The heart in ankylosing spondylitis

Heart disease is a well recognised complication of ankylosing spondylitis (AS). The most characteristic lesions are aortic incompetence and conduction defects. The spectrum of disease is wide and includes mitral valve disease,1 cardiomyopathy,2 and pericarditis.3 First described by Mallory in 1936,4 Bauer drew attention to the association of aortitis with spondylitis in 1951.5 The presence of aortic valve disease in a subgroup of patients with rheumatoid spondylitis was one feature which helped distinguish rheumatoid arthritis and AS as separate diseases.6

Prevalence of aortic incompetence increases with age, disease duration, and presence of peripheral arthritis.7 Graham found a prevalence of 10-1% in patients who had had spondylitis for 30 years, compared with 1% in those with five years of disease.7 Prevalence was approximately doubled in a subgroup of patients with peripheral joint disease (other than hip and shoulder involvement). Similarly, prevalence of atrioventricular block increased with duration of disease from 0-6% after five years of disease to 8-5% after 30 years. As with aortic incompetence, prevalence almost doubled if peripheral joints were affected. There is an apparently increased prevalence of pleuritis in those with cardiovascular disease,7 though this may be due to the effect of disease duration, with which both are associated.8 Studies show no increased prevalence of HLA-B27 in patients with lone aortic incompetence in the absence of spondylitis.9 There is, however, some evidence of an increased prevalence of HLA-B27 in men with complete heart block.10

Studies show a slightly increased mortality in those with AS, and cardiovascular disorders have been implicated as one cause of this.11 12 Court Brown studied cause of death in patients with AS treated with radiotherapy during 1935–54, who were followed up until 1960.11 Relative risk of death due to chronic endocarditis, not specified as rheumatic, was 2-8 and other, non-cerebrovascular circulatory disease 1-3. Radford looked at mortality in 836 patients with AS not given deep x ray treatment, diagnosed during 1935–57, who were followed up until 1968.12 There were no deaths due to endocarditis, but the relative risk due to other, non-cerebrovascular circulatory disease was 1-4 in men. In a retrospective series of 519 patients with AS7 aortic incompetence was present in 24 and progressed in 50% over a seven year period. Progression was associated with development of symptoms, including dyspnoea and angina. The spectrum of disease is wide, however, and acute cardiac decompensation may occur over a short period of time.13

The pathological features of aortitis in AS were characterised by Bulkley in 1973.14 The proximal aortic wall behind and above the sinuses of valsalva is thickened primarily because of adventitial scarring and intimal proliferation. The vasa vasorum are surrounded by plasma cells and lymphocytes, and their lumens often narrowed. Aortitis may extend below the aortic root to the base of the mitral valve and into the intraventricular septum. The adventitial scar tissue below the aortic valve may result in a subaortic fibrous ridge. The inflammatory process usually extends no more than a few centimetres distally into the ascending aorta, though there are reports of more extensive involvement.15 Syphilitic aortitis has identical histological features, though does not extend below the aortic valve.

Aortic valve disease occurs in 1-10% of patients with AS.5 7 16 Differences in patient groups, such as age and duration of disease, may partly explain the varying prevalence between series. It is typically a feature of long-standing disease in adults, though has been reported in young children.17 Graham found lone aortic incompetence present in 21 of 519 patients with spondylitis compared with three of 508 controls with RA.7 Cardiac conduction disturbances are reported in 1-33%.17 18 19 As with aortic valve disease, variations in prevalence may be due to differences in patient groups. As conduction disturbances may be intermittent, however, the frequency of electrocardiographic investigations and duration of follow up may also be important. Prevalence is higher in those with aortic valve disease.20 A wide variety of conduction disturbances have been described, including first, second, and third degree atrioventricular block, bundle branch block, fascicular block, and Wolff-Parkinson-White syndrome.18 20 21 Conduction disturbances may resolve spontaneously even in cases of complete heart block.18 Electrophysiological studies in patients with complete heart block suggest that the block is preferentially located in the atrioventricular node, though the conduction system may be widely affected.22 Mitral regurgitation is uncommon, though may be severe.1 The prevalence of mitral valve prolapse does not seem greater than that found in the general population.23 24

Over the past 10 years there has been increased recognition of myocardial disease in AS.24 25 Ribeiro found five of 28 patients with AS had a dilated and poorly contracting left ventricle.24 Brewerton demonstrated early diastolic abnormalities of the left ventricle on echocardiography in 16 of 30 male patients with AS who had no cardiorespiratory symptoms or known cardiac abnormalities.25 The findings were consistent with the presence of excess connective tissue in the myocardium. Computerised image analysis of myocardial tissue obtained at necropsy in 28 patients showed 30-7% interstitial reticulin compared with 17-7% in controls matched for age and sex (p<0-0001). Unlike rheumatoid

In patients with angina, the presence of a brachial orifice may be attributed to progression of previously diagnosed cardiac disease, particularly in severe aortic valve disease.25 In patients with heart disease exercises should be continued to maintain posture and spinal mobility. All patients with valvular lesions should be advised about prophylaxis against subacute bacterial endocarditis. When considering surgical replacement of an aortic valve, preoperative assessment and consultation with anaesthetic colleagues should be made because of the likely associated musculoskeletal, cardiovascular, and pulmonary problems.

Department of Rheumatology, St Vincent's Hospital, Elm Park, Dublin 4, Ireland

Correspondence to: Dr O'Neill.
The heart in ankylosing spondylitis.

T W O'Neill and B Bresnihan

doi: 10.1136/ard.51.6.705

Updated information and services can be found at:
http://ard.bmj.com/content/51/6/705.citation

**Email alerting service**
Receive free email alerts when new articles cite this article. Sign up in the box at the top right corner of the online article.

**Notes**

To request permissions go to:
http://group.bmj.com/group/rights-licensing/permissions

To order reprints go to:
http://journals.bmj.com/cgi/reprintform

To subscribe to BMJ go to:
http://group.bmj.com/subscribe/