Introduction:

Welcome to CUGH's bi-weekly clinical case-series, "Reasoning without Resources," by Prof. Gerald Paccione of the Albert Einstein College of Medicine. These teaching cases are based on Prof. Paccione's decades of teaching experience on the medical wards of Kisoro District Hospital in Uganda. They are designed for those practicing in low resource settings, Medicine and Family Medicine residents, and senior medical students interested in clinical global health. Each case is presented in two parts. First comes a case vignette (presenting symptoms, history, basic lab and physical exam findings) along with 6-10 discussion questions that direct clinical reasoning and/or highlight diagnostic issues. Two weeks later CUGH will post detailed instructors notes for the case along with a new case vignette. For a more detailed overview to this case-series and the teaching philosophy behind it, see Introduction to "Reasoning without Resources". Comments or question may be sent to Prof. Paccione at: gpaccion@montefiore.org

Note: If you would like to be notified when a new case is posted (along with instructor notes for the previous one), send your e-mail to Jillian Morgan at jmorgan@CUGH.org.

About the Author:

Dr. Gerald Paccione is a Professor of Clinical Medicine at the Albert Einstein College of Medicine in the Bronx, New York. His career has centered on medical education for the past 35 years – as a residency Program Director in Primary Care and Social Internal Medicine at Montefiore Hospital, and director of the Global Health Education Alliance at the school. He has served on the Boards of Directors of Doctors for Global Health, Doctors of the World USA, and the Global Health Education Consortium. Dr. Paccione spends about 3 months a year in Uganda working on the Medicine wards of Kisoro District Hospital where he draws examples for the case studies.

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CASE 41 – Pumping Fast

A 22 year old male, a farmer married with 1 child, was previously in good health until 6 months ago when he began to experience episodes of sharp right-substernal chest pain associated with "heart pumping very fast" that occurred especially when walking fast or working hard. The heart pumping would start suddenly, last approximately 1-15 minutes, and stop suddenly. It occurred about once every week 6 months ago, but now occurs every 2-3 days, more with exertion, but even while sitting and lying down.

Two weeks ago he began to feel increasingly fatigued and short of breath while working. Four nights ago he awoke from sleep with dyspnea, right substernal chest pain and the "heart pumping" which lasted longer than usual. He didn't work over the past 3 days. He's had no fevers, cough or sputum. He usually sleeps flat, and hasn't woken short of breath until the episode 4 nights ago.

Physical Exam:

Looks well, thin, in no distress sitting upright
BP 100/60 R 15, HR 80, regular; T 37
HEENT: conjunctiva normal without icterus, pallor; fundi benign; mouth no thrush;
neck: no JVP at 45 degrees, ⊕ HJR; no nodes; thyroid normal; carotid +2, sharp upstroke
lungs: clear
cor. PMI: non-sustained, vigorous LV heave, 2 cm lateral to the MCL, 4 cm diameter;
RV lift, asynchronous with LV heave, "rocking motion" of chest wall;
S_1 normal intensity; P_2↑'d left upper sternal border;
⊕ S_3 apex, ↑'d with expiration
4/6 holosystolic murmur apex → axilla, medium pitch
2/4 diastolic decrescendo high-pitched murmur LSB spanning diastole,
↑'d with hand-grip (held for 1 minute)
2/4 diastolic rumble apex without OS or pre-systolic crescendo; ↑'d with expiration
abdomen: normal, without hepato-splenomegaly or tenderness
extremities: no edema; pulses +2-3 throughout
Neurologic: mental status, CN, motor, sensory, cerebellum, gait intact

1. **What is the clinical significance of each of the findings on physical exam?**

- *no JVP, edema, or hepatic tenderness: indicate normal RV pressures*

- *⊕ HJR: Hepato-jugular reflux (HJR) can be seen with increased left or right ventricular end-diastolic pressures (LVEDP or RVEDP), and thus can be a sign of left and/or right-sided heart failure – either one or both. With RV failure, the already elevated right sided pressures become higher as applied abdominal pressure increases venous return to a heart that can't expand to accept it, and the neck veins engorge and pulsate. With left-sided CHF, the ↑'d LVEDP at rest pushes the septum to the right and decreases the RV's capacity to accept the increased venous return, thus inducing a + HJR as well.*

  When the JVP is normal, a (+) HJR is a valuable as an indicator of left-sided CHF. In right-sided CHF, the JVP is already elevated and so the +HJR doesn't add independent diagnostic information to what's already known about the (elevated) right sided pressure (although it can be useful in corroborating the (+)JVP). But in LV failure, the JVP is normal and the HJR maneuver "unmasks" the high LV pressures.

