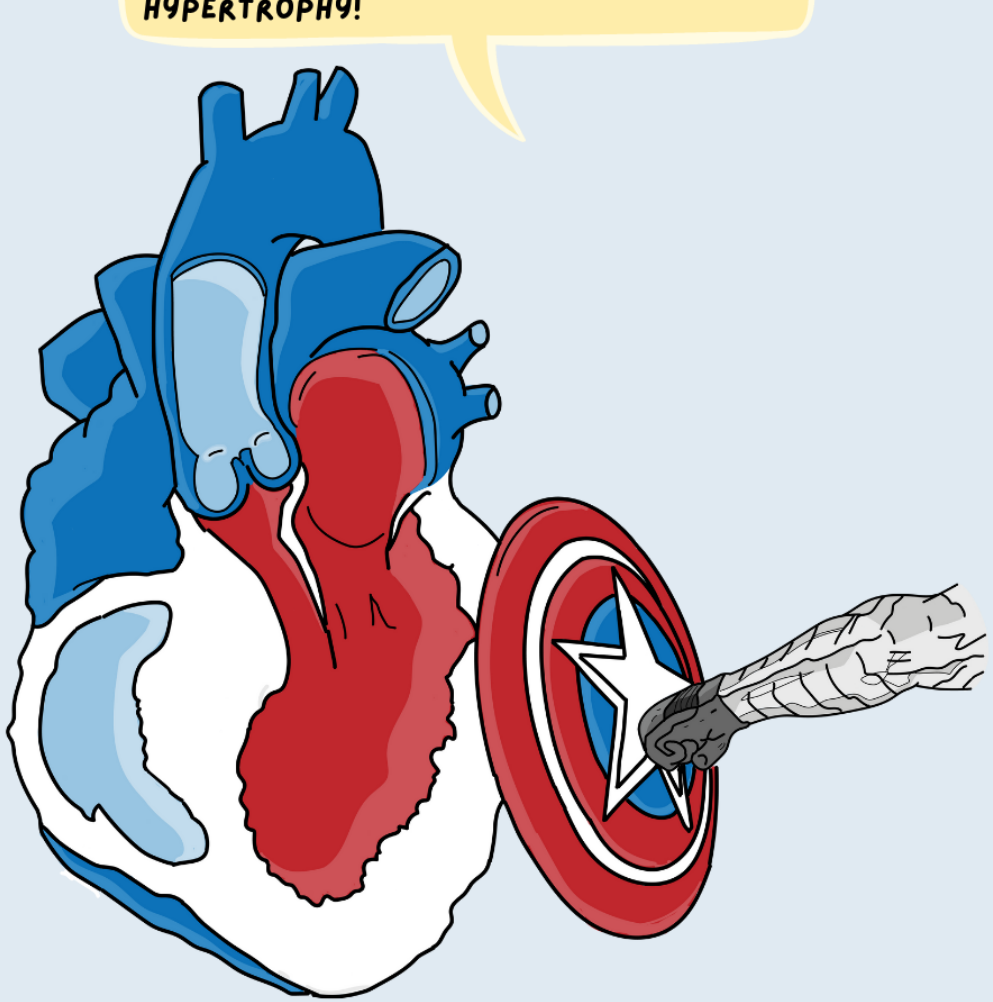

CARDIO NERDS
PROOF THAT TONY STARK HAS A HEART

HYPERTROPHIC CARDIOMYOPATHY



TYPICAL FEATURES

- MYOCARDIAL HYPERTROPHY (=>15MM WALL THICKNESS) DISPROPORTIONATE TO STIMULUS I.E ABSENCE OF HTN, AS, PRESSURE OVERLOAD
- ASYMMETRIC PROXIMAL SEPTAL HYPERTROPHY IS TYPICAL - SEPTAL TO POSTERIOR WALL RATIO 1.3:1

OTHER FEATURES

2D ECHO
- HYPERTROPHY OF INFERIOR, ANTERIOR, OR LATERAL WALLS
- APICAL HYPERTROPHY
- HYPERTROPHY OF RV
- ISOLATED HYPERTROPHY OF PAPILLARY MUSCLES (RARE)

DOPPLER
- CONTINUOUS WAVE DOPPLER FOR MID-CAVITARY OBSTRUCTION - LOBSTER CLAW FLOW AND/ OR LATE PEAKING FLOW

CONTRAST
- APICAL HYPERTROPHIC VARIANT - SPADE SHAPED
- APICAL HCM AND APICAL ANEURYSMS CAN BE MISSED WITHOUT CONTRAST!

