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PICTURE QUIZ

An unfortunate teenager

A 19 year old man presented to the emergency department with sudden onset shortness of breath and sharp left sided chest pain that was worse on inspiration. He had no history of trauma and had been otherwise well before the onset of these symptoms. However, a year ago he had twice presented to hospital with similar symptoms. He had also undergone surgery on one of his heart valves two years previously but was unclear about the details. The only drug he was taking was warfarin for long term anticoagulation.

The patient was tall, thin, and visibly short of breath but was able to complete full sentences. Closer examination showed a respiratory rate of 22 breaths/min, a non-deviated trachea, and reduced air entry over the left hemithorax with a resonant percussion note throughout. Precordial auscultation showed a soft systolic murmur and mechanical S2, which was loudest over the cardiac apex. The jugular venous pulse was not visibly raised, the patient was normotensive, and pulse oximetry measured his oxygen saturation at 95% on room air.

Twelve lead electrocardiography showed normal sinus rhythm with no other abnormalities. Full blood count, renal function, electrolytes, and clotting were all normal, with an international normalised ratio of 1.1 reflecting poor compliance with warfarin. Chest radiography was performed (fig).

1 What abnormalities are visible on the chest radiograph?
2 What unifying diagnosis explains all these abnormalities?
3 What criteria are used to establish a definitive diagnosis?
4 What are the common clinical manifestations of this syndrome?
5 What is the genetic basis of this disease?

Submitted by Zishan Sheikh, Shahnawaz Khan, and Afroze Khan
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STATISTICAL QUESTION

"n of 1" trials

Researchers investigated whether paracetamol was as effective as non-steroidal anti-inflammatory drugs (NSAIDs) in the treatment of pain and disability related to osteoarthritis of the hip or knee. A series of double blind, randomised "n of 1" controlled trials was performed. Each drug was taken for two weeks, administered for a maximum of five cycles. Thirteen patients in primary care who had regularly been using NSAIDS were selected. Patients received the same type of NSAID and in the same dosage, if possible, as they were taking before the start of the study. Outcome measures included severity of complaints of pain, stiffness, and limitations in daily functioning, together with satisfaction with drugs and side effects.

Seven patients completed the study, and it was recommended that six of these change to paracetamol. All the other patients continued with NSAIDs. Three months after the study finished, of the six patients for whom a change to paracetamol was recommended, four were taking NSAIDs for practical reasons or because of a perceived lack of efficacy of paracetamol. The authors concluded that n of 1 trials can be used to investigate the best treatment for patients with osteoarthritis of the hip or knee.

Which of the following statements, if any, are true?

a) Patients received an NSAID and paracetamol.
b) Each patient acted as his or her own control.
c) The optimum treatment was determined for each patient.
d) An n of 1 trial is a between-subjects design.

Submitted by Philip Sedgwick
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CASE REPORT **Exertional dyspnoea and syncope**

A 64 year old man presented with a two year history of exertional dyspnoea, which improved with rest. He reported laboured breathing after walking for about 10 minutes on the flat or after climbing one flight of stairs. He had no chest pain or other symptoms. His medical history included hypertension and pulmonary embolism. He was a retired mechanical engineer with no family history of cardiac disease and was a non-smoker.

On examination, his pulse was 60 beats/min and blood pressure 130/80 mm Hg. His jugular venous pressure was not raised, his heart sounds were normal, and he had no peripheral oedema. Electrocardiography showed sinus rhythm, with low voltage complexes in the limb leads, poor R wave progression, and no evidence of left ventricular hypertrophy. Chest radiography showed clear lung fields and increased cardiothoracic ratio.

Transthoracic echocardiography with harmonic adjustment showed grossly thickened myocardium with a "speckled" appearance, impaired long axis left ventricular function with an ejection fraction of 53%, thickened mitral and tricuspid valve leaflets, and evidence of diastolic impairment. We performed computed tomography to investigate differential diagnoses of breathlessness, including coronary artery disease and chronic pulmonary embolism. Computed tomography showed unobstructive mild calcification of the left anterior descending artery and no evidence of pulmonary embolism.

The patient collapsed after being given metoprolol as part of the computed tomography imaging protocol. An electrocardiogram taken after initial resuscitation showed atrial flutter with 3:1 block. Cardiac magnetic resonance imaging confirmed myocardial thickening seen on previous imaging and showed circumferential transmural delayed enhancement of the left ventricle and to a lesser extent the right ventricle.

1 What is the most likely underlying diagnosis?
2 Why did this patient collapse?
3 What is the initial treatment for atrial flutter?
4 How would you further manage this patient?

Submitted by Jason M Tarkin, Andrew Kelion, Tarun Mittal, and Miles C Dalby
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