:24　3. Undifferentiated autoimmune rheumatic and connective tissue disorder (Overlap syndrome) with heavy proteiuria: a case report and literature review

陳世杰　林清淵

中國醫藥大學附設醫院兒童腎臟科

09:24 － 09:42　4. Catastrophic Antiphospholipid Syndrome after Infection: A Case Report

陳炯霖　林軒名　張志宗　黃秋錦

中國醫藥大學附設醫院 內科部腎臟科暨腎臟醫學中心

09:42 － 10:00　5. Salmonella Mycotic Aneurysm in a Man with Lupus Nephritis

李振豪[1,2]　張家築[2]

大林慈濟醫院腎臟內科[1]　彰化基督教醫院腎臟內科[2]

10:00 － 10:20　Break

【病例討論 II】　主持人：張家築

10:20 － 10:38　6. Chylous ascites and chylothorax: complications of nephrotic syndrome associated with membranous nephropathy

柯志霖　詹尚儒　邱炳芳　張家築

彰化基督教醫院內科部腎臟科

10:38 － 10:56　7. Successful treatment and complete remission in a 60-year-old female with idiopathic crescentic gromerulonephritis

林芳芳[1,2]　張淀榮[1,3]
中山醫學大學附設醫院[1]腎臟科　[2]小兒腎臟科　[3]中山醫學大學醫研所

10:56 － 11:14　8. Paroxysmal Nocturnal Hemoglobinuria and Acute Renal Failure: a Case Report and Literature Review

王彩融[1]　文美卿[2]　鄭志雄[1]　吳明儒[1]　陳呈旭[1]　游棟閔[1]　莊雅雯[1]
黃士婷[1]　徐國雄[1*]
台中榮民總醫院內科部 腎臟科[1]　台中榮民總醫院病理部[2]

11:14 － 11:32　9. A Case of Life-threatening Hypokalemia

張維誠　黃文德
國軍岡山醫院. 國軍左營醫院

Ellis-van Creveld 症候群之 nephronophthisis:壹兒童病例報告
Nephronophthisis in Ellis-van Creveld syndrome: a pediatric case report

顏廷芳[1], 文美卿[2], 陳呈旭[3], 賀昊中[4], 傅令嫻[1]
Ting-Fang Yen[1], Mei-Chin Wen[2], Cheng-Hsu Chen[3], Hao-Chung Ho[4], Lin-Shien Fu[1]
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[1]Division of Nephrology, Department of Pediatrics, [2] Department of Pathology,[3] Division of nephrology Department of internal medicine, [4]Division of Urology, Department of Surgery, Taichung Veterans General Hospital, Taichung, Taiwan

Ellis-van Creveld (EvC) syndrome, a chondral and ectodermal dysplasia characterized by short ribs, polydactaly, growth retardation, ectodermal and heart defects, is a rare autosomal recessive disorder. In previous literatures, the association between renal insufficiency and EvC syndrome has not been well described. We herein report a nine-year-old girl with typical EvC syndrome diagnosed since infancy presented with renal insufficiency during her childhood. The initial presentations were hypertension, heavy proteinuria and impaired renal function. Histopathological findings from biopsy specimen were indicative of nephronophthisis. Progressive renal failure and uremia occurred in months and she received peritoneal dialysis. Later, she received cadaveric renal transplantation at 11-year-old. The post-transplantation course was uneventful with fair renal function and no episode of rejection. The serum creatinine decreased to 1.0 mg/dl after renal transplantation for two years. Inspite of its rarity, we concluded nephronophthisis with subsequent renal insufficiency may be present in patients with EvC syndrome.