PITFALLS

- SEPTAL TO POSTERIOR WALL RATIO ALONE SHOULD NOT BE USED AS A MARKER OF HCM
- A NORMAL AGING HEART & OTHER DISEASE STATES MAY RESULT IN A SIMILAR SEPTAL TO POSTERIOR WALL RATIO!

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DIFFERENTIAL FOR HYPERTROPHIC HEART

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ATHLETES' HEART (YOUNGER PATIENTS)
- TYPICAL WALL THICKNESS <=13MM IN ATHLETES' HEART AND >15MM IN HCM

HYPERTENSIVE HEART DISEASE WITH LVH (OLDER PATIENTS)
- HIGHER THRESHOLD OF SEPTAL TO POSTERIOR WALL RATIO = 1.5:1 TO DX HCM
- STRAIN RATE IMAGING TO DIFFERENTIATE NON-OBSTRUCTIVE HCM FROM HYPERTENSIVE LVH

SUB-VALVULAR FIXED AORTIC STENOSIS (ANY AGE)
- HIGH INDEX OF SUSPICION - MEMBRANE MAY BE HARD TO VISUALIZE
- ECHO - EARLY PEAKING FIXED SIGNAL (EARLY SYSTOLE) VS HCM (DYNAMIC LATE SYSTOLE)

