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Prevalence and natural history of left ventricular apical aneurysms in hypertrophic cardiomyopathy

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Purpose: To define prevalence and natural history of patients with hypertrophic cardiomyopathy (HCM) and apical aneurysms (AA).

Methods: A single center-cohort consisting of 423 patients (49.3±17.2 years, 66.2% male) was followed up for a median of 84 months (range 6 to 480 months). A left ventricular (LV) AA was defined as a discrete, thin-walled dyskinetic or akinetic apex with a relatively wide communication to the LV cavity recognized both by means of echocardiography and magnetic resonance imaging, Figure 1. Cumulative SD event rates through follow up were estimated by Kaplan-Meier method and differences were assessed by log rank test.

Results: AA were recognized in 11 out of 423 patients in our cohort (2.6%). Nine out of 11 patients (81.8%) with AA presented also with midventricular obstruction (MVO), with apical aneurysm formation identified in more than one fourth of patients with MVO (26.5%). During follow up, 4 out of 11 patients with an AA (36.4%) experienced progression to end stage HCM (burnt out) or death due to heart failure (HF). With the survival rates from HF and associated death being significantly lower in the aneurysm group [5 year survival 74.1% [95% CI (58-90.2)] for the aneurysm group compared to 99% [95% CI (98-98.8)] of the rest of the cohort, log rank p=0.001. Marginal non significant differences were recorded concerning the survival from sudden death and surrogate endpoints [5 year survival 97.6% [95% CI (96.8-98.4)] for the non aneurysm group and 80% [95% CI (67.4-92.6)] for the AA group, log rank p=0.056.

Conclusions: AA is a distinct phenotypic characteristic of HCM associated with an unfavorable prognosis in terms of progression to end stage HCM (burnt out) or death due to HF. Additionally, there is a clear connection between MVO and apical aneurysms.

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Prevalence and outcome of cardiogenic shock in patients with tako-tsubo cardiomyopathy


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Purpose: Tako-tsubo cardiomyopathy (TTC) is regarded a benign disease since left ventricular (LV) function returns to normal within a short period of time. However, severe complications have been described in a limited number of patients (pts).

This study evaluated the frequency and outcome of cardiogenic shock in a large TTC registry.

Methods: From 37 heart centres, 324 pts (296 f, 28 m, age 68 ± 12 years) were included in the registry of the Arbeitsgemeinschaft Leitende Kardiologische Krankenhausärzte according to the following criteria: 1) acute chest symptoms, 2) ischaemic ECG changes, 3) reversible LV akinesia corresponding to a single coronary artery territory, 4) absence of coronary artery stenoses. Complete data on complications were available in the last 209 registry pts.

Results: Complications developed in 108/209 pts (52%) within 2.6 ± 2.9 days (median 1 [IQR 1-3] days) after symptom onset; 51 of these pts (24%) experienced > 1 and 23 (11%) > 2 complications. Most complications (77%) occurred within 3 days after symptom onset, however, 23% developed later (from day 4 to 56). Fourteen of 209 pts (7%) experienced cardiogenic shock which developed on the day of admission in 11 pts (7%) and from day 2 to day 4 after admission in 3 pts (21%). Seven of these patients (50%) were also in pulmonary oedema. The ECG on admission showed a higher heart rate (92 ± 24 vs. 76 ± 17 beats per minute, p < 0.001) and more Q waves in pts with cardiogenic shock (54% vs. 27%, p = 0.043). Cardiac markers were significantly higher (creatinine kinase 4.3 ± 3.57 vs. 1.8 ± 3.8 [p < 0.05] and troponin 62.1 ± 109 vs. 10.7 ± 11.9 [p < 0.001] times the upper limit of normal, and ejection fraction lower (38.9 ± 5.1 vs. 51.1 ± 8.9, p < 0.005). Intracavitary balloon pumping was applied in 2/14 pts, and catecholamines were administered in 8 pts. Four of 14 TTC pts with cardiogenic shock (29%) died, 2 from multiple organ failure and 1 pt each from refractory cardiogenic shock and LV rupture. In the latter pt ST-segment elevation persisted for 3 days when myocardial perforation occurred.

Conclusion: Cardiogenic shock occurs in 7% of pts with TTC. The prevalence of cardiogenic shock is similar to findings in reported myocardial infarction (6-10%). However, the mortality of cardiogenic shock in TTC pts appears to be lower (29%) than reported in reperfused patients with ST-elevation myocardial infarction (42-62%). This may be due to the early spontaneous reversibility of LV dysfunction in TTC.
Cardiomyopathies: from pathogenesis to treatment

function and histological characteristics. Colonization of endothelial markers and mesenchymal markers were identified using confocal immunofluorescence staining. Western blot was performed to evaluate the TGF-β/Smad and Wnt/β-catenin signaling pathway.

Results: A mice model of CBB3 myocarditis was made and BMP7 was administrated to CBB3-infected mice. Histological data demonstrated that BMP7 administration increased inflammatory cells accumulation and cardiac fibrosis in response to CBB3 challenge. Echo data described cardiac dysfunction was recovered after treatment of BMP7 intervention. Double labeling of endothelial and mesenchymal markers showed the treated mice had significantly reduced the double-positive cells. Western blot described that TGF-β/Smad and Wnt/β-catenin signaling pathway was involved in this pathogenesis.

Conclusions: BMP7 interventions TGF-β1 induced endothelial-to-mesenchymal transition in viral cardiomyopathy through both TGF-β1/Smad and Wnt/β-catenin signaling pathway. The research is supported by grant from the Health Joint-research Program of China and Canadian Institutes of Health Research (81010007) and National Natural Science Foundation of China (31070786).

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Systematic review of atrial fibrillation and stroke in patients with hypertrophic cardiomyopathy

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Background: Hypertrophic Cardiomyopathy (HCM) is a common inherited myocardial disorder associated with arrhythmia. Current guidelines for the management of Atrial Fibrillation (AF) do not give detailed advice on management in HCM patients because of a lack of robust clinical prediction tools that can estimate risk of developing AF and an absence of adequately powered studies.

Objective: To summarise and critically review literature on AF and stroke in HCM patients and to perform a meta-analysis to determine clinical risk factors for AF and stroke.


Study selection: Published articles on clinical studies addressing the topic of HCM, AF and stroke in human subjects. Data extraction: 2 investigators independently extracted data from identified articles. Studies in English language investigating AF and stroke in HCM as primary or secondary endpoint were included. Reviews, case reports, abstracts were excluded.

Data synthesis: A random effect meta-regression model was used to determine the pooled overall prevalence and incidence of AF and stroke. The I2 statistic was used to estimate the proportion of total variability in the data attributed to the heterogeneity between the studies.

Results: In a total population of 5397 patients in 29 retrospective studies, the overall prevalence of AF was 21.61% (95% CI 19.30-23.92, I2=67.3%, p<0.001). The overall prevalence of thromboembolism in HCM patients with AF was 30.65% (95% CI 23.35-37.95, I2=85%, p<0.001). The overall incidence of AF was 2.99% (95% CI 1.81-4.14, p<0.001). The overall incidence of all thromboembolic events in HCM patients with AF was 4.35% (95% CI 2.91-5.79, I2=62.4%, p<0.005). LA size and age were predictive for the development of AF and stroke in the majority of studies. Meta-analysis revealed an LA diameter of 38.03 mm (95% CI 34.62-41.44) in SR patients and 45.37 mm (95% CI 41.64-49.04) in AF patients. There were no randomized controlled trials of therapy: antiarrhythmic was associated with a lower incidence of stroke in a small number of studies, but data on the effects of pharmacological therapy and radiofrequency ablation were limited and contradictory.

Conclusions: AF is common in HCM and associated with a high risk of thromboembolism. LA size and age are independently associated with AF, but the published literature is insufficient to create a robust tool for the prediction of AF or thromboembolic risk in patients in SR. There are no trials of therapy, but most data suggest that once patients have AF, they should be anticoagulated.

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Value of magnetic resonance and exercise echocardiography to predict outcome in patients with hypertrophic cardiomyopathy

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1University Hospital A Coruña. Department of Radiology, A Coruña, Spain; 2University Hospital A Coruña. Department of Cardiology, A Coruña, Spain; 3Wall Motion Abnormalities (WMAs) during Exercise Echocardiography (EE) are associated to events in Hypertrophic Cardiomyopathy (HCM). We aimed to evaluate Magnetic Resonance (MR) and EE to predict outcome in HCM.

Methods: In 131 HCM patients (56% and MR (perfusion and delayed hyperenhancement (DHYPER) were performed in 148 pts with HCM (age 51±15 years), normal LV function (LVEF≥50) and absence of history of coronary disease.

Results: During follow-up (5.9±2.7 years), there were 6 hard events (4 cardiac failure, 1 stroke, 1 appropriate discharge of defibillator) and 25 combined events (including new atrial fibrillation and syncope). WMAs at EE were seen in 13 cases (8.7%), perfusion defects (PDef) in 11 (7.4%) and DHYPER in 47 pts (31.8%). WMAs were seen in 50% of pts with hard events vs. 7% of those without hard events (p<0.009), and in 20% and 7% of pts with and without combined events (p<0.046). A PDef and/or DHYPER in ≥3 segments (sg) was seen in 50% and 15% of pts with and without hard events (p=0.035), and in 36% and 12% of pts with and without combined events (p=0.003). Univariate predictors of hard events were peak double product (DP) (p=0.03), peak wall motion score index (PWMSI) (p<0.001). Univariate predictors of combined events were a NYHA functional class ≥2 (p=0.02), left atrial size (p=0.03), DP (p=0.03), resting WMSI (p=0.03), PWMSI (p<0.001), and MR data (no. sg with PDef, p=0.003; no. sg with DHYPER, p=0.004; PDef and/or DHYPER in ≥3 sg and WMs at ExE, the group with negative results by both techniques had better outcome (log rank test 12, p=0.008).

In conclusion, PDef and/or DHYPER by MR are associated to WMAs during EE in pts with HCM. MR may help to predict outcome in them.

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Presence of chronic kidney disease is associated with poor clinical outcomes during hospitalization in patients with takotsubo cardiomyopathy: multi-center registry from tokyo CCU network

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Background: It is well known that the presence of chronic kidney disease (CKD) increases the risk of the cardiovascular disease. However, whether the presence of CKD adversely affects in-hospital clinical outcomes in patients with Takotsubo cardiomyopathy (TC) remains to be determined.

Methods: We investigated 219 patients of TC from Tokyo CCU Network database, comprising of 67 cardiovascular centers in the metropolitan area during January 1, 2010 to December 31, 2011. We estimated the glomerular filtration rate (eGFR) in these patients in whom serum creatinine level was measured. We attempted to characterize TC complicated by CKD by comparing patients of TC complicated by CKD with non-complicated TC patients. We evaluated the differences in clinical outcomes in patients according to the presence of CKD.

Results: A total of 131 patients were included in the analysis. The prevalence of CKD was 31.8% (95% CI 25.5-38.1). The mean age was 65.2±13.5 years and the gender was 80.6% (95% CI 75.1-85.1) male. The mean eGFR was 65.6±22.7 ml/min/1.73m2. The overall prevalence of AF was 21.6% (95% CI 18.6-24.6). The mean heart rate (HR) was 104±26.4 bpm. The mean left ventricular ejection fraction (LVEF) was 36.2±12.9%. The mean number of hospitalization was 3.5±2.9. The mean number of days of mechanical ventilation was 2.5±2.3. The mean number of days of intravenous positive pressure ventilation was 2.3±1.9. The mean number of days of inotropic support was 2.3±1.9.

Conclusion: The presence of CKD is associated with poor clinical outcomes during hospitalization in patients with Takotsubo cardiomyopathy (TC) remains to be determined.