- *PMI 4cm, displaced: indicates LV dilation;*
  non-sustained, vigorous: indicates a "volume load", not a "pressure load" on the ventricle.
  "Pressure loads", as with hypertension or aortic stenosis, cause concentric hypertrophy in which the LV muscle thickness increases out of proportion to the LV chamber and produces a sustained, relatively non-displaced PMI. "Volume loads" as occur in valvular disease, produce a vigorous contraction that's non-sustained.

- *RV lift/LV heave, asynchronous: The asynchronous heave-lift combination suggests significant MR.*
  A parasternal (RV) lift can be caused by an excessive volume/pressure load on the right ventricle causing RV dilatation and/or hypertrophy - as in tricuspid regurgitation, cardiac shunts (e.g.ASD), or pulmonary hypertension. However, it can also result from left atrial expansion from severe MR. The LA lies behind the RV, and its expansion from a regurgitant bolus of blood from the LV pushes the RV forward causing a late-systolic parasternal lift.

  The left chest thus seems to rock back and forth: first the LV heave, followed by the parasternal lift. This finding suggests severe MR.

- *S_3: Localized to the LV by its increase with expiration, the S3 is caused by sudden deceleration of blood flowing into the ventricle, tensing of the mitral chordae, and/or tapping of a dilated LV against the chest wall at the end of the rapid filling phase of diastole.*
  The S3 is a sign of a dilated ventricle and usually dysfunction/CHF in adults older than 35 years. However, an S3 can be heard normally in the young (he is 22 y.o.) and

in severe MR without implying ↑'d LVEDP. However in AI, it suggests either an ↑'d LVEDP (i.e.CHF) or severe valvular insufficiency.

- *↑ P_2: Suggests increased pulmonary artery pressure (PAP), due to both increased pulmonary venous pressures from the ↑'d LAP/ LVEDP, and reflex pulmonary arteriolar vasoconstriction to protect the pulmonary capillary bed.*

- *Murmur 1: holosystolic, from apex to axilla: indicates MR, radiating along the ribs; 4/6, medium pitch: suggests severe MR. A murmur of grade 3 intensity or louder suggests severe MR (LR 4.4, McGee p.496). Medium to low pitched MR murmurs suggest a large regurgitant volume with a decreased pressure differential between the LV and LA. The more common high-pitched murmurs of MR are usually hemodynamically insignificant.*

- *Murmur 2: diastolic decrescendo, high pitched, left sternal border (LSB): Indicates AI; 2/4, spanning diastole: Suggests moderate – severe AI, duration of an AI murmur correlates with the severity of the lesion; Increased with handgrip: Handgrip, (without breath-holding, and maintained for >20 seconds), increases peripheral vascular resistance and afterload, accentuating the regurgitant murmurs of both AI and MR. Application of bilateral BP cuffs inflated above systolic pressure is a better, but somewhat more cumbersome maneuver.*

- *Murmur 3: 2/4 diastolic rumble at the apex: this is probably an "Austin – Flint" plus mitral inflow murmur (see below). The increase with expiration indicates it's left-sided.*

2. What is the differential diagnosis of left-sided apical "rumbles" and how do you differentiate them?
What does the rumble heard in this patient most likely represent?

- *Pitch of a murmur reflects the pressure gradient across the valve: "rumbles" or low-pitch murmurs imply a low pressure gradient and high flow (e.g. between the LA and LV in diastole). "Rumbles" are heard with the bell lightly applied to the chest wall, often to a very focal area near the apex. They can be clearly and then barely audible within a centimeter.*

- *The 3 principle causes of mitral rumbles are:*
  - *Mitral Stenosis;*
  - *Austin Flint (AF) murmur of AI, caused by a moderate-to-severe regurgitant jet from the aorta pushing the anterior mitral leaflet into the mitral inflow tract inducing a partial obstruction and leaflet fluttering.*
  - *mitral inflow murmur accompanying severe MR, without obstruction.*

- *In a patient with an AI murmur, an associated Austin-Flint murmur is more likely than associated Mitral Stenosis if:*
    - *no loud S1 or Opening Snap of mitral valve is heard (but AI can decrease both the S_1 and an OS)*
    - *S_3 is present: implies dilated LV, therefore less severe (if any) MS (although AI can cause dilatation and an S3 by itself)*
    - *no pre-systolic crescendo to the rumble*

