**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 31 – Chest Pain for 14 Years

A 40 year old woman presents to Kisoro District Hospital after months of progressive abdominal pain, and abdominal and leg swelling.

She first began experiencing chest pain 14 years ago after birth of the last of her 3 children. When asked to describe the "pain", she fluttered her hand and recounted transient episodes of "heart racing" lasting minutes to hours, occurring weekly to monthly. Intermittent bouts of heart racing were her main symptom until ~ 3-4 years ago when she noticed gradually progressive dyspnea on exertion and decreased energy, and, over the past year, shortness of breath at night intermittently causing her to sleep with pillows.

Three months ago, fatigue and weakness increased, accompanied now by diffuse, constant abdominal pain, and abdominal and bilateral leg swelling. There was no change with food. She recently began to cough producing scant clear-yellow sputum. She has not felt "hot". She recalls no prolonged period of pain in her joints causing functional limitation when younger.

**Physical Exam:** Appearing tired and thin, in no acute distress

RR 24, T: 38, BP 110/70; HR regular ~110; occasional premature contractions (PCs)

Mouth: poor dentition, no thrush

Neck: JVP to angle of the jaw; regular cannon V waves

+ hepato-jugular reflux (HJR)

no lymphadenopathy, no thyromegaly

Lungs: crackles, intermittent wheezes ↑ ½ lung fields bilaterally

Heart: Right parasternal lift;

PMI/heave: 5 cm diameter, in 2 interspaces, anterior to mid-axillary line

Right parasternal/left PMI/heave are dyssynchronous with "rocking" motion to chest wall

$S_1$, ↓ $S_2$;

⊕ $S_3$ in left lateral decubitus position at apex, ↑'d with expiration;

⊕ $S_3$, when supine, at left lateral sternal border, ↑'d with inspiration

Gr 3/6 low-medium pitched, holosytolic murmur to axilla/back

- Post- PC compensatory pause – no significant change in intensity of murmur

Gr 2/6 holosystolic murmur, LLSB to 3rd rib, ↑'d with inspiration

Abdomen: pulsatile liver; tender to percussion in RUQ

distended, ⊕ shifting dullness

Extremities: edema: 3mm to knees bilaterally

Musculo-skeletal and Neuro exams unremarkable

1. 1. The complaint of "chest pain" can carry different meanings in Africa than in the U.S. What other symptoms might chest pain encompass in rural Africa? Why?

How should the health provider get at the root of the complaint?

- *Commonly in Africa, and sometimes in the West, particularly when histories are translated, "pain" is the word used to describe various types of discomfort in the chest including palpitations and labored breathing. All feel bad, arouse concern, threaten life and the heart at its very core.*
  *In this case, "pain" indeed referred to palpitations – the uncomfortable, conscious sensation of the heart beating.*

- *The complaint of "chest pain" always requires further inquiry with <u>open-ended</u> questions. E.g. "Tell me more, what do you mean by 'chest pain'?"…. etc.*

2. What is the *pathophysiologic significance* of the time course or natural history of the patient's symptoms?

- *The chronicity of 14 years makes any pain complaint less likely to be immediately dangerous or, in the case of chest pain, to reflect ischemia.*
  *If the "pain" signifies "palpitations" as it turned out to be in this case, it's less likely to be from worrisome arrhythmias like ventricular tachycardia, but rather from bouts of supraventricular tachycardia (SVT) or paroxysmal atrial fibrillation (PAF) - self-limited, probably due to primary conduction abnormalities, atrial enlargement or atrial scarring.*

- *Although she dates her problem to her "last child", many complaints are linked by patient memory or causal reasoning to important life events, and the two are often NOT biologically linked.*
  *In this case, with only the stable, very long palpitation history - without other symptoms of cardiac disease - post-partum cardiomyopathy would be a very unlikely explanation for her symptoms. However, during pregnancy the increase in blood volume may have exacerbated underlying atrial distention and facilitated reentrant (e.g. SVT/PAF) arrhythmias in predisposed atria.*

- *Just 3-4 years ago progressive dyspnea on exertion and orthopnea, symptoms of left ventricular failure, developed. With primary "volume loads" on the myocardium characteristic of chronic mitral regurgitation (MR) or aortic insufficiency (AI), it usually takes many years before symptoms of left ventricular failure due to an increased left-ventricular end diastolic pressure (LVEDP) develop – even with severe lesions.*

- *Three months ago, signs of right ventricular failure developed.*
  *RV failure, due to chronically increased pulmonary venous and then pulmonary arterial pressures, often follows LV failure by years.*

3. In patients who complain of chronic SOB and/or edema, what is a common but often overlooked cause of *abdominal pain*? How is it diagnosed?

