showed myocardial hypertrophy and granular appearance of the myocardium should be considered in the diagnosis of CA. Cardiac magnetic resonance imaging is valuable in the diagnosis of CA.

**GW26-e2244**  
**Gene Mutations in Chinese with Arrhythmogenic Right Ventricular Dysplasia/Cardiomyopathy—a cohort registry study**  
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**OBJECTIVES**  
Arrhythmogenic right ventricular dysplasia/cardiomyopathy (ARVD/C) is an inherited cardiac disease associated with an increased risk of arrhythmic sudden death. Mutations in desmosomal genes and some extra-desmosomal genes have been identified to associate with ARVD/C. Previously we identified 5 novel plakophilin (PKP2) mutations in a cohort of Chinese patients with ARVD/C. Our present study is to determine the prevalence of other associated gene mutations in this ARVD/C registry study and explore the potential genotype-phenotype relationship.

**METHODS**  
Genotypic and phenotypic profiles were studied in a cohort of 22 symptomatic Han Chinese with clinical diagnosis of or suspected ARVD/C according to modified international Task Force criteria in 2010. Direct sequencing of 5 desmosomal genes and 3 extra-desmosomal genes was performed by 3730XL DNA Analyzer.

**RESULTS**  
22 mutations including 13 novel (13/22, 59.1%) in 5 desmosomal genes PKP2, Desmplakin (DSP), Desmoglein-2 (DSG2), Desmocollin-2 (DSC2), Plakoglobin (JUP) were identified in 20 (20 of 32, 55.6%) patients in our cohort. No mutations were found in extra-desmosomal genes. Among 32 patients, 11 (1 of 32, 34.4%) patients have PKP2 mutations, 3 (9.4%) DSP, 3 (9.4%) DSG2, 6 (18.8%) DSC2 and 4 (12.5%) JUP. Multiple mutations were found in 6 subjects (6 of 32, 18.7%). In which, 3 have PKP2 mutation, 3 DSC2, 3 DSP and 3 JUP. Genotype-phenotype analysis indicates compound multiple mutations may predict major structural abnormalities.

**CONCLUSIONS**  
PKP2 mutation is the most common gene mutations in our ARVD/C cohort. A higher percentage of DSC2 and JUP mutations were identified in the cohort compared with previous reports. Compound multiple mutations are common and may indicate major structural abnormalities. Extra-desmosomal gene mutations are rare in our Chinese ARVD/C cohort.

**GW26-e2283**  
**Incremental Value of Contrast Echocardiography in the Diagnosis of Left Ventricular Noncompaction**  
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**OBJECTIVES**  
Contrast echocardiography with left ventricular opacification (LVO) can improve endocardium definition and potentially becomes supplement of conventional two-dimensional echo (2DE) in the diagnosis of noncompaction of the ventricular myocardium (NCVM). This study aimed to access the feasibility, accuracy, reproducibility of LVO & its incremental value than 2DE in NCVM diagnosis.

**METHODS**  
LVO & 2DE were performed in 85 patients (54 men, mean age 40±20 years) with suspected NCVM (NCVM Gp), and 2DE were performed in 40 healthy volunteers (Normal Gp, 20 men, mean age 40±23 years). The LV chamber size and LV ejection fraction derived from Biplane Simpson’s formula were compared among LVO-NCVM Gp, 2DE-NCVM Group & 2DE-Normal Gp. The location and extent of NCVM were evaluated based on AHA/ACA 16 segment model for LV segmentation, and the thickness ratio of noncompacted to compacted myocardium (NCR) were assessed on LVO & 2DE by 2 independently blinded experienced echo-cardiologists.

**RESULTS**  
(1) Compared with the Normal Gp, the NCVM Gp showed larger LVEDV (89.8±11.6ml vs. 45.1±15.3ml), LVEDV (121.1±15.5ml vs. 95.1±14.8), LVEFS (43.0±3.44mm vs. 33.4±0.4mm), LVEFS (74.3±3.94ml vs. 44.4±4.3ml), lower LVEF (40.8±13.2% vs. 65.6±7.1%), and E/A ratio (0.82±0.32 vs. 1.62±0.5) using 2DE method (p<0.05).  
(2) Within the NCVM Gp, compared with the values from 2DE method, LVEDV (65.2±7.8ml vs. 58.9±11.6ml), LVEDV (162±14.8ml vs. 121.1±15.5ml), LVEFS (47.8±5.67mm vs. 43.0±3.44mm), LVEFS (84.7±2.46ml vs. 74.3±3.94ml) derived from LVO method were larger and LVEF (38.2±12.4 vs. 40.8±13.2) on LVO was slightly lower (p<0.05).  
(3) Among the whole 1360 LV segments in NCVM Gp, there were more segments adequately visualized for analysis on LVO than on 2DE (1278 vs. 1143, 93.97% vs. 86.99%). There were no more noncompaction areas detected on LVO than on 2DE (314 vs. 239, 29.09% vs. 19.26%). Of the 921 segments interpreted as normal on 2DE, 52 segments (5.65%) were noncompacted on LVO. NCVM on LVO were majorly located in medium (53.18%), apical (46.15%) segments and lateral wall (99.81%); rarely involved in basal segment (0.64%).  
(4) NCR on LVO was greater than that on 2DE (4.21±1.3 vs. 3.3±1.2, P<0.0001), but they are highly related and both showed excellent interobserver consistency. The coefficient of inter-observer variability of NCR was slightly smaller using LVO than 2DE (5.2% vs. 6.6%).

**CONCLUSIONS**  
Contrast echocardiography can clinically improve the diagnosis of NCVM in accuracy, sensitivity & reproducibility, and act as a useful supplement to the routine two-dimensional transthoracic echo.