

A potpourri of fascinating cases to learn from

Hey Doc – I can't do what I used to.....

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No Disclosures



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Terry Jones RIP
Feb 01, 1942 - Jan 21, 2020



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Case



58 year old Tri-athlete presents with 6/12 history of decrement in performance

- competes in 4-5 Olympic triathlons a year (1.5km swim, 40km bicycle, 10km run).
- competes on team USA for Aquabike



He has dropped from the top 5th percentile in his age to the bottom 50th percentile in triathlon. Main symptom is SOB with occasional light-headedness with maximal exertion.

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Case



Pmed/Sur Hx:

- Not relevant

Fam Hx:

- Father MI age 68 (smoker).
- No family history of premature SCD, CAD, aortopathy or valvular disease.

Social History:

- Non-drinker/non-smoker.
- Attorney in Medical Negligence.
- Life-long Athlete

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Physical examination

BP 122/78mmHg

Pulse 56bpm

Slightly delayed carotid upstroke.

S1 normal. S2 present but soft, grade III/VI ESM throughout the precordium.

No change with dynamic maneuvers.

No radio-fem delay

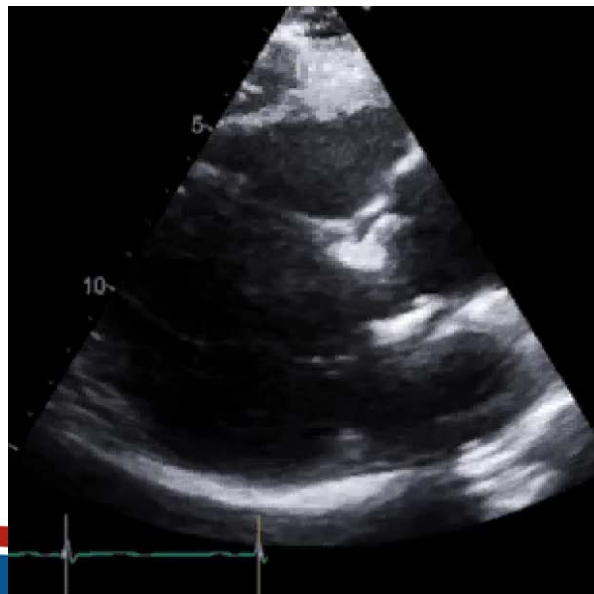
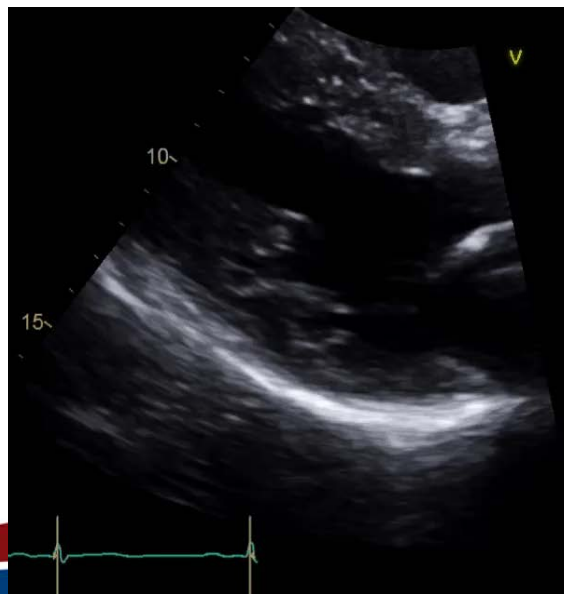
ECG



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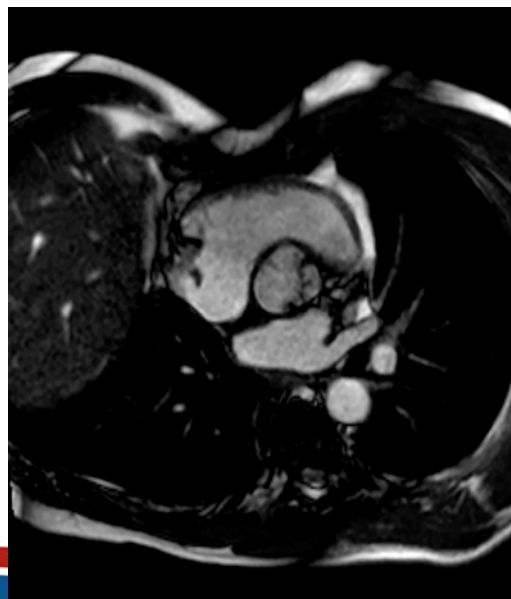
Echo

