Case 1: A 31-Year-Old Man with Fever and Rapidly Progressive Dyspnea

A 31-year-old man with a history of ankylosing spondylitis is brought to the Emergency Department by a friend. He reports one week of low-grade fevers (100.5 °F), cough, progressive shortness of breath, nausea, diarrhea, and occasional vomiting. He has noticed a rapid decline in his exercise tolerance in the past week and states he can't even walk more than 10–20 ft without getting very short of breath. The patient also complains of a frothy cough sometimes laced with blood, paroxysmal nocturnal dyspnea, and orthopnea. He denies any chest pain.

On physical examination, blood pressure is 110/40 mmHg, pulse rate is 110 bpm, and oxygen saturation is 98%. His lungs are clear and he has a diastolic murmur along the right upper sternal border. After an echocardiogram, he is referred for cardiac catheterization. What is the diagnosis based on the aortogram below (Figure 1.1)?

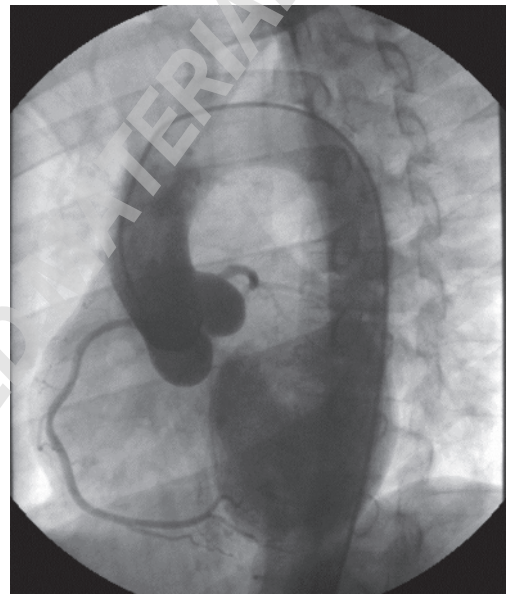

Figure 1.1 Aortogram performed in an LAO (left anterior oblique) projection. The pigtail catheter is located right above the aortic valve during injection of contrast dye.

Aortic Regurgitation

The pigtail catheter is in the aortic root and a power injection of a high volume of contrast is performed. This results in filling of the left ventricle, consistent with aortic regurgitation (AR). Angiographic classification of severity of AR is based on the amount of contrast dye entering the left ventricle during an aortogram:

- 1+ some dye enters ventricle but clears with every systole
- 2+ the ventricle becomes completely opacified after several heartbeats and remains opacified throughout the cardiac cycle
- 3+ after several heartbeats – the ventricle becomes as dark as the aorta
- 4+ after several heartbeats – the ventricle becomes darker than the aorta

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Note that in the 2014 AHA/ACC Valvular Heart Disease Guideline, transthoracic echocardiography is the first line diagnostic modality for AR as it provides information on the cause of regurgitation, regurgitant severity, and LV (left ventricular) size and systolic function. Cardiac magnetic resonance imaging is indicated in patients with AR in whom echocardiographic images are suboptimal. Cardiac catheterization was performed in this patient to evaluate coronary anatomy prior to surgery.

AR is a condition of increased afterload with associated hemodynamic changes varying depending on the time–course of the valve dysfunction. If AR develops rapidly (i.e. acute or subacute AR), the left ventricle is unable to handle the pressure and volume overload causing a rapid increase in left ventricular pressures during diastole, markedly elevated pressures at end diastole, and premature closure of the mitral valve. Systemic diastolic pressures may be low but generally there is a minimal increase in pulse pressure; in very severe cases of acute AR, cardiac output may fall leading to a decrease in pulse pressure and/or hypotension. In chronic AR, stroke volume increases to maintain effective forward flow. This leads to dilation of the left ventricle, leading in some patients to the development of a massively dilated left ventricle, termed *cor bovinum*.

In this patient, an echocardiogram showed severely decreased left ventricular contraction with left atrial dilation. The aortic valve was trileaflet with inadequate coaptation and severe AR (Table 1.1). Right ventricular contractile performance was decreased. Hemodynamic changes of severe AR were found at cardiac catheterization including elevated left ventricular end diastolic pressure (38 mmHg), equalization of aortic and left ventricular pressures during diastole (Figure 1.2), and premature closure of the mitral valve during diastole. There was moderate pulmonary hypertension (50/33 mmHg) and reduced cardiac output (2.3 l/min by assumed Fick Method). The patient underwent successful aortic valve replacement with a 25 mm St. Jude prosthesis.

Ankylosing spondylitis is an autoimmune spondylarthropathy that is characterized by chronic, painful, degenerative inflammatory arthritis primarily affecting the lower spine and sacroiliac joints. There is a strong association with the class I major histocompatibility complex antigen HLA-B27. In patients with ankylosing spondylitis the incidence of AR is approximately 10%.

