Aortic valve disease

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AORTIC STENOSIS

Clinical suspicion of significant aortic stenosis even in the presence of a normal chest X-ray and electrocardiogram should lead to careful 2-D echocardiographic examination. The lesion can be adjudged mild and the subject probably certificated if the recording shows normal septal and posterior wall thickness, normal left ventricular dimensions and fractional shortening and a valve opening > 15 mm. In cases of doubt or where technically satisfactory echocardiograms are unobtainable, cardiac catheterization is likely to be mandatory. For unrestricted certification the peak systolic gradient across the valve should be < 20 mmHg and the LV end-diastolic pressure < 12 mmHg at rest, and both must remain within these limits on exercise to three times resting oxygen consumption.

AORTIC REGURGITATION

Any abnormality of electrocardiogram at rest or on exercise, or of the chest X-ray, any increase in left ventricular dimensions on echocardiography, a diastolic pressure below 65 mmHg and/or pulse pressure > 55 mmHg is likely to lead to cardiac catheterization. For full certification any angiographic regurgitation should be trivial.

Introduction

Valvular heart disease causes between 1 and 4% of sudden deaths [1, 2], and clinical experience shows that sudden incapacity is even more common. The main causes of incapacity in valve disease are (1) syncope or near-syncope and (2) cerebral embolism with neurological deficit leading to (1). The first risk outweighs the second in aortic valve disease, and syncope, near-syncope and sudden death are much more frequently caused by stenotic than regurgitant lesions [3]. Many mechanisms for syncope in aortic valve disease have been suggested. These include exercise-induced vasodilation and brady- or tachyarrhythmias combined with a nearly fixed and reduced cardiac output. Once a patient with aortic valve disease is symptomatic the risk of sudden death is extremely high [4, 5], and although reduced considerably by aortic valve replacement or valvotomy this risk still remains [6].

Because of the risk of sudden incapacitation, significant aortic valve disease precludes fitness to fly. Generally there is no difficulty in recognizing clinically the patient with severe symptomatic aortic valve disease, which is most frequently due to degenerative changes in a congenitally abnormal and often bicuspid valve. Rheumatic aortic valve disease is much less common and will often be combined with some evidence of mitral valve disease. It is generally accepted that significant aortic valve disease is unacceptable in aviators [7, 8] and therefore the definition of what comprises significant disease is important.

Aortic valve disease can be considered significant in two different respects.

1. AT INITIAL EXAMINATION

Even mild aortic disease can progress in the young adult over a period of a few years to severe disease [9, 10]. The chance of such progression is not remote, and 30% of patients with only an ejection sound and no murmur in one study developed murmurs and 5% required surgery during a ten-year follow-up period [10]. In view of these figures it would seem sensible to discourage persons with any evidence of aortic valve disease from starting on a career which could be cut short on medical grounds after a few years.

Bicuspid aortic valves

Approximately 2% of the male population have bicuspid aortic valves [11]. These may produce no clinical sign and even with echocardiography only
70% will be detected. Often the only physical sign is an aortic ejection sound. This may be misdiagnosed as a split first heart sound even by experienced cardiologists, and it is likely that however good the medical surveillance, many ejection sounds will go undetected. Follow-up studies of asymptomatic patients with aortic ejection sounds show that a significant proportion will develop important aortic stenosis or regurgitation or subacute bacterial endocarditis during a 10-year follow-up period. The high risk of aortic valve disease developing which is severe enough subsequently to disqualify from flying implies that the presence of a definite aortic ejection sound combined with echocardiographic evidence of bicuspid valve at initial assessment justifies discouraging a flying career. Discovery of an ejection sound on initial examination requires a high quality simultaneous echocardiogram and phonocardiogram to show that the onset of the ejection sound occurs at the exact moment that the aortic valve becomes fully open. The presence of a bicuspid valve may then be demonstrated as an eccentric aortic valve closure line on the M-mode echocardiogram, which identifies 70% of patients. A transverse section of the aortic valve with a 2-D echocardiogram will identify the number of aortic valve cusps in most subjects in whom a satisfactory recording can be obtained.

**Mild aortic valve disease and innocent flow murmurs**

Many young adults have soft murmurs of no prognostic or haemodynamic significance. It is important to differentiate these murmurs from mild aortic valve disease. The following features are important for a murmur to be diagnosed as 'innocent' and the following criteria should be satisfied.