ASE American Society of Echocardiography



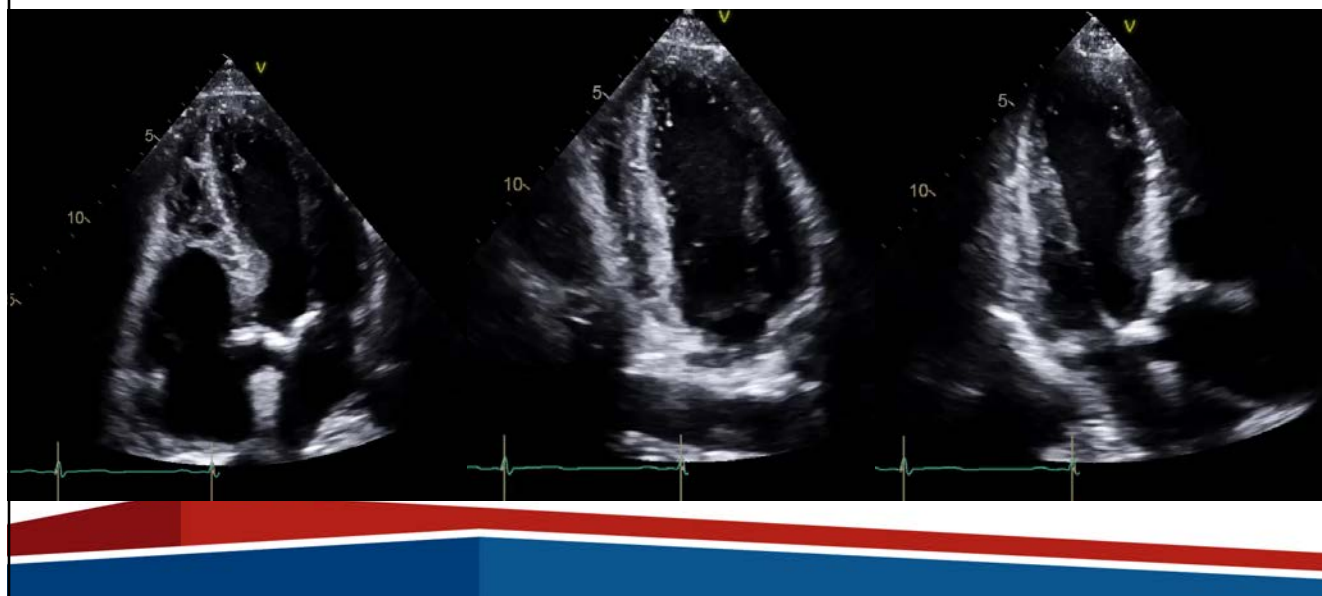
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Echo/CMR



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Echo



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Echo report



Normal biventricular function

Low normal LV size with LVH (septum 1.6cm)

Bicuspid Aortic Valve with Moderate Aortic Stenosis

Mid ascending aorta 4.4cm

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Diagnosis: BAV, moderate AS and dilated aorta.



Which of the following is true?

