

## P H O T O   Q U I Z

# Radiographic conundrum: what is the cause of enlarged heart in this patient with Down's syndrome?

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## Case history

A 19 year-old man with Down's syndrome was suffering from high fever and cough after a minor throat infection. His relevant past history mentioned a small, asymptomatic ventricular septal defect (VSD). Antibiotics (amoxicillin-clavulanic acid) were prescribed at an early stage. After a few days, doxycycline was added because his temperature remained above 39 °C. Ten days after onset of this illness he was transferred to our hospital, having developed fatigue, persistent cough and progressive shortness of breath. On admission, physical examination revealed tachycardia (135 bpm),

he was afebrile (37.4 °C), tachypneic, and had a normal blood pressure. Furthermore, a raised jugular venous pressure and a grade 5/6 rough systolic murmur at the left sternal border (4<sup>th</sup> intercostal space) were found. No diastolic murmur was heard. There were no peripheral stigmata of infective endocarditis or other relevant findings. A chest X-ray was taken (figure 1).

## What is your differential diagnosis and what would be your next step?

Answer on the next page.



Figure 1. Chest X-ray on admission

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## Answer to photo quiz (on page 151)

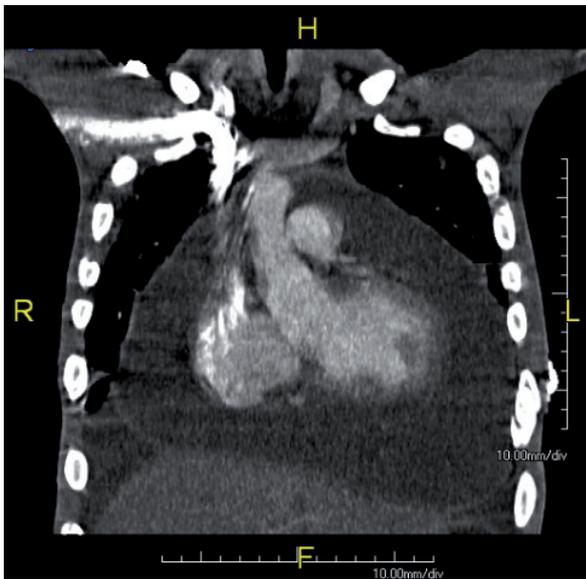


Figure 2. An AP view, reconstructed from the raw data of the chest CT-scan, to give a similar view to the chest X-ray

Chest radiography shows overt cardiomegaly with a cardiothoracic ratio of more than 60%. The incidence of congenital cardiac anomalies in Down's syndrome is up to 50 percent and the most commonly found congenital heart defects are endocardial cushion defect, VSD, secundum atrial septal defect, tetralogy of Fallot, and isolated patent ductus arteriosus [1]. About 30% of patients have multiple cardiac defects. Our patient's previous medical history included a small, asymptomatic VSD. Recently Concolino et al have shown that mild to moderate pericardial effusion is found in almost 30% of Down's syndrome patients [2]. Some of them have hypothyroidism. In our patient Thyroid-Stimulating Hormone (TSH) measurement excluded this diagnosis.

Our next step was a two-dimensional echocardiographic examination. Echocardiographic examination demonstrated significant mitral incompetence, with vegetative growths on the valvular cusps, and 3 cm of pericardial exudate surrounding the heart; the aortic valve appeared normal, other valves were not visualized. Pericardiocentesis produced 1500 ml of frank pus from the pericardial sac that was sent for culture. Gram stain of the pericardial fluid revealed polymorph nuclear cells and Gram-positive cocci. Antibiotic treatment was promptly changed and vancomycin intravenously was added to the regimen. Doxycycline was discontinued. Cultures of the

pericardial fluid showed heavy growth of *Staphylococcus aureus*. Based on antibiotic susceptibility tests vancomycin was discontinued and flucloxacillin begun (12 g continuously/24 hours) and gentamycin 4 mg/kg per day, amoxicillin-clavulanic acid was stopped. Unfortunately, a few days later he exhibited early signs of cardiac tamponade, a pulsus paradoxus, for which he underwent pericardiectomy with drainage of a large, circumferential pericardial effusion. In preparation for surgical intervention a CT-scan of the thorax was performed (Figure 2).

### Conclusion

The chest X-ray did not in fact show a greatly enlarged heart but a massive volume of pericardial effusion. This case of purulent pericarditis and bacterial endocarditis was caused by *Staphylococcus aureus* which is reported to be the most common causative agent. Other causative agents include *Haemophilus influenzae* type b and *Streptococcus pneumoniae*. Purulent pericarditis starts with infection elsewhere in the body which is usually transmitted to the pericardium by direct bacterial dissemination or via the bloodstream [3]. Emergency pericardial drainage in combination with intravenous antibiotics is mandatory in such cases. The postoperative course of our patient was uneventful.

### References:

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2. Concolino D, Pascuzzi A, Pietragalla E, Lia R, Canepa S, Strisciuglio P. High prevalence of isolated pericardial effusion in Down syndrome. *Am J Med Genet A*. 2005;132:331-2.
3. Seferović PM, Ristić AD, Imazio M, Maksimović R, Simeunović D, Trincherro R, Pankuweit S, Maisch B. Management Strategies in Pericardial Emergencies. *Herz* 2006;31:891-900