an alanine tovaline substitution A1594V in CACNAIC. The proband (subject III-9), subjects II-5, II-11, and III-4 carried both mutations. Subjects II-13, III-7, and III-15 carried only MYH7-V878A and subjects III-2, III-16 and IV6 carried CACNA1C-A1594V. We did not found the two mutation in other members.

The family members with two mutations all showed inverted T-waves and ST segments depression in ECG recordings, severe left ventricular mashypertrophy [maximal left ventricular wall thickness (MLVWT) >18mm] in echocardiography and myocardial fibrosis in CMR. Among them, the subject III-9 was the most severe with MLVWT 28mm, LV outflow tract pressure gradient (LVOT_PG) 101mmHg, who received surgical septal reduction therapy. In the members with only MYH7-V878A mutation, the subject III-7 showed inverted T-waves, MLVWT 24mm and myocardial fibrosis, while subjects II-13 and III-15 all showed some abnormal repolarization, borderline MLVWT (11-12mm) and normal findings in CMR. In addition, the family members with only CACNA1C-A1594V mutation showed nearly normal in all examinations, except for subject IV-6 with inverted T-waves in ECG. Finally, the subjects with two mutations displayed more LV hypertrophy (MLVWT and LV mass index) and elevated LV filling pressure (E/e' ratio and LA diameter) than those with one mutation and non-mutation carriers ($p<0.05$).

CONCLUSIONS The subjects with MYH7-V878A mutation showed moderate or mild symptoms, and the subjects with CACNA1C-1594V mutation showed asymptomatic or very mild yet in this family. The pathogenesis effect of MYH7-V878A and CACNA1C-1594V mutations is thought to be cumulative, which lead to more severe symptoms in HCM.

GW27-e0526

Plasma level of big endothelin-1 in patients with hypertrophic cardiomyopathy

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OBJECTIVES The clinical expression of hypertrophic cardiomyopathy (HCM) is variable and largely unpredictable. Big endothelin-1 (ET-1) is the precursor of endothelin-1, which induce cardiomyocyty hypertrophy and intracellular myofibrillar disorganization. The purpose of this study is to observe the relationship between ET-1 and the prognosis of HCM patients.

METHODS A total of 245 consecutive patients with HCM were enrolled from 1999 to 2011 and partitioned to quartiles according to their plasma big endothelin-1 levels. The following clinical variables were recorded: medical history, symptoms, and alcohol intake, and smoking habit, current medications for coronary heart disease, hyperlipidemia, and diabetes. All patients underwent a complete cardiac evaluation, including physical examination, 12-lead electrocardiogram, M-mode, 2-dimensional and Doppler echocardiogram. At the entry and during follow up, the primary events were all-cause mortality, including cardiovascular death (SCD, heart failure-related death and fatal stroke) and non-cardiovascular deaths.

RESULTS At baseline, big endothelin-1 was positively correlated with N-terminal B-type natriuretic peptide ($r = 0.291$, $p < 0.001$), late gadolinium enhancement on magnetic resonance imaging ($r = 0.222$, $p = 0.016$) and the presence of NYHA class III/IV ($r = 0.192$, $p = 0.003$). During a follow-up of 5.1±3.0 years, big endothelin-1 level was positively associated with the risks of all-cause mortality, cardiovascular death, heart failure-related death and progression to heart failure. After adjusting for multiple factors related to survival and cardiac function, the significance remained in the association of big endothelin-1 with the risk of all-cause mortality (hazard ratio (HR) = 1.72, 95% confidence interval (CI) 1.11-2.67, p=0.016) heart failure-related death (HR = 1.86, 95%CI 1.08-3.19, p=0.025) and progression to heart failure (HR = 1.48, 95%CI 1.01-2.16, p=0.043).

CONCLUSIONS Our study showed that increased plasma big endothelin-1 was associated with poor prognosis in patients with HCM. The therapeutic effect of ET-1 blockers should be evaluated in HCM patients.

