10. Markby DP. Lyme disease facial palsy: differentiation from Bell's palsy. BMJ. 1989; 299(6699):605–606.
11. Monsanto RD, Bittencourt AG, Bobato Neto NJ, Beilke SC, Lorenzetti FT, Salomone R. