ATRIAL FIBRILLATION IN HYPERTROPHIC CARDIOMYOPATHY

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HYPERTROPHIC CARDIOMYOPATHY

- Common genetic disorder
- Autosomal dominant
- 1:500
- Major cause of sudden death
- Mutations in sarcomeric proteins in 55%
- A left ventricular (LV) wall thickness ≥15 mm not explained by loading conditions is considered diagnostic.

Elvis Presley
January 8, 1935 – August 16, 1977
Singer, Musician, Actor

DNA analysis reveals genetic heart defect that could have killed Elvis

- Elvis Presley died of a heart attack in August 1977, aged 42
- DNA from his hair offers new theory as to the cause of his death
- Scientists found that he was genetically prone to heart muscle disease
- Analysis also found ‘mutations’ causing obesity, glaucoma and migraines
AF and HYPERTROPHIC CARDIOMYOPATHY

• AF is the most common arrhythmia in HCM
• Increased risk of stroke and heart failure
• Thromboembolic events
• 48 hour Holter monitoring is recommended in initial assessment of HCM
• Atrial fibrosis is the main pathophysiological process
AF and HYPERTROPHIC CARDIOMYOPATHY

- 12 to 28% of HCM patients have AF
- AF was the initial disease presentation in 10% of HCM
- Asymptomatic AF may lead to underestimation of true prevalence
Predictors of AF in HCM

- Age
- Genetics: The Arg663His (rs371898076) mutation in the myosin heavy chain beta (MYH7) gene
- CHF
- RV involvement
- LA size
- Biomarkers

Am J Cardiol 1999;83:13–18H.
Outcomes of AF in HCM

- A combined cohort from Italy and the USA demonstrated an increased risk of HCM-related death in patients with comorbid AF (OR 3.7, 95% CI [1.7–8.1]).
- In a sub-group analysis, those who developed AF at ≤50 years of age had an increased risk of HCM-related mortality and progression of symptoms.
- Increase risk of composite endpoint in patients undergoing myectomy who developed perioperative AF.
- Stroke associated with AF was found to be the cause of 13% of HCM related deaths in a consecutive cohort of 744 patients with HCM.
A.

Proportion Surviving Free of All-Cause Deaths

Years from Initial Evaluation

p=0.34

Red line: HCN with AF
Blue line: HCN without AF
p=0.46
Medical Treatment of AF in HCM

• Rate vs. Rhythm control strategy?
• Beta-blockers, diltiazem and verapamil for rate control
• Amiodarone for rhythm control (side effect??)
• Disopyramide recommended as a second line especially in symptomatic LVOTO
• Caution is needed in light of the potential for enhanced AV conduction and associated increased ventricular rate in AF.
Catheter ablation of AF in HCM

- Small limited studies showed some benefit
- Redo procedures were required in 52% of cases
- Continuous antiarrhythmic medications were required in 54% of cases
- HCM was an independent risk factor for AF recurrence following multiple procedures (HR 2.42, 95% CI [1.06–5.55])
Surgical Management of AF in HCM

HCM and atrial fibrillation surgical management algorithm

NYHA III-IV
Refractory or intolerant to medical treatment
No atrial fibrillation detected

isolated extended septal myectomy

LVOT obstruction gradient ≥50 mmHg
(Fixed or Progressive)

NYHA ≥ II or syncope or angina +
Atrial Fibrillation

Extended Septal Myectomy and AF management

paroxysmal AF

LAD <45 mm
Bipolar Radiofrequency
Pulmonary vein isolation

LAD >45 mm
Full Biplateral Cox-Maze
(Either Cox-Maze IV or
cut-and-sew Maze)

persistent AF
Long-standing
persistent AF
Permanent AF

LAD <60 mm
LAA amputation or
occlusion
Full Biplateral Cox-Maze
procedure

LAD >60 mm

Coronary sinus
Circumflex coronary artery

Mitr al Annulus

Tricuspid Annulus

SVC

IVC

Cryoblation

Surgical incision

Radiofrequency Ablation

doi: 10.21037/acs.2017.05.08
Catheter Ablation vs Maze for AF in HCM
Stroke Risk in HCM

- Incident rates of stroke in HCM, irrespective of AF diagnosis, have been estimated as 2.5%/year.
- Compared with patients with HCM in sinus rhythm, those in AF were shown to have an eightfold increase in stroke risk (21% versus 2.6%) in a 480-patient cohort (107 AF cases) over a follow-up period of 12.6 ± 7.7 years.
- A metanalysis showed an overall increase risk of 3.75%.
- Thromboembolic events in patients with AF occurred on average 3.5 ± 3.4 years after AF diagnosis.
Thromboembolism occurred significantly less frequently in patients taking anticoagulant prophylaxis. Left atrial size, number of AF episodes, and AF category (paroxysmal or permanent) were not reliable markers of the risk of stroke. Furthermore, of the 18 study patients with AF (6%) who had an embolic stroke, 5 experienced this event after a single episode of AF. As in previous HCM studies, the CHA₂DS₂-VASC score did not predict the stroke risk. Therefore, because of the potentially devastating effects of thromboembolic events, anticoagulation should be taken into consideration in every patient with HCM and AF, independently of left atrial size and number and duration of AF episodes.
<table>
<thead>
<tr>
<th>Guideline</th>
<th>Issuing body</th>
<th>Year</th>
<th>Patients requiring anticoagulation</th>
<th>Anticoagulation agent (1st line)</th>
<th>Strength of recommendation</th>
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<tr>
<td>ESC guidelines on diagnosis and management of hypertrophic cardiomyopathy</td>
<td>ESC</td>
<td>2014</td>
<td>All patients with HCM and AF</td>
<td>VKA</td>
<td>Class I</td>
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<td>ESC guidelines for the management of AF</td>
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<td>No preference between VKA and NOAC for HCM</td>
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<td>2011</td>
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<td>No specification of VKA or NOAC</td>
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<td>Guidelines for diagnosis and treatment of patients with HCM</td>
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<td>2012</td>
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<td>Guidelines for the pharmacotherapy of AF</td>
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<td>2013</td>
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<td>Class Ia</td>
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ACC = American College of Cardiology; AHA = American Heart Association; ESC = European Society of Cardiology; HCM, hypertrophic cardiomyopathy; HRS = Heart Rhythm Society; JCS = Japanese Circulation Society; NOAC = non-vitamin K antagonist oral anticoagulant; VKA = vitamin K antagonist.
Stroke Prevention in HCM: VKA or NOAC??

- No data
- All NOACs major trials excluded HCM
- In a retrospective cohort of patients with HCM on anticoagulation and found no significant difference between NOACs and vitamin K antagonists in the rate of ischaemic stroke (HR 1.37, 95% CI [0.40–4.67]) or major bleeding (HR 0.75, 95% CI [0.36–1.57]).
- A recent post hoc subgroup analysis of the (RE-LY) study has shown that the presence of LV hypertrophy determined by ECG criteria lead to decreased warfarin efficacy (dabigatran 150 mg versus warfarin HR 0.48, 95% CI [0.29–0.78]).
AF in HCM: Aswan Heart Centre Experience

- 649 HCM patients (Aswan and Cairo)
- 65 patients (10%) have documented AF
  - Younger group
  - 28% of total patients have palpitations without documentation
  - Need more Holter monitoring
  - genotype??
- Paroxysmal AF: 80% (mean age: 39.5, mean LA diameter: 42.6)
- Permanent AF: 20% (mean age: 46.6, mean LA diameter: 44.2)
AF in HCM: Aswan Heart Centre Experience

- All strokes reported were before start of anticoagulation
- All on anticoagulations
- NOACs or Warfarin according to patient
- No documented denovo persistent or paroxysmal Afib post myectomy after 6 months follow up
- Amiodarone for short term for post myectomy Afib, long term on beta blockers
- No ablations, 1 biV pacemaker
AF in HCM: Future Directions

- AF ablation in selected-high volume centres
- LA fibrosis by MRI-LGE and risk of recurrence
- Maze procedure concomitant with myectomy
- Genotype-phenotype association
- Ablate and pace
- NOACs
AF in HCM:
Final thoughts

• A common clinical finding
• Prognosis of AF in HCM has improved with current management strategies
• Room for improvement regarding catheter ablation results
• Anticoagulate regardless of CHADSVASC Score
• Screening and early detection is key to safe and effective management