Key words: Ellis-van Creveld syndrome, Chronic renal failure,
Renal transplantation
關鍵字:Ellis-van Creveld 症狀，慢性腎臟衰竭，腎臟移植

Small bowel perforation: a late and fatal complication of encapsulating peritoneal sclerosis

包囊性腹膜硬化症合併小腸破裂之病例報告

郭韋宏　陳德全　吳建興　楊智超　許國泰　李建德

Wei-Hung Kuo, Te-Chuan Chen, Chien-Hsing Wu, Chih-Chau Yang, Kuo-Tai Hsu, Chien-Te Lee

高雄長庚紀念醫院　內科部　腎臟科

Division of Nephrology, Department of Internal Medicine, Chang Gung Memorial Hospital-Kaohsiung Medical Center, Kaohsiung, Taiwan

Encapsulating peritoneal sclerosis (EPS) is a rare and serious complication in patients undergoing peritoneal dialysis (PD). The incidence of EPS increases with duration of PD. The rate of EPS-associated mortality rate is high, ranging between 24 and 56%. We encountered a 51-year-old female with history of uremia and had received PD therapy since 1999. There were three episodes of PD-related peritonitis and she was thus shifted to hemodialysis (HD) in 2008 due to dialysis inadequacy and ultrafiltration failure. Her HD schedule was trice weekly with four hours per session. Latest Kt/V was 1.28 and URR was 72.5%. Prior to this hospitalization, EPS was diagnosed and exploratory laparotomy with enterolysis was performed in July, 2008. In October, 2009, she was sent to emergency room with acute onset abdominal pain. Upon arrival, profound hypotension (77/42 mmHg), abdominal distention with rebounding pain were noted. Laboratory tests revealed: leukocytosis (WBC:10300/mm^3), hemoglobin 11.6 g/dL, CRP 12.5 mg/L, lactate 32 mg/dL, BUN 82 mg/dL, creatinine 9.0 mg/dL, Na 133 meq/L, and K 6.5 meq/L. Abdominal computed tomography demonstrated perforation of bowel loops with pneumoperitoneum and ascites. Despite emergent laparotomy and antibacterial therapy, the patient died 2 days later. The culture of paracentesis yielded *E. coli, Enterococcus faecalis, Enterobacter cloacae, and Clostridium perfringens*. We concluded that EPS can still progressed in uremic patients even PD had been withdrawn. Our patients died of bowel perforation with associated overwhelming sepsis though dialysis had been switched from PD to HD.

Key words: Encapsulating peritoneal sclerosis, bowel perforation, peritoneal dialysis

混合性結締組織病合併嚴重蛋白尿的病例報告及文獻回顧

Undifferentiated autoimmune rheumatic and connective tissue disorder (Overlap syndrome) with heavy proteiuria: a case report and literature review

陳世杰, 林清淵
Chen Shih-Chieh, Lin Ching-Yuang
中國醫藥大學附設醫院兒童腎臟科 Pediatric nephrology, China University hospital

A 20 year-old boy had pain at both ankles of leg for four days. According to him, he had swelling of both ankles of leg since four days ago. The swelling of ankle was no reddish and affected his movement. He also had back pain especially at morning and holding one posture for a while. About 1 year ago, he had the similar symptoms, and he went to hospital for help. The dignosis was ankylosing spondylitis at that time. By the way, he had vomit, abdomen distention for 7 days. He also had abdominal pain at the area from left flank to epigastric quadram. The vital sign was T/P:R: 36.4C/84/20 BP:155/93mmHg. Physical examinations showed bilateral lower legs edema. There was no Raynaud's phenomenon, no swollen hand. He had mild myalgia and arthralgias over bilateral lower extremities. Urine was yellow, but was foamy. After admission, serial examinations were done. The immunologic studies only showed increased Jo-1 titre without other ENA elevation nor hypocomplementemia. Serum biochemistry showed hypoalbuminemia (1.6g/dl) with hyperlipidemia (TC/TG:477/283 mg/dl). Low-dose methylprednisolone pulse therapy and immunosuppresants with Tacrolimus and Cellcept as well as albumin infusion were administered. Renal biopsy was also done on 2010-12-20 and showed thrombotic microangiopathy (TMA) and mild dilated glomerular microangiopathy. The H&E stain showed mesangial proliferation while the IF stain showed C3 and fibrinogen staining over mesangial and perivascular areas. The 24hr urine protein was 27.5g and the CCr was 134ml/min/1.73m2 (GFR= 112ml/min/1.73m2). Pitting edema improved gradually. During follow-up, he had frequent relapsing.