(a) Soft murmur (2/4 or less in intensity) usually musical in quality; (b) no associated ejection sound; (c) no diastolic component; (d) normal electrocardiogram, chest X-ray and M-mode echocardiogram.

If any of these criteria is not satisfied, then further investigations to confirm that the aortic valve is normal, e.g., 2-D echocardiography or combined echo- and phonocardiography may be necessary. Non-invasive investigations, particularly the echocardiogram and clinical examination, are very accurate in showing even minor abnormalities of the aortic valve, although they often cannot define accurately minor haemodynamic effects. In approximately 20% of subjects satisfactory echocardiograms cannot be obtained for technical reasons and cardiac catheterization may be necessary to distinguish mild aortic stenosis from a flow murmur. If a definite early diastolic component is present, cardiac catheterization and aortography is unnecessary, even if the echocardiogram is unsatisfactory, as the aortic valve is the only likely source of such a murmur. Isolated early diastolic murmurs, in otherwise normal subjects, do not originate from the pulmonary valve.

2. **Suspicion of aortic valve disease detected in a licensed pilot**

From time to time, aircrew will have murmurs detected at routine medical examinations which suggest the possibility of aortic valve disease. Many will have congenitally bicuspid aortic valves.

**Evidence of a bicuspid valve**

There is no evidence that a bicuspid valve constitutes any risk beyond the subsequent development of significant aortic valve disease which should be detected by regular follow-up. Sudden severe aortic regurgitation due to prolapse of a non-infected bicuspid aortic valve has been reported but is exceedingly rare. If a bicuspid valve, producing only an ejection sound and no murmur is documented, this should be documented as outlined above and reassessed regularly by a cardiologist. Because of the risk of infective endocarditis, suitable antibiotic prophylaxis should be recommended for dental and/or urinary tract manipulation.

**Murmurs suggesting an aortic valve lesion**

The risk of incapacitating symptoms is related to the severity of the haemodynamic effects of obstruction and regurgitation. Although some authors have reported conduction abnormalities (prolonged H-V time) to be common and related to the severity of aortic stenosis there is no evidence that these changes have any clinical significance or that electrophysiological studies are necessary or helpful in patients with mild aortic disease. The level of obstruction or regurgitation which should be regarded as significant needs review. The invasive criteria quoted at the 8th Bethesda Conference in 1975 are consistent with what prognostic information is available and also with clinical and pathological experience. Aortic stenosis is judged to be sufficiently mild to permit a pilot to continue flying if the peak systolic gradient across the valve is < 20 mmHg and the left ventricular end-diastolic pressure is > 12 mmHg at rest and on exercise to three times resting oxygen consumption. Aortic regurgitation was not judged to constitute a risk if on cardiac catheterization the regurgitant fraction is < 25% of the left ventricular stroke volume. In addition there should be a mean pulmonary artery pressure of < 20 mmHg and a mean pulmonary wedge pressure of < 15 mmHg.
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Although these criteria are arbitrary they have served and are in line with clinical experience. No better data on the risks of sudden incapacity in mild aortic valve disease have appeared since the Bethesda conference and thus the fitness criteria dictated by cardiac catheterization in pilots or potential pilots with aortic valve disease are the same in 1982 as they were in 1975. The main advance during this time has been the development of and experience with non-invasive methods of diagnosis, of which echocardiography is the most important. If the severity of suspected aortic valve lesions can be assessed accurately by non-invasive means without recourse to cardiac catheterization this has obvious advantages. If the following clinical and non-invasive criteria are satisfied then cardiac catheterization should be unnecessary.

Aortic stenosis

**PHYSICAL EXAMINATION**

(a) There should be no fourth heart sound, no 'abnormality of the cardiac impulse (double character, palpable fourth heart sound), displacement beyond the mid-clavicular line or increased prominence or alteration in the character of the carotid pulse.

(b) The second heart sound should split normally. A single second sound or one with reversed splitting on respiration suggests an aortic lesion may be significant.

**THE ELECTROCARDIOGRAM, CHEST X-RAY AND ECHOCARDIOGRAM**

These should be normal. Both the chest X-ray and electrocardiogram can be normal in the presence of haemodynamically significant aortic stenosis\(^{[19, 30]}\). The echocardiogram is more sensitive to minor changes in left ventricular wall thickness and cavity size and is therefore of great importance. The following echocardiographic criteria should be satisfied.