GW27-e0637

Zinc Levels in Left Ventricular Hypertrophy

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OBJECTIVES Zinc is one of the most important trace elements in the body and zinc homeostasis plays a critical role in maintaining cellular structure and function. Zinc dyshomeostasis can lead to many diseases, such as cardiovascular disease. Our aim was to investigate whether there is a relationship between zinc and left ventricular hypertrophy (LVH).

METHODS Design and Methods: A total of 519 patients was enrolled and measured their serum zinc levels in this study. We performed analyses the relationship between zinc levels and LVH and the four LV geometry pattern patients: normal LV geometry, concentric remodeling, eccentric LVH and concentric LVH. We performed further linear and multiple regression analyses to confirm the relationship between zinc and left ventricular mass (LVM), left ventricular mass index (LVMI) and relative wall thickness (RWT).

RESULTS Our data showed that zinc levels were 710.2±243.0μg/L in control group and were 641.9±215.2μg/L in LVH patient. We observed that zinc levels were715±243.5μg/L,694.2±242.7μg/L,643.7±225.0μg/L and 638.7±197.0μg/L in normal LV geometry, concentric remodeling, eccentric LVH and concentric LVH patients, respectively. We further found that there was a significant inverse linear relationship between zinc and LVM (p=0.001), LVMI (p=0.000) but did not show a significant relationship with RWT (p=0.561). Multiple regression analyses confirmed the linear relationship between zinc and LVM and LVMI remained inversely significant.

CONCLUSIONS The present study revealed that serum zinc levels were significantly decreased in the LVH patients, especially in the eccentric LVH and concentric LVH patients. Furthermore, zinc levels were significantly inversely correlated with LVM and LVMI.

GW27-e0884

The analysis of PECAM-1 and high sensitivity C-reactive protein in Hypertrophic cardiomyopathy patients

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OBJECTIVES To observe the correlation between inflammatory factor PECAM-1 and high sensitivity C-reactive protein (hs-CRP) in hypertrophic cardiomyopathy (HCM) patients.

METHODS 156 cases of patients with HCM were selected from two hospitals from January 2008 to December 2014. HCM diagnostic criteria in accordance with the ACC/ESC expert consensus hypertrophic cardiomyopathy (2003). Records of all patients age, sex, medical history, NYHA classification of cardiac function, the main indicators of ECG and echocardiography to exclude diabetes, infectious diseases, cancer, collagen diseases, application of immunosuppressive drugs. The contents of PECAM-1 and hs-CRP in all patients were determined by ELISA.

RESULTS The median value of PECAM-1 of 156 patients was 476.38μg/ml (475.61±39.47μg/ml); hs-CRP median value was 1.97 mg/L (1.12±2.75 mg/L), two indicators were higher than normal. 51 cases (32.7%) underwent coronary angiography showed that 12 cases (7.7%) had coronary heart disease, these patients and those without coronary heart disease had no statistically significant difference between the two indicators. The levels of PECAM-1 and hs-CRP increased associated with the largest increase in wall thickness ($P < 0.05$); significant T-wave inversion ($P < 0.01$); chest pain or syncope symptoms ($P < 0.05$).

CONCLUSIONS HCM may be associated with the inflammatory response, particularly in those patients with wall thickening, significantly T wave changes, chest pain or syncope symptoms.

GW27-e0893

Clinical and MRI characteristics of end-stage hypertrophic cardiomyopathy

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OBJECTIVES The end-stage hypertrophic cardiomyopathy (ES-HCM) is part of the clinical spectrum of HCM. Systematic evaluation of

clinical and magnetic resonance imaging (MRI) characteristics of this spectrum has not been performed. The purpose of this study was to clarify the clinical and MRI characteristics in ES-HCM patients.