The pathology of overlap syndrome is pleomorphic with the glomerular lesions resembling the spectrum found in SLE and vascular lesions resembling those found in scleroderma. Our patient showed thrombotic microangiopathy. The anti-Jo-1 antibodies are present in 20% to 25% of adult myositis patients and are highly specific for myositis associated with a constellation of symptoms including skin involvement, lung disease, Raynaud's phenomenon, inflammatory arthritis, and fever that may manifested with overlap syndrome. Overlap syndrome is not a benign disease and may evolve to other connective tissue disease. The mortality rates have been found to range from 15% to 30% at 10 to 12 years with patients with more clinical features of scleroderma and polymyositis having a worse prognosis.

Key words: Overlap syndrome,, thrombotic microangiopathy, nephrotic syndrome
關鍵字: 混合性結締組織病, 血栓性小血管病變, 腎病症候群

感染後引發的災難性抗磷脂症候群：病例報告

Catastrophic Antiphospholipid Syndrome after Infection: A Case Report

陳炯霖, 林軒名,張志宗 ,黃秋錦
Chiung-Lin Chen, Hsuan-Ming Lin, Chiz-Tzung Chang, Chiu-Ching Huang
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Background: Antiphospholipid syndrome (APS) is a systemic autoimmune disorder characterized by a combination of arterial and/or venous thrombosis, pregnancy morbidity, thrombocytopenia and the appearance of serum antiphospholipid antibody. Catastrophic APS (CAPS) is a variant of APS, which is usually life-threatening with rapid progressive multiple organ failure. Infection is the most common trigger factor.

Case Report: A 33 year-old woman had acute hepatic dysfunction, pulmonary edema and general edema after an episode of urinary tract infection. APS was favor according to her clinical presentation and laboratory examination. Pulse steroid was administered. Severe thrombocytopenia, hemolytic anemia, acute renal failure, and cerebral vascular thrombosis developed despite high dose steroid therapy. We prescribed daily plasma exchange and anticoagulation therapy for her catastrophic APS. Thrombocytopenia, anemia, renal dysfunction, and liver dysfunction recovered gradually after titers of antiphospholipid antibnody improved.

Conclusion: For unusual thrombocytopenia and multiple organ dysfunction after an infection episode, CAPS should be considered and plasma exchange can be applied if there is no contraindication.

Key words: Catastrophic antiphospholipid syndrome, infection, multiple organ failure, thrombocytopenia, plasma exchange..

沙門氏菌感染性動脈瘤發生在紅斑性狼瘡腎炎病人

Salmonella Mycotic Aneurysm in a Man with Lupus Nephritis

李振豪 Chen-Hao Li[1,2], 張家築 Chia-Chu Chang[2]
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彰化基督教醫院腎臟內科

A 69-year-old man presented with intermittent fever for three days on 2010/12/24. He had history of systemic lupus erythematosus (SLE) and lupus nephritis class V, diagnosed in 2007. The current medication included prednisolone 10mg per day, azathioprine 50mg per day, allopurinol 100mg per day.

He had dry cough, and mild pain at left upper chest wall. There were no oral ulcer, malar rash, skin lesion, lymphadenopathy, abdominal tenderness, or symptoms of urinary or gastroenteral tracts. The complete cell count showed white cell count 7900/μL, Segment form 77.2%, lymphocyte 9.7%, hemoglobin 13.3 g/dl, and platelet 200,000/μL. The Erythrocyte sediment rate was 80 mm per hour and C-reactive protein was 16 mg/dL. The basic biochemistry data disclosed sodium 132 mmol/L, potassium 3.2 mmol/L, blood urea nitrogen 11 mg/dL, and creatinine 0.90 mg/dL. The urinalysis showed proteinuria 1+, RBC 8.4 per high power field, WBC: 4.4 per high power field. The blood culture grew Gram negative bacilli (GNB) three days later. The chest plain film on 12/27/2010 found a bulging lesion at aortic arch. Gallium scan disclosed intensely increased gallium uptake involving left upper anterior chest wall. The identification of GNB bacteremia proved Salmonella enteritidis C1 infection. The antibiotic had been shifted to ceftriaxone 2000mg ivd Q12H on 12/27. Because of Salmonella bacteremia, bulging mass at aortic arch on chest plain film, and intensely gallium uptake involving left upper anterior chest wall, mycotic aneurysm of aortic arch is highly suspected. Chest computed tomography (CT) with contrast enhancement on 12/29/2010 showed atherosclerosis with suspicious aortitis with pseduoaneurym formation. He was discharged after effective parenteral antibiotic therapy for 4weeks.

