Coarctation of the Aorta

Case Report, by Rebecca Blue, MD, MPH

This case report reviews a case of coarctation of the aorta with surgical repair in a student pilot applicant. A comprehensive review is provided of this cardiovascular abnormality, surgical correction, associated pathology, and concerns for medical certification. This review illustrates that, while there are circumstances in which coarctation or associated disease may be disqualifying, successful management of this problem can provide the opportunity for successful medical certification.

History

A 21-year-old male presented to his aviation medical examiner for a first-time application for a third-class medical certificate to begin student pilot training. Review of the applicant's medical disclosures revealed that he had undergone surgical repair for coarctation of the aorta at age seven months. He was unable to provide a surgical report but reported primary end-to-end anastomosis. He also reported a history of a persistent left superior vena cava.

The applicant stated that he had been followed by pediatric cardiology throughout childhood without further problems, and he denied any history of hypertension, restenosis, or aortic aneurysm. He stated that he was generally well and exercised regularly in aerobic activity and high-intensity exercises, with no reported symptoms of chest pain, palpitations, or syncopal episodes.

On exam, he was found to have a well-healed surgical scar measuring 2 centimeters on the left lateral thorax consistent with the stated surgery. His heart rate was 68 beats per minute, with a blood pressure of 125/65. Pulses were equal in all extremities; the remainder of his exam was otherwise relatively unremarkable.

Angiography had been performed two years prior to the patient's presentation, with no evidence of residual coarctation. A recent echocardiogram demonstrated a normal valvular examination, a dilated coronary sinus consistent with a left superior vena cava, a descending aorta with a slightly decreased diameter distal to the subclavian artery, with peak velocity of 2.28 cm/sec and a peak gradient of 20.83 mmHg. Normal intracardiac anatomy and function was demonstrated, with an ejection fraction of >60%. A bubble study found no evidence of right-to-left shunting.

Aeromedical Issues

Patients with coarctation of the aorta may have significant issues regarding the sequelae of cardiac outflow obstruction.[3,7] including chronic and refractory hypertension that may be poorly controlled with medication. Ventricular hypertrophy and valvular disorders, particularly mitral and aortic stenosis, may occur as a result of chronic obstruction.[3]

While hypertension may be the presenting symptom for an airman with an undiagnosed coarctation, it is incumbent upon the evaluating physician to identify the etiology of the hypertension and any related sequelae, such as hypertrophy or valvular disorders, particularly in a younger applicant. Although waivers may be granted for hypertension and associated medication use, a missed diagnosis of coarctation may have such serious consequences as syncopal episodes, severe ventricular hypertrophy, heart failure, aortic dissection, infective endocarditis, premature coronary artery disease, and mortality from any of these etiologies.[3,4,7]

Outcome

This applicant had already undergone significant evaluation prior to his presentation to the aviation medical examiner (AME), including a current echocardiogram showing no significant sequelae and an angiogram demonstrating excellent blood flow with no evidence of restenosis. The AME appropriately deferred the patient to the FAA's Aerospace Medical Certification Division, which, after review of all documentation, authorized a special issuance for a third-class airman medical certificate. Follow up was required for two years after issuance, at which time the airman was to provide a current cardiac status report, with any imaging deemed necessary by his cardiologist.

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Aortic coarctation is a birth defect that leads to the constriction of a part of the aorta, disrupting normal blood flow and requiring an increase in cardiac output to overcome this obstruction.[1] It is a relatively common defect, seen in as many as 5-8% of congenital heart defects and can be found in association with other cardiac defects or as a singular disease.[1] The most common location for a coarctation is at the level of the ductus arteriosus and may be associated with a patent ductus, generally presenting early in infancy.[2] Other etiologies include inflammatory and autoimmune reactions (in which case the coarctation may occur anywhere along the thoracic or abdominal aorta) or traumatic dissection with acute obstruction caused by disruption of the true aortic lumen. The latter is considered a surgical emergency and requires immediate intervention.[1,7]

Due to the obstructive nature of the lesion, coarctation causes left ventricular hypertrophy by way of significantly increased afterloads.[1, 3] Presenting symptoms may range from unequal pulses through acute decompensated congestive heart failure.[1,3] In particular, neonates that present with symptoms due to ductus closure (with resultant increases in constriction and, therefore, afterload) often demonstrate signs of severe congestive heart failure and frank shock.[2,3] Other patients may initially be asymptomatic, and present much later in the disease course after the development of severe left ventricular hypertrophy and resultant hypertension or with the presentation of a heart murmur related to obstructed flow.[4]

Interestingly, the development of hypertension does not necessarily relate to the degree of aortic obstruction.[5] Furthermore, many patients present with increased peripheral resistance even distal to the coarctation. The etiology is most commonly believed to be a result of reduced renal blood flow and resultant activation of the renin-angiotensin system.[5,6]

Adults presenting with undiagnosed coarctation of the aorta may be asymptomatic and are most commonly diagnosed based on evidence of hypertension not otherwise explained, particularly with a pressure difference between the upper and lower extremities.[4,7] Higher blood pressure noted in the upper extremities, with a difference of greater than 20 mmHg, is considered diagnostic.[7] Adult patients may also present with undifferentiated systolic murmurs.[7]

Coarctation should be evaluated with an echocardiogram and/or angiography.[3,4] Surgical intervention or balloon angioplasty (with or without stenting) is often required, commonly for patients with early presentations of coarctation during infancy or childhood.[8,9] Post-surgical evaluation of patients, such as this applicant, should again include echocardiograms and/or angiography to identify any restenosis or obstruction and to evaluate the heart and heart valves for evidence of pathology, such as aortic stenosis, mitral valve disease, left ventricular hypertrophy, or other cardiac defects.[3,8,9] Even after successful surgical repair of the lesion, patients may have chronic hypertension and require even life-long therapy, most commonly with beta-blockers, ACE inhibitors, or angiotensin II antagonists.[3,4] Patients should receive endocarditis prophylaxis before any invasive procedures.[3]

The prognosis for patients, such as this applicant, with good surgical outcomes and normotension is generally good, with no restrictions to their activity levels. Complications in surgery, restenosis, aneurysms, and chronic hypertension have variable prognoses.[8,9]

Of note, a left superior vena cava, while considered a morphological abnormality, is usually a benign finding, except that is often associated with other cardiac abnormalities, particularly congenital defects.[10]

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