METHODS We collected 72 ES-HCM patients characterized by systolic dysfunction (left ventricular eject function < 50%). According to the cardiac structure and hemodynamics, ES-HCM patients were divided into those with dilated phenotype (D-ES, n=42) and those with restrictive phenotype (R-ES, n=30). All of the patients underwent MRI.

RESULTS ES-HCM developed at a wide age range (12 to 70 years), with 35% of patients < 40 years old. The clinical manifestation did not differ significantly between the two groups, including chest distress (41), dyspnea (49), palpitation (20), amaurosis (7), syncope (7) and edema of lower extremity (18). The New York Heart Association functional class≥2 in all patients. The incidence of atrial fibrillation, intraventricular block, nonsustained ventricular tachycardia were 46%, 28%, 18% respectively. The maximum wall thickness were over 15mm in 67 patients. Regional LV wall thickness was decreased in 26 patients (23 in D-ES vs. 3 in R-ES, $p < 0.05$). The left and right atrial anteroposterior diameter (LAD, RAD) were 47.33mm, 44.67mm in D-ES and 54.57mm, 63.57mm in R-ES. The left and right ventricular short axis diameter were 65.86mm, 31.33mm in D-ES and 48.10mm, 32.40mm in R-ES. There was significant difference between the two groups in LV end diastolic volume (130.77 ml/m2 in D-ES, 61.12 ml/m2 in R-ES, $p < 0.05$). Late gadolinium enhancement (LGE) demonstrated focal or diffuse LGE at the LV wall in 100% of ES-HCM. LGE volume fraction was significantly larger in D-ES (37.9%) than in R-ES (19.6%) ($p < 0.05$).

CONCLUSIONS This study demonstrated that patients with end-stage HCM have expanded clinical expression and MRI characteristics, including dilated phenotype and restrictive phenotype. Besides, the D-ES patients had more LGE distributed diffusely throughout ventricular septum and LV free wall than the R-ES patients. MRI has an important application value in the diagnosis and evaluation of ES-HCM.

GW27-e0966

Treatment evaluation by exercise-stress tests of catecholaminergic polymorphic ventricular tachycardia

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OBJECTIVES Catecholaminergic polymorphic ventricular tachycardia (CPVT) is a lethal autosomal dominant heritable arrhythmia syndrome. Patients diagnosed with CPVT are at high risk of life-threatening ventricular arrhythmias if untreated. Beta-blockers or a combination with calcium channel blockers/flecainide are cornerstone in China, but the the efficacy for prevention of arrhythmic events in CPVT is unkown. This study aimed to evaluate treatment efficacy by exercise-stress tests.

METHODS A retrospective analysis of all exercise-stress test data during following-up for 20 CPVT probands (12 male, 60.0%) diagnosed in the Department of Cardiology, of Beijing Tsinghua Changgung Hospital and Peking University People's Hospital between September 2006 and March 2016.

RESULTS Followed changing lifestyle, all patients received enduring largest dose beta-blockers, four patients combination with calcium channel blockers, and one combination with flecainide. Exercise-stress tests after medication showed increasing maximum workload ($p<0.05$), improving ventricular arrhythmia score significantly ($p<0.05$) and non-sustained ventricular tachycardia (NSVT) were suppressed in 14 patients (14/20,70.0%). However the threshold of ventricular arrhythmia had a lower trend and bigeminal/frequent VPBs decreased significantly (137.3±19.6 vs.122.5±13.5, $p<0.05$). ICD implantation was recommended in five patients who experienced NSVT despite optimal medical management and changed lifestyle, only one receving dual chamber pacemaker. During the follow up of 44.7±66.6 months, emotion stress induced sudden cardiac death associated with noncompliance occurred in two patient and the remaining 18 patients would not experience syncope.

CONCLUSIONS Conventional therapy beta-blockers is incompletely effective in preventing arrhythmic events in patients with CPVT, combination with calcium channel blockers or flecainide is needed.