Reviewing literature, this case report reminded us of susceptibility of Salmonella bacteremia in patient with SLE. It's reasonable to screen for mycotic aneurysm in patient elder than 50 years old or with atherosclerotic risks.

關鍵字: 沙門氏菌, 紅斑性狼瘡, 感染性動脈瘤
Key words: Salmonella, Lupus, Mycotic aneurysm

乳糜性腹水和乳糜胸：膜性腎病變相關之腎病症候群的併發症

Chylous ascites and chylothorax: complications of nephrotic syndrome associated with membranous nephropathy

柯志霖，詹尚儒，邱炳芳，張家築
Chih-Lin Ke, Shang-Ju Chan, Ping-Fang Chiu, Chia-Chu Chang
彰化基督教醫院內科部腎臟科
Division of Nephrology, Department of Internal Medicine, Changhua Christian Hospital

Chylous ascites has been reported as complication of nephrotic syndrome. However, coexistence of chylous ascites and chylothorax is a rarely reported condition in adult nephrotic syndrome associated with membranous nephropathy.

A 58-year-old woman presented with lower leg edema, heavy proteinuria, hypoalbuminemia and hyperlipidemia in December, 2008. She was diagnosed with nephrotic syndrome and the pathology of kidney biopsy revealed membranous nephropathy. However, dyspnea and abdominal discomfort developed in October, 2009. Chest radiography revealed right pleural effusion and abdominal ultrasonography demonstrated massive ascites. Thoracocentesis and paracentesis were performed and both disclosed white milky transudative fluids with triglyceride levels of 77 and 72 mg/dl, respectively, which were negative for culture and cytology. Lymphoscintigraphy showed no evidence of lymphatic leakage in the lung field. Technetium-99m phytate was injected into peritoneal cavity and revealed increased radioactivity in the right thoracic cavity which consistent with peritoneal-pleural shunting. Serial examination, including chest computed tomography, abdominal magnetic resonance imaging and positron emission tomography, did not show any cause of chylous ascites and chylothorax. Video-assisted thoracoscopic surgery with mechanical pleurodesis was performed, but no obvious diaphragmatic defect was found. After the procedure, clinical condition was improved by albumin infusion coupled with diuretics for nephrotic syndrome and she was discharged uneventfully.

The pathogenesis of chylous ascites in nephrotic syndrome is unknown. Hypoalbuminemia-induced bowel edema may predispose to change the permeability of mucosal or serosal lymphatics. Chylous ascites and chylothorax may be present concurrently in nephrotic syndrome. The mechanism is uncertain and it may be associated with or without a transdiaphragmatic shunt.

關鍵字: 乳糜性腹水，乳糜胸，腎病症候群，膜性腎病變
Keywords: chylous ascites, chylothorax, nephrotic syndrome, membranous nephropathy

一位六十歲原發性半月型快速進行性腎絲球腎炎患者之成功治療經驗

Successful treatment and complete remission in a 60-year-old female with idiopathic crescentic gromerulonephritis

林芳芳 [1,2]，張浤榮 [1,3]
Lim Fong Fong[1,2], Horng-Rong Chang[1,3]
中山醫學大學附設醫院 [1] 腎臟科 [2] 小兒腎臟科 [3] 中山醫學大學醫研所
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[3] Institute of Medicine, Chung Shan Medical University, Taichung