- *in this patient with obvious LV enlargement, a prominent murmur of MR, and no prior history of LVF symptoms (seen early in MS), significant MS is unlikely. The "rumble" is due to a combination of Austin-Flint from AI plus mitral inflow from MR vibrating the mitral valve leaflets.*

3. What are indicators of severity in aortic insufficiency and what complicates their interpretation in this patient? (from McGee, Ch. 41)

The indicators of severity of AI (with their respective Likelihood Ratios (LR)) include:

- *diastolic BP <50 (LR 19); pulse pressure >80 (LR 11)*
    - *if DBP >70 or pulse pressure (PP) <60, it's unlikely to be severe AI.*

- *Hill's test: foot-arm <u>simultaneous</u> SBP difference >60 (normal <20) (LR 17)*

- *Austin-Flint suggests moderate-severe AI and↑ LVEDP*

- *S_3 suggests mod-severe AI and/or ↑ LVEDP*

In this patient, MR complicates the picture, increasing the diastolic rumble, contributing to the S_3, enlarging the PMI, and decreasing the PP (by providing an "escape route" for blood exiting the LV and thereby diminishing the SBP).

4. What is the specific cause of this patient's symptoms – the shortness of breath, "heart pumping very fast" and the chest pain? Why was he in good health until recently?

- *This young man has severe RHD causing principally MR and AI. However, through "eccentric hypertrophy" (in which the chamber expands in proportion to the wall thickness), the heart can tolerate and adapt to a severe volume overload for 10-20 years before LVEDP rises and CHF develops. In this patient, myocardial decompensation and left-sided heart failure is now occurring with increasing dyspnea.*

- *His "heart pumping very fast", usually precipitated by exertion but recently occurring at rest and waking him from sleep, is probably due to <u>paroxysmal SVT</u>, a reentrant arrythmia in a dilated LA. This is probably more likely than sinus tachycardia given the history of its <u>discrete onset and termination</u> - the most valuable clinical clue in*

differentiating an arrhythmia from sinus tachycardia. AI is a common cause of palpitations due to the increased LV volume and, without the recent history of rest episodes, would be the logical etiology of the palpitations otherwise.

- *"Chest pain" probably refers to the disagreeable sensation of palpitations and the pounding of the dilated LV against the chest wall. With long diastoles in AI, diastolic perfusion pressure can drop too low for subendocardial perfusion resulting in ischemia (often when sleeping, when the heart slows down), but that's not suggested by the description and pain setting in this case. In Uganda, chest pain is often the translation for vague discomforts in the chest.*

5. What is the therapy available to him in Uganda?

- *He is clearly in need of valve replacement surgery given his symptoms and signs of increased LVEDP and cardiac failure. Medical management is very difficult. Beta-blocker therapy may help palpitations due to SVTs, but it will increase AI (regurgitation increases with longer diastoles); ACE-inhibitors or hydralazine/nitrate afterload reduction would seem to make physiologic sense with 2 regurgitant lesions, but they unfortunately don't make a significant difference in symptoms or survival in MR, and are of little benefit in AI.*

Suggested Reading:

McGee, S. Evidence Based Physical Diagnosis (2nd Edition, 2007); Saunders Elsevier
Enriquez-Sarano, M., et. al. Mitral regurgitation Lancet 2009; 373: 1382–94
Lembo, N.J., et.al Bedside Diagnosis of Systolic Murmurs NEJM 1988; 318:1572-8
McGee, S. Etiology and Diagnosis of Systolic Murmurs in Adults American Journal of Medicine (2010) 123, 913-921
Bonow RO, Lakatos E, Maron BJ, et al. Serial long-term assessment of the natural history of asymptomatic patients with chronic aortic regurgitation and normal left ventricular systolic function. *Circulation* 1991;84:1625-1635.
Siemienczuk D, Greenberg B, Morris C, et al. Chronic aortic insufficiency: factors associated with progression to AVR. *Ann Intern Med* 1989;110:587-592.
Rapaport E. Natural history of aortic and mitral valve disease. *Am J Cardiol* 1975;35:221-227.
McKay CR, Rahimtoola SH. Natural history of aortic regurgitation. In: Gaasch WH, Levine HJ, eds. *Chronic Aortic Regurgitation*. Boston: Kluwer Academic, 1980:1-17.