

$p=0.0032$ ), 5 out of 7 patients with renal involvement in the rituximab group were eventually hemodialysis dependent.

**Conclusions** In summary, our data indicated that add-on rituximab in the background of conventional therapy may improve the overall but not the renal survival in SLE-TTP patients.

### 199 AGE ADJUSTED CHARLSON'S COMORBIDITY INDEX SCORE PREDICT MAJOR ADVERSE CARDIOVASCULAR EVENTS IN SYSTEMIC LUPUS ERYTHEMATOUS

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**Background and aims** Cardiovascular events are the leading cause of death or disability in people with systemic lupus erythematosus (SLE). However, the real mechanism that causes the risk increments for the major adverse cardiovascular events (MACEs) is still not fully understood. The Age-Adjusted Charlson Comorbidity index (ACCI) score is a prognostic classification that was initially developed for a patient who may have a range of co-morbid conditions and has been validated in many clinical settings.

**Methods** The data for this study were collected from Taiwan's National Health Institute Research Database (NHIRD) for the years 2004 to 2007, 5998 participants were enrolled. All patients' sequential clinically diagnosed SLE data were reviewed from 2002 to 2009 in order to determine their MACEs risk by using ACCI score.

**Results** Cox proportional hazard ratio model showed that AAC score as a continuous variable conferred 25% increased risk of MACE in average for each AAC point starting from zero. And when patients were divided into different groups by AAC scores, patients with AAC score more than 6 conferred an adjusted hazard ratio 4.88 (95% CI 3.84–6.19,  $p<0.001$ ), compared to those with AAC score 0–1.

**Conclusions** This is a national cohort study for the evaluation of MACEs for SLE patients. Our results demonstrated that SLE patients with higher ACCI score show significantly higher risk of cardiovascular events and the ACCI score could be applied as an index for MACEs evaluation.

### 200 PAN-DYSAUTONOMIA IN SYSTEMIC LUPUS ERYTHEMATOUS: TWO CASES OF SUCCESSFUL TREATMENT AND A REVIEW OF LITERATURE

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**Background and aims** Pan-dysautonomia hasn't been well studied in patients with systemic lupus erythematosus (SLE) and the optimal treatments are still to be determined. We report two cases of SLE with pan-dysautonomia and a review of the literature.

**Methods** Two cases with SLE complicated with pan-dysautonomia were carefully documented and described. Medline, EMBASE, Web of Science and PubMed were searched and 8 other cases were reviewed.

**Results** Case 1: A 23 year-old female SLE patient with central and peripheral nervous system (CNS and PNS) involvements for 5 years, presented pan-dysautonomia, including severe orthostatic hypotension, dysuria and gastro-intestinal pseudo-obstruction. Acetylcholine-receptors (AChR) antibody was detected in her cerebrospinal fluid (CSF). After treating with prednisone, tacrolimus, midodrine and fludrocortisone, her symptoms gradually improved in 12 months. Case 2: A 44 year-old female presented with pan-dysautonomia, anti-nuclear and anti-Ro antibody positivity and hypocomplementemia. AChR antibody titer also elevated in her CSF. After similar treatment, and additional plasma exchange for 6 times, all symptoms improved in 3 months. Combined with other 8 reported cases, 9 were female, 4 and 3 cases complicated with CNS and PNS involvement respectively, 3 cases had AChR antibody detected and all were positive, 4 cases received methylprednisolone pulse therapy. Cyclophosphamide ( $n=3$ ), azathiopurine ( $n=3$ ), and tacrolimus (our case 1) were also used. Plasma exchange was applied in our case 2.

**Conclusions** Pan-dysautonomia can be an initial symptom of SLE. AChR antibody may be useful in early recognising this rare disease. Plasma exchange along with prompt immunosuppressive therapy seem to lead to an early remission of the disease.

Abstract 200 Table 1 A review of 10 SLE cases complicated with pan-dysautonomia.

Case No	Case 1	Case 2	Patient 1	Patient 2	Patient 3	Patient 4	Patient 5	Patient 6	Patient 7	Patient 8
Author and year	Presenting, 2016	Presenting, 2016	Dale et al, 2012	Wong et al, 2011	Yukawa et al, 2008	Law et al, 2006	Jodo et al, 1992	Otokida et al, 1990	Arruda et al, 1989	Hoyle et al, 1985
Age/gender	23/F	44/F	13/F	14/F	32/F	28/F	42/F	37/M	72/F	21/F
Autoantibodies										
ANA	1:128	1:320	1:2560	1:1280	1:320	1:800	1:320	1:320	1:128	positive
Anti-dsDNA	negative	negative	positive	positive	positive	positive	positive	positive	negative	negative
Anti-Ro	positive	positive	negative	negative	negative	positive	positive	NM	negative	positive
Anti-La	positive	positive	negative	negative	negative	negative	positive	NM	negative	positive
Complement C3/C4	decreased	decreased	decreased	decreased	decreased	decreased	decreased	NM	normal	normal
CNS involvement	yes	no	no	no	yes	no	no	yes	no	yes
PNS involvement	sensory+ motor	no	no	no	sensory	no	no	sensory+ motor	no	no

NM: Not mentioned