(a) High quality recordings must be obtained. This is possible in about 80% of subjects.

(b) The left ventricular posterior wall thickness and the thickness of the interventricular septum must be normal. These features are best seen using M-mode echocardiograms. Most centres accept 12 mm as the upper limit of septal and posterior wall thickness measured at end-diastole.

(c) The left ventricular dimensions and fractional shortening must be normal.

(d) The aortic valve will probably be thickened even if no significant stenosis is present and may be obviously bicuspid. Any restriction of valve motion detected by either the M-mode or 2-D echocardiogram should be regarded with suspicion. The M-mode echocardiogram occasionally shows normal valve motion in patients with a mobile but stenotic valve. This problem can be resolved by the 2-D echocardiogram. This must however show the valve cusps clearly in both longitudinal and transverse sections and the echo beam must be scanned up, down and across the valve. If these precautions are taken then a valve that shows a 15 mm opening in all views can be regarded as non-stenotic\(^{[31, 31]}\). Two-dimensional echocardiography often overestimates the severity of aortic stenosis but does not underestimate it if these criteria are used.

**EXERCISE TESTING**

There are no systematic data on the value of exercise testing in the prediction of the severity and risk of aortic valve disease in the adult population. It has been used however in children, with some success, to assess the severity of aortic stenosis. It seems reasonable to demand a completely normal maximal exercise test (electrocardiogram, blood pressure and heart rate response and exercise tolerance) before accepting a non-invasive assessment that aortic stenosis is mild.

Aortic regurgitation

Although the risk of sudden incapacitation is lower in aortic regurgitation than stenosis it is nevertheless desirable that similar criteria for investigation should be applied. In addition the diastolic blood pressure is of importance. If this is \(< 65\) mmHg or the pulse pressure \(> 55\) mmHg, invasive assessment of severity is necessary\(^{[38]}\). Non-invasive criteria are also similar; a normal electrocardiogram, chest X-ray and echocardiogram should rule out any increase in left ventricular dimensions or associated valve stenosis shown by restricted valve motion.

A final caution is required in the case of aortic regurgitation. The development of a new murmur of aortic regurgitation should always raise the suspicion of an abnormality of the aortic root of which dilatation associated with a connective tissue disorder such as Marfan's syndrome, or a small asymptomatic aortic dissection are the most important. A subject with obvious Marfan's syndrome is an unlikely candidate for initial certification; however, a 'forme fruste' of the syndrome may occur and first come to light with the development of an aortic regurgitant murmur.
Therefore a new murmur of aortic regurgitation requires.

1. Clinical examination to exclude Marfan's syndrome usually in a 'forme fruste' (e.g. length of fingers, high arch palate, abnormality of the ocular lens).
3. An assessment of aortic root anatomy. Echocardiographic assessment with the 2-D system is good at showing aortic root anatomy whereas the M-mode echocardiogram is poor in this respect.
4. The mitral valve should be normal, since mitral valve prolapse is another common feature of Marfan's syndrome.

If these stringent non-invasive criteria are satisfied then cardiac catheterization is unnecessary. A single abnormality may require catheterization, and the criteria already described should be applied to decide whether the individual is fit to fly.

Associated mitral valve disease

If there is evidence of associated rheumatic mitral valve disease the risks are considerably increased, and such pilots should not be licensed to fly professionally. Even mild rheumatic mitral valve disease carries a significant risk of cerebral embolism.

Re-examination

Once mild aortic valve disease has been established, regular follow-up is required by a cardiologist, as the condition is progressive. The non-invasive investigations (i.e. electrocardiogram, chest X-ray, echocardiogram and exercise test) should be repeated at yearly or six-monthly intervals and any change should raise the possibility that invasive assessment is required.

Table 1  Recommendations relating to fitness to fly

<table>
<thead>
<tr>
<th>Condition</th>
<th>Type of Licence</th>
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<tbody>
<tr>
<td>Bicuspid aortic valve with no evidence of stenosis or regurgitation</td>
<td>Unrestricted</td>
</tr>
<tr>
<td>'Mild' aortic stenosis or regurgitation</td>
<td>Restricted to fly as or with co-pilot or safety pilot</td>
</tr>
<tr>
<td>'Significant' aortic valve disease</td>
<td>None</td>
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Recommendations relating to fitness to fly in aircrew with the abnormalities outlined above are summarized in Table 1.

References