A 60-year-old female with normal renal function previously had foamy urine for one month and gross hematuria for ten days before admission. Elevated serum creatinine level to 2.4 mg/dl and normocytic anemia (Hb 8.3 gm/dl) were noted. Her urinalysis showed proteinuria (protein ++) and hematuria (occult blood +++, urine RBC >100 /HPF). 24-hour urine protein was 1.44 gm/day and creatinine clearance (CCr) was 26.6 ml/min. Renal sonogram revealed mild increased echoic pattern in both kidneys with right kidney 11.53 cm in size and left kidney 11.57 cm in size. The reports of renal biopsy disclosed pauci-immune focal necrotizing and crescentic glomerulonephritis, with mild tubular atrophy/interstitial fibrosis.

After treatment according to the recommended protocol, oral prednisolone and cyclophosphamide for four months, her serum creatinine recovered to 1.2 mg/dl. The urinalysis showed proteinuria (protein +) and hematuria (occult blood +, urine RBC 15-20 /HPF). 24-hour urine protein was 0.42 gm/day and CCr was 77.1 ml/min. The reports of second renal biopsy revealed focal sclerotic glomerulonephritis, consistent with inactive pauci-immune focal necrotizing glomerulonephritis. The patient has been maintained with azathioprine and the renal function remained normal with nephrological follow-up for two years.

關鍵字：半月型快速進行性腎絲球體腎炎、蛋白尿、血尿。
Key words: crescentic gromerulonephritis, proteinuria, hematuria, cyclophosphamide, azathioprine.

陣發性夜間血尿症與急性腎衰竭: 一個病例報告與文獻回顧

Paroxysmal Nocturnal Hemoglobinuria and Acute Renal Failure: a Case Report and Literature Review

王彩融[1]、文美卿[2]、鄭志雄[1]、吳明儒[1]、陳呈旭[1]、游棟閔[1]、莊雅雯[1]、黃士婷[1]、徐國雄[1*]

Tsai-Jung Wang [1], Mei-Chin Wen[2], Chi-Hung Cheng[1], Ming-Ju Wu[1], Cheng-Hsu Chen[1], Tung-Min Yu[1], Ya-Wen Chuang[1], Shih-Ting Huang[1], Kuo-Hsiung Shu[1*]

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Division of Nephrology[1], Department of Internal Medicine, Taichung Veterans General Hospital, Taichung, Taiwan

Department of Pathology[2], Taichung Veterans General Hospital, Taichung, Taiwan

Paroxysmal nocturnal hemoglobinuria(PNH) is an uncommon, acquired clonal disorder of hematopoietic stem cells. It is presented by intravascular hemolysis, recurrent infection, thrombosis, and bone marrow failure. A somatic mutation in the phosphatidylinositol glycan class A (PIG-A) gene renders the red blood cells particularly sensitive to lysis via the alternative pathway of complement activation. Kidney involvement secondary to chronic tubular deposition is usually benign. Acute renal failure occurred during a hemolytic crisis is extremely rare.

We report a case of a 71-year-old woman who was admitted to this hospital because of epigastric pain and flu-like symptoms for five days. She had taken Chinese herb after the onset of symptoms. Decreased urine output was noted one day before admission. The initial Cr was 2.9 mg/dl with rapid progressed to 10.7 mg/dl within one week. Laboratory studies showed normocytic anemia with a hemoglobin of 9.6 g/dl, reticulocyte count of 0.68%, elevated LDH, mild unconjugated hyperbilirubinemia, and low haptoglobin. Urinalysis revealed hematuria, glucosuria and proteinuria. Light microscopy of the kidney biopsy specimen demonstrated acute tubular injury and the iron stain showed increased hemosiderin pigments in tubular cells. Further evaluation for the cause of hemolysis revealed no fragment RBCs on the peripheral blood smear but the presence of a PNH clone (absent CD55 and CD 59 by flow cytometry assay on RBCs). The patient was adequately hydrated. Finally, the renal functions test was recovered three weeks later without renal replacement therapy.