- *Hepatic congestion due to RV failure distends the pain-sensitive hepatic capsule, and causes diffuse abdominal pain, often accompanied by chronic nausea and anorexia. Frequently diffuse and poorly-localized by history, the right-upper quadrant is tender on exam.*

  *Probably the most specific exam technique to use is percussion: first, of the left upper quadrant (as a control) and then of the right-upper quadrant (RUQ). Observe for wincing elicited over the RUQ with at first gentle and then heavier percussion notes (moving from more specific/less sensitive to more sensitive/less specific maneuvers) and ending with even less specific "punch-tenderness": the examiner hitting the back of his/her own hand over the left and then the right upper quadrants.*

4. What are the key findings on the PE and their pathophysiologic significance?

- *Vital Signs: RR 24, HR 110: increased respiratory and heart rates are seen in both decompensated CHF, via catecholamines, or an underlying pneumonia, which itself can cause cardiac decompensation;*

  *$T^0$ 38: a low grade fever, is also consistent with CHF or infection;*

- *JVP to the angle of the jaw: increased right-sided pressures, likely RV heart failure*

- *regular cannon V waves in the neck: Tricuspid regurgitation (TR) (TR is often better seen than heard i.e. neck pulsations may be more evident than the associated low-pitched murmur)*

- *lungs: crackles/wheezes: probably mean LV failure in this context: acute transudation of capillary fluid into the alveoli overwhelms the capacity of the pulmonary lymphatics to "drain" the lungs. Crackles follow.*
  *Of course, crackles are also consistent with pneumonia, more so if unilateral.*

  *LVF can induce reflex bronchospasm, which causes wheezing.*

- *PMI, LV heave, MCL-AAL, 5 cm diameter: the normal PMI is < 2 cm; A PMI > 3 cm is associated with LV dilatation, and if also "heaving" (i.e. forceful, suggesting*

*relatively preserved systolic function) is likely to be due to primary LV volume overload due to valve incompetence - as in MR or AI.*

- *Dyssynchronous RV lift/LV heave: the LA lies behind the RV, and an RV lift occurring late in systole after an LV heave is a sign of severe MR: the MR causes the LA (filling with blood from the LV) to push the RV forward against the precordium. Thus in "primary MR" due to valve pathology – i.e. not ("secondary") mitral annular dilatation from systolic dysfunction - the precordium seems to rock back and forth dyssynchronously: LV heave – RV lift.*
  *When the RV and LV are fairly synchronous, biventricular (usually dilated) cardiomyopathy involving both ventricles is usually responsible.*

- *medium-low pitch of the holosystolic murmur, radiating to the back: signifies more severe MR. Pitch is related to the pressure gradient between chambers. Most MR is high-pitched (blood flowing backwards from the high pressure LV to the low pressure LA producing a high gradient). However, when MR (holosystolic, radiating to the back) is lower pitched, more severe pathology is suggested – i.e. a lower gradient implies either a high LA pressure from a wider incompetent mitral orifice and more flow, and/or a ventricle failing to contract. Thus, while most MR is high pitched, lower pitch signifies increased severity of the regurgitation.*

- *post-PC compensatory pause, no change in the murmur's intensity: No change in intensity post-compensatory pause favors MR over AS.*
  *In both lesions, the longer RR interval leads to more volume in both the LV and the LA, and lower aortic diastolic pressure given more time for arterial run-off peripherally. But in MR, the lower aortic and higher atrial pressures accompanying the long diastole leads to proportionately more blood being ejected forward into the aorta rather than backward into the full (more resistant) left atrium. Thus, although in MR there is indeed more blood overall ejected from the left ventricle after a longer diastole, the backward flow remains relatively constant and there's no audible change in murmur intensity.*

  *In AS however, there is only one way out and, with longer time to fill the ventricle, more blood is ejected through the diseased AV increasing the intensity of the murmur.*

- *S3's: In this patient there are two distinct S3s - one from the LV at the apex increasing with expiration, the other from the RV at the lower left sternal border (LLSB), increasing with inspiration. The two S3's are signs of dilatation and*

*dysfunction of both ventricles. (N.B. in severe MR, an S3 can be heard with preserved systolic function.)*

- *↑with inspiration of 2/6 murmur, LLSB: an increase with inspiration is a sign of RV murmurs (TR). LV murmurs increase with expiration.*

- *pulsatile liver: indicates TR. A pulsatile liver can be appreciated visually in thin, supine patients by simple observation; it's best palpated by a hand gently placed over the RUQ with mild pressure applied to the abdomen.*

- *ascites, edema, tender liver: These are all signs of RV failure (due to ↑'d PA pressures, in turn due to severe MR for years). PA pressures increase with MR due to the LA pressure increase, reflex vasoconstriction to protect the pulmonary capillary bed from transudation and edema, and pulmonary arteriolar hypertrophy/sclerosis over time. Often the initial LV CHF symptoms of PND and orthopnea decrease as the RV fails and thereby ejects less blood into the congested lungs, although fatigue and symptoms of decreased cardiac output increase.*