PEARL: AORTIC REGURGITATION IS RARE IN HCM BUT COMMON IN PATIENTS WITH A FIXED OUTFLOW OBSTRUCTION

CONSIDER MIMICS

- LV INFILTRATION - AMYLOIDOSIS, GLYCOGEN & LYSOSOMAL STORAGE DISEASE

PEARL: PROMINENT INTRAMYOCARDIAL VASCULATURE IN THICK WALLS FAVORS HYPERTROPHIED MYOCARDIUM OVER AN INFILTRATIVE PROCESS

- LVH WITH ANTEROSEPTAL ISCHEMIA
- STRESS CARDIOMYOPATHY
- MITOCHONDRIAL CYTOPATHIES
- FRIEDREICH'S ATAXIA

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OBSTRUCTIVE PHYSIOLOGY

LVOT GRADIENT >30MMHG AT REST

QUANTIFICATION OF OUTFLOW TRACT OBSTRUCTION

PULSED WAVE DOPPLER

ALIASING

MECHANISM: EJECTION VELOCITY > NYQUIST LIMIT

CONTINUOUS WAVE DOPPLER

DAGGER-SHAPED MID- TO LATE-SYSTOLIC PEAK

MECHANISM: MAXIMAL GRADIENT AFTER > LV STROKE VOLUME HAS BEEN EJECTED

DDX: AS OR MR - SYMMETRIC PEAK

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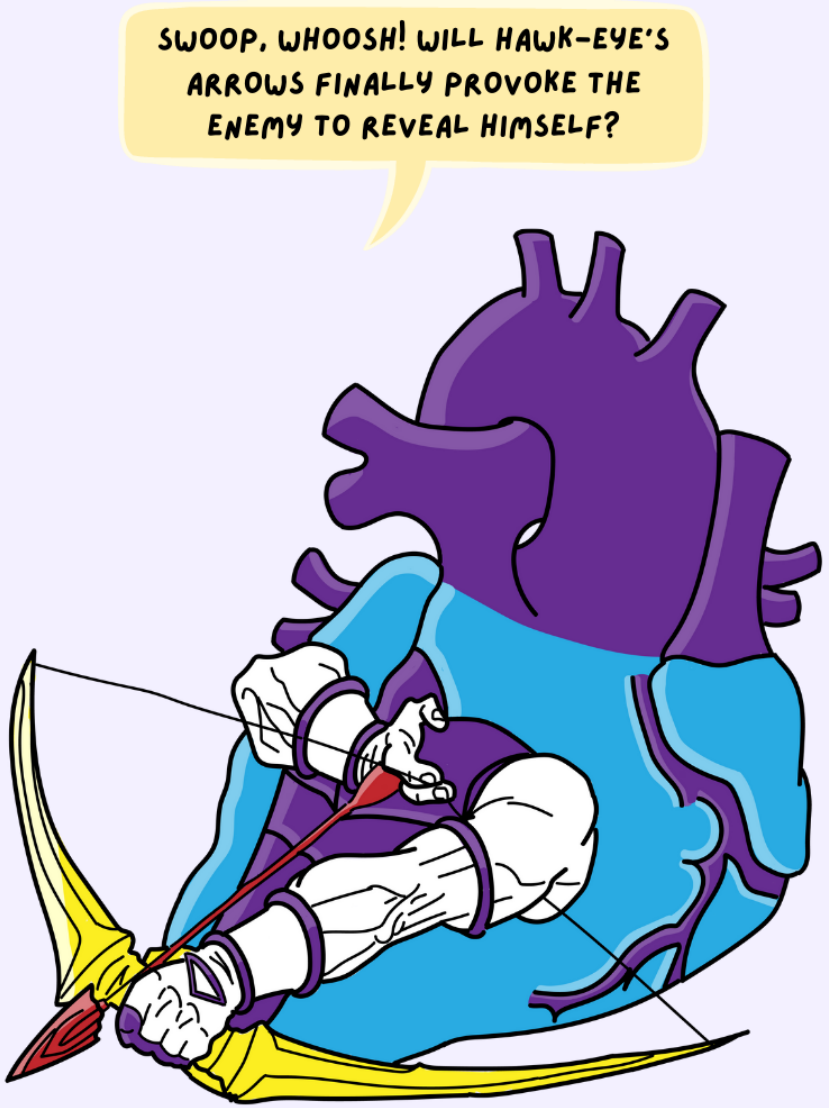
OBSTRUCTIVE WITH PROVOCATION



OBSTRUCTIVE WITH PROVOCATION
LVOT GRADIENT >50MM HG

NON- PHARMACOLOGIC >>	PHARMACOLOGIC	PHYSIOLOGIC
•EXERCISE STRESS ECHO USEFUL WHEN SYMPTOMS OCCUR AFTER MEALS •VALSALVA USEFUL IN THOSE WHO CANNOT EXERCISE	•AMYL NITRATE •ISOPROTERENOL •DOBUTAMINE	•FOLLOWING A PVC/POST-COMPENSATORY PAUSE •PHYSIOLOGIC STRESS

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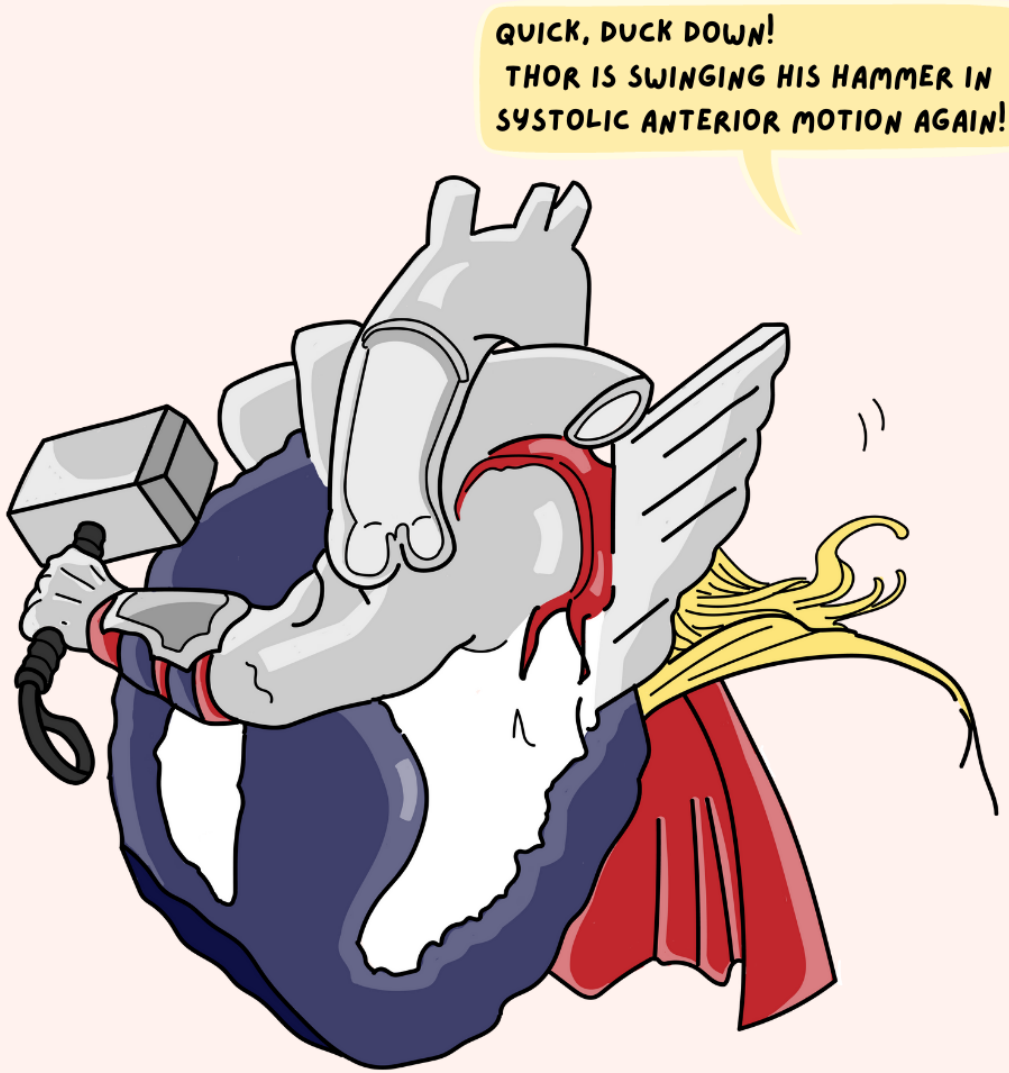
SYSTOLIC ANTERIOR MOTION OF MITRAL VALVE



MECHANISM

DRAG FORCES DURING VENTRICULAR SYSTOLE > VENTURI FORCES (LIFT!) WITH ANTERIORLY DISPLACED PAPILLARY MUSCLES

ON ECHO?

- MID-SYSTOLIC NOTCHING OF THE AORTIC VALVE
- CONTACT OF ANTERIOR MITRAL LEAFLET/CHORDAE WITH SEPTUM
- GREATER OBSTRUCTION WHEN MITRAL LEAFLET IN CONTACT WITH VENTRICULAR SEPTUM FOR >40% OF SYSTOLIC CYCLE