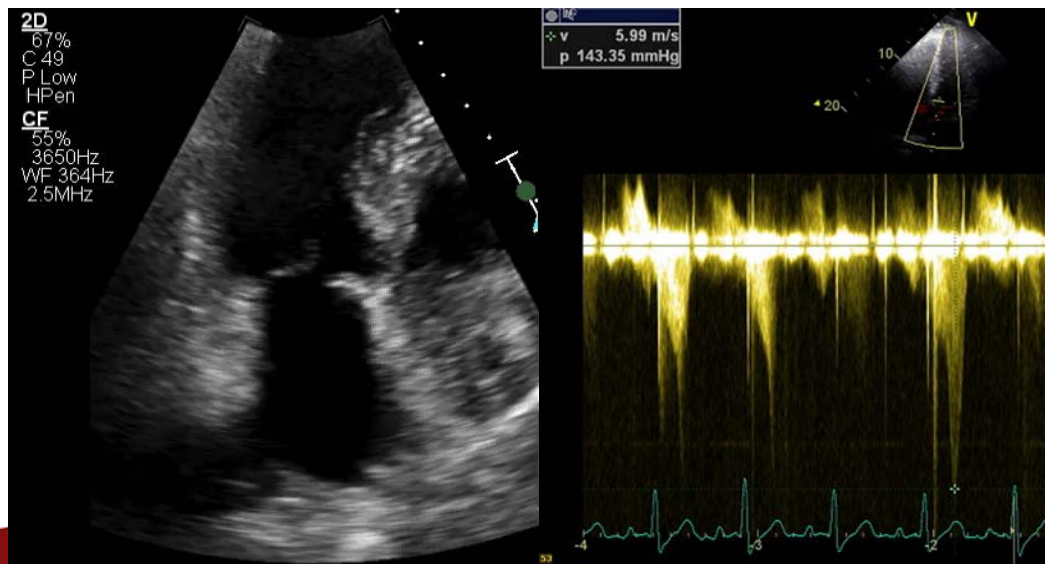
1. Athletes with moderate AS should not participate in any sports with the exception of 1A
2. Exercise testing to at least the level of activity in competition is appropriate.
3. Athletes with BAV and any dilation of the aorta should not participate in any competitive sports.

Athletes with moderate AS (stage B) can participate in low and moderate static or low and moderate dynamic competitive sports (classes IA, IB, and IIA) if exercise tolerance testing to at least the level of activity achieved in competition and the training regimen demonstrates satisfactory exercise capacity without symptoms, ST-segment depression, or ventricular tachyarrhythmias, and with a normal blood pressure

For athletes with a BAV and a mild to moderately dilated aorta (score 2 to 3.5 or aortic root or ascending aortic diameters measuring 40 to 42 mm in men or 36 to 39 mm in women) and no features of associated connective tissue disorder or familial TAA syndrome, participation in low and moderate static and dynamic competitive sports with a low likelihood of significant bodily contact (classes IA, IB, IC, IIA, IIB, and IIC) may be considered. For these athletes, avoidance of intense weight training should be considered (Class IIb; Level of Evidence C).

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Stress echo



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Stress Echo Report



9.9 METS (Average). 96%MAPHR.

No Ischemia (ECG or Echo).

Normal BP response.

No tachyarrhythmias.

Systolic Anterior Motion (SAM) of the MV with dynamic gradient of approximately 140 mmHg.

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Diagnosis: BAV, moderate AS, dilated aorta and dynamic LVOT obstruction

Which of the following is true?

1. He most likely has genetic HOCM.
2. An ICD is appropriate.
3. He should definitely not play any sports.
4. Septal reduction surgery and AVR is indicated
5. Trial of medical management is appropriate

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Cardiac MRI



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Cardiac MRI



The left main travels between the aorta and the right ventricular outflow tract at the level of the pulmonary valve, for a distance of 13 mm from its origin, before bifurcating into the LAD and circumflex.

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Diagnosis: BAV, moderate AS AND dilated aorta AND dynamic LVOT obstruction AND an anomalous coronary artery

Which of the following is true?

1. He is at risk of SCD due to the anomalous coronary artery and this should drive the decision regarding surgery.
2. According to ACC guidelines a BAV with aorta of 4.7cm can participate in 1A sports.
3. He is a no risk of continuing with Aquabike (he is a medical negligence lawyer – can't foresee any issues with this).
4. Surgery is not indicated at this time.

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Next Steps...



Trial of BB – did not tolerate. Switched to diltiazem and returned for stress echo 3 months later.

“On his initial evaluation he achieved 9.9 Mets at which time his dynamic gradient was approximately 140 mmHg. With introduction of diltiazem he increased his functional capacity to 15 METS with a reduction in the dynamic gradient (~30mmHg).”

Respectfully declined any advice on exercise limitations and continues to compete. No issues in 5 years of follow up.

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Follow up 2019



- **Feels well. Mean gradient across the AV now 45mmHg at rest**
- **Still achieves 13.5 METS on stress testing – dynamic gradient ~90mmHg with stress.**
- **BP response is normal.**
- **BNP normal.**
- **Plans to compete in a triathlon next weekend**
- **I persuade him to have surgery AVR/myectomy/ascending aorta replacement(?indication)**
- **Biopsy result: TTR amyloid!!**

You know
nothing
John Snow



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