The 4 stages of AR according to the 2014 AHA/ACC Valvular Heart Disease Guideline are: Stage A: No current AR but at risk of developing AR (including bicuspid aortic valve, aortic valve sclerosis, diseases of the aortic sinuses or ascending aorta, history of

Table 1.1 Echo criteria for grading severity of aortic regurgitation (AR).

Echo criteria	Mild AR	Moderate AR	Severe AR
Doppler jet width	≤25% of LVOT	25–64% of LVOT	≥65% of LVOT
Vena Contracta	<0.3 cm	0.3–0.6 cm	>0.6 cm
Regurgitant volume	≤30 ml/beat	30–59 ml/beat	≥60 ml/beat
Regurgitant fraction	≤30%	30–49%	≥50%
Effective regurgitant orifice (ERO)	<0.1 cm^2	0.1–0.29 cm^2	≥0.3 cm^2
Miscellaneous			Holodiastolic flow reversal in the proximal abdominal aorta

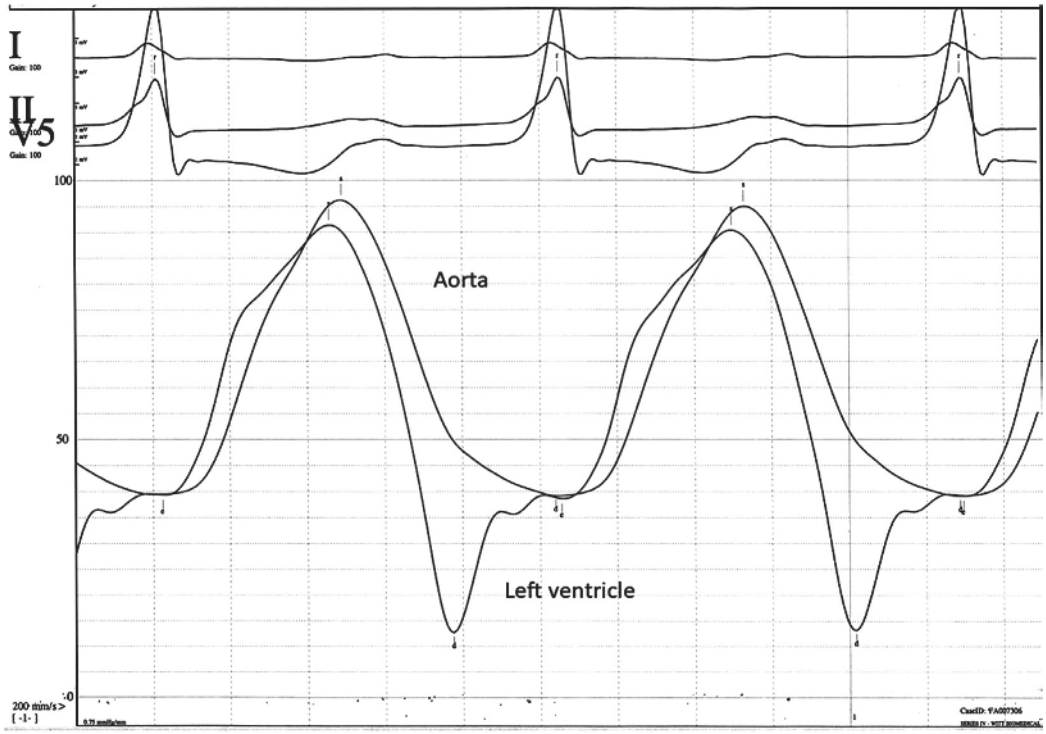

Figure 1.2 Simultaneous recording of pressures in the aortic root and left ventricle.

rheumatic fever or known rheumatic heart disease, infective endocarditis); Stage B: progressive mild-to-moderate AR; Stage C: severe asymptomatic; and Stage D: symptomatic AR.

Class 1 indications for aortic valve replacement in AR include: (i) Symptomatic patients with severe AR regardless of systolic function; (ii) Asymptomatic patients with chronic severe AR and LV systolic dysfunction (LVEF 50 mm or indexed LV end-systolic dimension [LVESD] >25 mm/m^2); and (iii) Patients undergoing cardiac surgery for other indications with severe AR (stage C or D). Class 2a indications include: (i) Asymptomatic patients with severe AR with normal LV systolic function (LVEF ≥50%) but with severe LV dilation (LVESD >50 mm or indexed LVESD >25 mm/m^2) and (ii) moderate AR (stage B) while undergoing surgery on the ascending aorta, coronary artery bypass grafting (CABG), or mitral valve surgery. Class 2b indication includes Asymptomatic patients with severe AR, normal LV systolic function at rest (LVEF ≥50%, stage C1), but with progressive severe LV dilatation (LV end-diastolic dimension >65 mm) if surgical risk is low.

Further Reading

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