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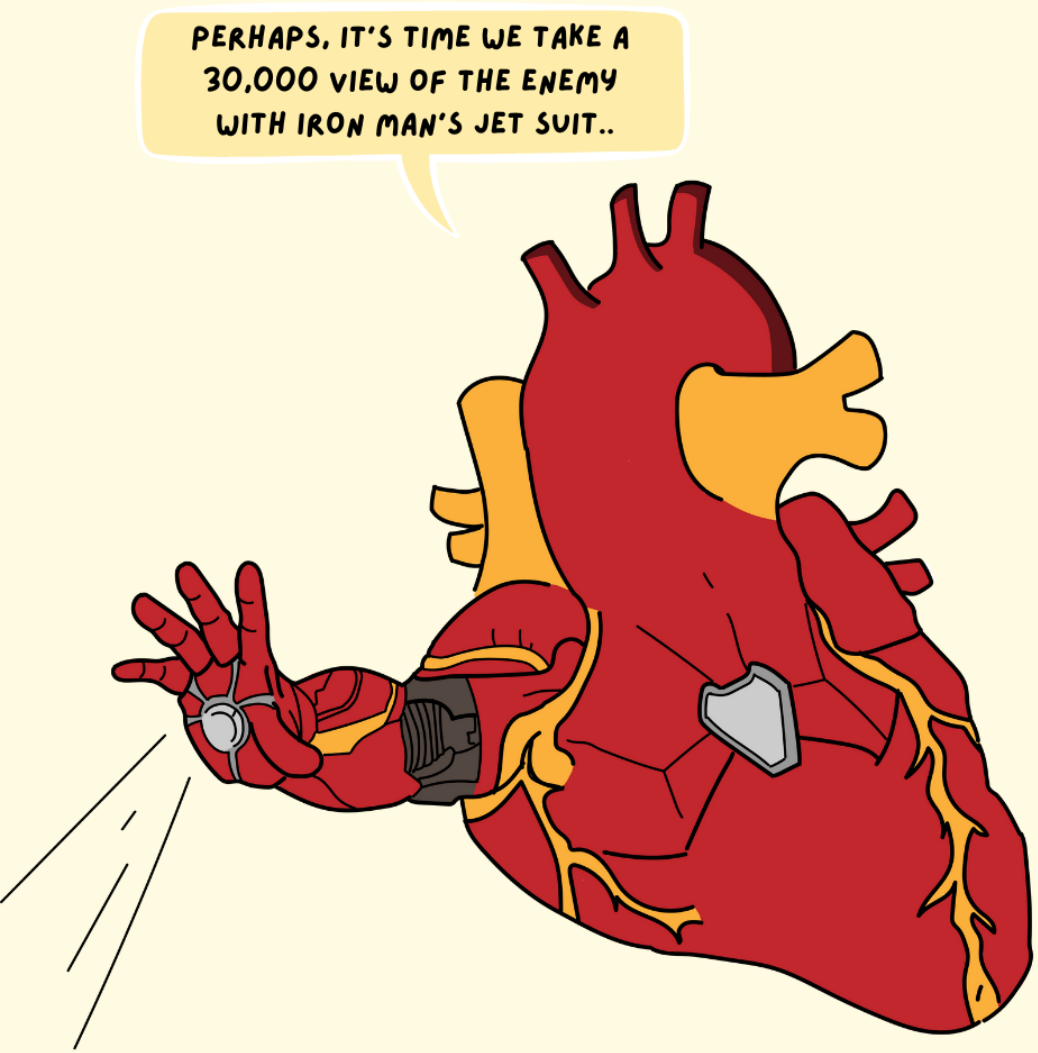
MITRAL REGURGITATION



MITRAL REGURGITATION

MECHANISM?

MAL-COAPTATION OF MITRAL VALVE LEAFLETS DURING SAM

ON ECHO?

- POSTERIORLY DIRECTED JET
- MID- TO LATE- SYSTOLIC PEAK (VS EARLY IN STRUCTURAL MR)
- LATE PEAKING VELOCITY >6M/S (VS LVOT)