GW27-e1008

Native T1 and T2 Mapping by Cardiovascular Magnetic Resonance in detecting altered myocardial composition with increased afterload in left and right heart diseases

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OBJECTIVES Recent developments in CMR imaging techniques are enabling rapid parametric mapping of magnetic relaxation properties (T1 and T2 relaxation times). Quantified myocardial T1 values, T2 values could demonstrate the underlying composition of the myocardium. It is still not completely clear how these relaxation times compare in patients with right sided afterload increase such as pulmonary arterial hypertension (PAH) and left sided afterload increase such as severe aortic stenosis (AS). The aims of this study were to observe whether cardiovascular magnetic resonance could characterize native myocardial T1 values, T2 values in patients with pulmonary arterial hypertension (PAH) and severe aortic stenosis (AS) in comparison to healthy volunteers and to evaluate the association of T1 values, T2 values with ventricle function and remodeling.

METHODS CMR was performed for in 16 patients with PAH (62±10 years, 9 female), 17 patients with AS (85±5 years, 7 female) and 5 volunteers (38±17 years, 2 female) on a 1.5T MRI scanner. T1 and T2 mapping in addition to ventricular volumes and function of the left and right ventricle (LV and RV) were performed and the values were compared using Student t tests and Pearson's correlation.

RESULTS Compared with healthy volunteers, patients with PAH had significantly elevated T1 at the septum (1072.4±64.3 ms vs 1023.2±15.3 ms, $p=0.017$) and RV insertion points (1137.8±81.8 ms vs 1037.6±8.4 ms, $p=0.00$). They also have higher T2 values at RV insertion points (54.1±5.4 ms vs 42.6±1.5 ms, $p=0.001$) and RV free wall (46.6±3.3 ms vs 40.5±1.4 ms, $p=0.021$). Patients with AS had significantly elevated T1 values at the LV free wall (1026.8±53.0 ms vs 966.9±18.8 ms, $p=0.006$). At the septum, T1 showed a trend of increase in patients with AS ($p=0.053$) and T2 showed the same trends at RV insertion in patients with AS ($p=0.068$). No correlation was found between RV insertion point T2 or septal T1 and RV volume, or ejection fraction. A correlation trend was found between RV insertion point T1 and RV mass Index ($p=0.06$). No correlation was found between LV free wall T1 and LV volume, ejection fraction, or LV mass index.

CONCLUSIONS T1 and T2 values are both elevated in the RV insertion points and the RV free wall of patients with PAH, but only T1 values were elevated in in the free LV wall in patients with severe aortic stenosis. This combination of T1 and T2 values demonstrate potentially different remodeling process in the right compared to the left ventricle with increased afterload.

GW27-e1045

Alteration in right heart function upon high-altitude exposure and its roles in Acute mountain sickness

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OBJECTIVES We aimed to observe the alterations of right heart function by different altitudes and time phase, explore its roles in AMS and try to provide new clues and direction in clinical diagnosis, treatment and prevention for AMS.

METHODS The echocardiography examinations were performed by ultrasonography using the S5-1 cardiac probe (CX50, Philips, USA) by a senior technician. Most of the valuable parameters have been measured including right atrial diameter (RA), right ventricular end-diastolic diameter (RV), pulmonary artery (PA), pulmonary acceleration time (AT), tricuspid blood flow velocity E peak, tricuspid blood flow velocity peak A, the right ventricular outflow tract blood flow to the end time (b), tricuspid blood flow velocity at the a peak termination to the next new Tung Chau tricuspid blood the velocity E peak time interval (a) ECG R-wave peak to the right ventricular outflow tract ejection terminate at the time interval (c). Each parameter has been measured four consecutive cardiac cycles. Myocardial performance index (MPI) Tei index, mean pulmonary artery pressure (mPAP), pulmonary vascular resistance (PVRTot), blood flow velocity in the tricuspid peak E\A peak were calculated according the parameters abovementioned.