**5. What is the most likely *specific etiology* of this patient's underlying disease?**

*<u>Chronic mitral regurgitation due to rheumatic heart disease.</u>*

*The chronicity of symptoms - starting with episodes of palpitations for many years, gradual left heart failure for a few years, and recent right ventricular failure - is consistent with the progression of chronic MR as discussed above, and the physical exam is classic for chronic MR.*

*Rheumatic heart disease is a major cause of heart failure in Africa and the rest of the developing world, although it's rare now in developed nations. Caused by a genetically-linked autoimmune cross-reaction between group A streptococcal antigens and the human heart, acute rheumatic fever (ARF) is thought to complicate 3% of strep pharyngitis in epidemics, but far less frequently when sporadic or "endemic". Its decrease in the developed world is attributed to improved housing and less overcrowding (less transmission), decreased rheumatogenicity of group A strep strains, and prompt antibiotic treatment of strep pharyngitis. Of note, most cases of ARF are complications of*

*asymptomatic strep infections, and, although the heart is involved in at least 50% of patients with symptomatic ARF, most patients with later complications of rheumatic heart disease can't recall an illness consistent with ARF.*

*Recent echocardiographic, population-based studies in Africa reveal an alarming prevalence of RHD of 2-3% of school children, ten times the prevalence of RHD that was previously detected in asymptomatic children by physical exam (NEJM 357:470, '07).*

**6. What potentially reversible cause of clinical decompensation in this disease "can't be missed"?**

- *Infection, particularly bacterial pneumonia taking root in the "wet" lungs of CHF, can't be missed.*
  *Infection is both a common complication and a precipitant of CHF.*

  *In Uganda, besides acute pyogenic pneumonia, TB is a very prevalent "community-acquired pneumonia" and can insidiously worsen CHF and cause diuretic resistance. Thus monitor temperature, check WBC and perhaps ESR (which is often low in decompensated CHF, so if the ESR is elevated, it suggests infection). Also, consider a chest x-ray if available.*

- *Other common precipitants of CHF in Africa include arrhythmia (atrial fibrillation) and anemia. Ischemia is rare, and hyperthyroidism, prevalent and untreated in rural Africa, less likely simply because the others are so common.*

- *For this patient with end-stage MR, because of the low grade fever, scant sputum, and crackles, antibiotics were given – without effect – and sputum was checked for TB, but was negative. The ESR was low (N.B. as mentioned, low ESRs can sometimes be seen in recent right ventricular failure). A CXR showed a massively dilated heart, but no discrete infiltrate.*

**7. How can this disease be prevented?**

- *Patients who've had rheumatic carditis once, have a 50% chance of having recurrent carditis after another group A strep pharyngitis even if mild, and it's thought that most*

*symptomatic chronic RHD results from repeated (usually asymptomatic) bouts of post-strep rheumatic carditis causing progressive scarring of the valve. Thus key to prevention of chronic heart disease, is chronic prophylaxis against group A strep pharyngitis.*

- *In patients with valvular disease due to rheumatic fever, prophylaxis is recommended for at least 10 years after the last episode and at least until age 40, whichever is longer. In areas of high epidemic potential like rural Africa, some advocate lifelong prophylaxis. Prophylaxis is ideally with benzathine penicillin G, 1.2 M units monthly IM (or if allergic, daily sulfadiazine 0.5-1.0 daily for patients, depending on weight (< or > than 60 Kg).*

- *Primary prevention, i.e. treatment of group A strep pharyngitis, effectively prevents ARF if administered within 1-2 weeks of the onset of symptomatic infection. Treatment is with Benzathine penicillin G, 600,000 units IM (or erythromycin for 10 days if allergic).*

**Suggested Readings:**


Silwa, K., et al. Epidemiology and Etiology of Cardiomyopathy in Africa *Circulation.* 2005;112:3577-3583

Essop, M.R., et. al, Rheumatic and Nonrheumatic Valvular Heart Disease: Epidemiology, Management, and Prevention in Africa Circulation. 2005;112:3584-3591

Marijon, E., et. al Prevalence of Rheumatic Heart Disease Detected by Echocardiographic Screening N Engl J Med 2007;357:470-6.

Carapetis, J.P. Rheumatic Heart Disease in Developing Countries NEJM 2007, 357;5

Constant, J. *Bedside Cardiology* 4$^{TH}$ Ed. Little, Brown and Co. 1993

Enriquez-Sarano, M., et.al; Mitral Regurgitation Lancet 2009; 373: 1382–94

Marcus RH, et.al. The spectrum of severe rheumatic mitral valve disease in a developing country. Correlations among clinical presentation, surgical pathologic findings, and hemodynamic sequelae. Ann Intern Med 1994; 120:177.

Chockalingam A, Clinical spectrum of chronic rheumatic heart disease in India. J Heart Valve Dis 2003; 12:577.