ABNORMAL STRUCTURE OF MITRAL VALVE

- HYPERTROPHY OF PAPILLARY MUSCLES

  OTHERS: ACCESSORY PAPILLARY MUSCLE, ANOMALOUS INSERTION OF PAPILLARY MUSCLE

- INCREASE IN LEAFLET AREA
- LEAFLET ELONGATION

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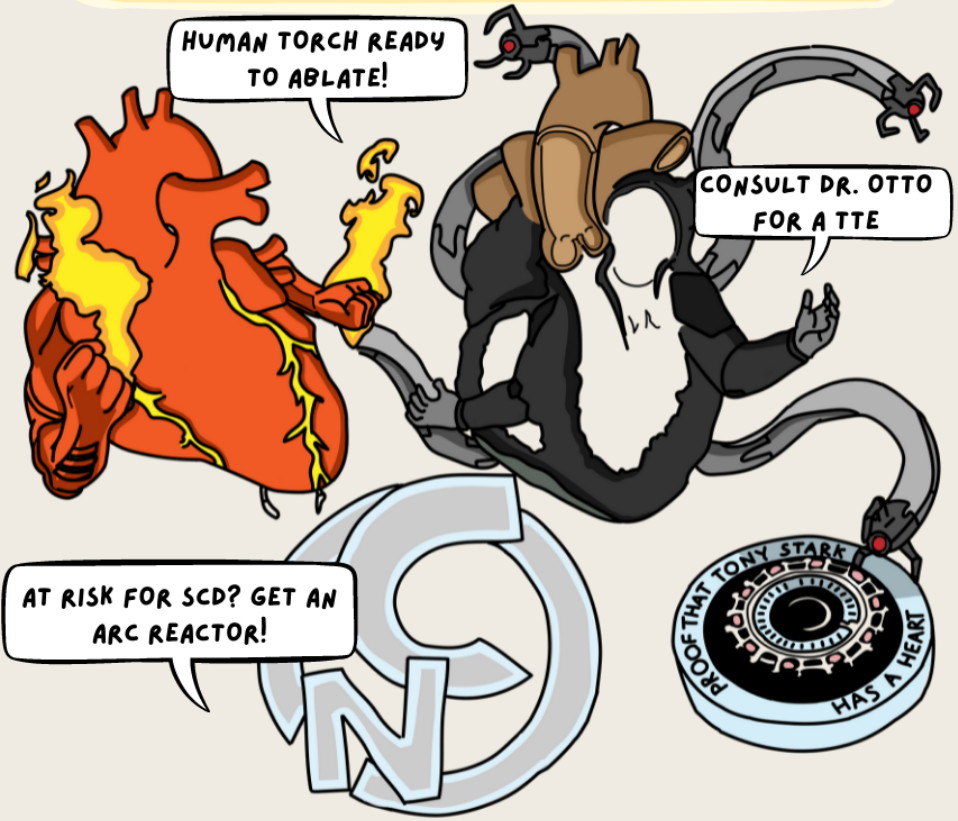
PROGNOSIS

HCM IS INHERITED AS A AUTOSOMAL DOMINANT DISORDER AND GENETIC COUNSELLING SHOULD BE PROVIDED TO ALL PTS & FAMILIES!
GENETIC TESTING CAN BE PERFORMED IN ACCORDANCE WITH WISHES OF THE FAMILY, BUT IS NOT ROUTINE UNLESS A PATHOGENIC VARIANT HAS BEEN IDENTIFIED IN THE PROBAND!



DISEASE PROFILE	ON ECHO	MANAGEMENT
BENIGN/STABLE	•HYPERCONTRACTILE NON-DILATED LV WITH DIASTOLIC DYSFUNCTION	•SERIAL TTE EVERY 1-2 YRS.
PROGRESSIVE HF (OBSTRUCTIVE)	•DYNAMIC LVOT OBSTRUCTION WITH MR AT REST OR EXERCISE PROVOCATION •REST OR PROVOKED GRADIENT =>30MMHG PREDICTS FUTURE HF PROGRESSION FROM NYHA CLASS I/II TO III/IV	•DRUGS: NEGATIVE INOTROPES - B-BLOCKERS, VERAPAMIL, DISOPYRAMIDE; MYOSIN INHIBITORS - MAVACAMTEN! •CANDIDATES FOR SURGICAL MYOMECTOMY/ALCOHOL SEPTAL ABLATION
ADVANCED HF & END-STAGE (NON-OBSTRUCTIVE)	•SYSTOLIC DYSFUNCTION EF <50% WITH REMODELING (VENTRICULAR ENLARGEMENT WITH LV WALL THINNING)	•CANDIDATES FOR HEART TRANSPLANTATION •CARDIAC RESYNCHRONIZATION THERAPY BRIDGE TO TRANSPLANT
HIGH RISK FOR SCD	•MASSIVE LVH > 30MM •UNEXPLAINED SYNCOPE •HX OF SCD, VF OR SUSTAINED VT •FAMILY HX OF SCD FROM HCM •EF <50% •LV APICAL ANEURYSM •CMR WITH LGE >15% •NSVT	•PRIMARY PREVENTION ICD

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