Key words: Acute renal failure, paroxysmal nocturnal hemoglobinuria, hemolysis

關鍵字：急性腎衰竭，陣發性夜間血尿症，溶血

致命低血鉀病例報告

A Case of Life-threatening Hypokalemia

張維誠　黃文德
Cheng-Wei Chang, MD[1], Wen-Te Huang, MD[2]
國軍岡山醫院. 國軍左營醫院
[1]Division of Nephrology, Department of Medicine,
Gangshan Armed Forces Hospital

A 16-year-old previous healthy female presented to the emergency department because of consciousness disturbance. Blood biochemistry tests showed potassium: 1.8 mmol/L and sodium:129 mmol/L. Blood gas analysis : PH: 7.595. PaCO2: 30.2 mmHg, PaO2:98 mmHg, Hco3-: 28.6mmol/L. Urine biochemistry revealed: sodium:110 mmol/L, potassium:34 mmol/L, chloride: 105 mmol/L, creatine: 154 mg/dl, osmolality: 385 mos/kg H2O. EKG revealed low voltage and multiple U waves. After emergent KCL (10 meq) administration, she suddenly developed cardiac arrest and emergent ACLS and ET-tube intubation was performed then. After 4 days ventilator support, ET-tube was successfully removed on the 4 hospitalized day

The diagnosis of anorexia nervosa precipitated severe hypokalemia is easy missing in ER, especially in young female. Due to anorexia nervosa rarely cause of life-threatening hypokalemia in the literatures. However, our case highlights the importance that early recognition of cause of hypokalemia and appropriate management may improve the clinical outcomes. Physicians should keep in mind that anorexia nervosa *per sec* is as a potential cause of severe hypokalemia.

Key words: **Hypokalemia,**

台灣腎臟醫學會

ᘓᘓ 第 69 次透析人員繼續教育課程 ᘓᘓ

【中部場次】

時　間：民國 100 年 6 月 12 日上午 9:00—12:00
地　點：中國醫藥大學 立夫教學大樓 101~104 教室
主　題：腫瘤腎臟學(Onco Nephrology)

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主持人： 張浤榮

09:00－09:40　1、腎切除後之慢性腎臟病(Chronic kidney disease after Nephrectomy)
張浤榮　主任
中山醫學大學附設醫院　腎臟科

09:40－10:20　2、腫瘤溶解症候群 (Tumor Lysis syndrome)
丁羿文　醫師
中國醫藥大學附設醫院　腎臟科

10:20－10:40　休　息

主持人：吳明儒

10:40－11:20　3、化學治療的腎毒性 (Chemotherapy related Nephrotoxicity)
李文欽　主任
彰化秀傳醫院　內科部

11:20－12:00　4、慢性腎臟病與腫瘤 (Cancer and CKD)
吳明儒主任
台中榮民總醫院　一般醫學內科

台灣腎臟醫學會

≈ 第 69 次透析人員繼續教育課程 ≈

【南部場次】

時　間：民國 100 年 6 月 5 日上午 09:00—12:00
地　點：高雄榮民醫院　第一、二會議室
主　題：腫瘤腎臟學(Onco Nephrology)

♬♬♬

主持人：黃偉傑

09:00－09:40　1、慢性腎臟病與腫瘤 (Cancer and CKD)
黃偉傑　醫師
高雄榮民總醫院　腎臟科

09:40－10:20　2、腫瘤溶解症候群 (Tumor Lysis syndrome，包括 Paraneoplastic syndrome)
張子爰　醫師
高雄榮民總醫院　腎臟科

10:20－10:40　休　息

主持人：郭健群

10:40－11:20　3、化學治療的腎毒性 (Chemotherapy and Radiation related Nephrotoxicity)
郭健群　醫師
高雄長庚紀念醫院　腎臟科

11:20－12:00　4、腎切除後之慢性腎臟病(Chronic kidney disease after Nephrectomy)
郭弘典　醫師
高雄醫學大學附設醫院　腎臟科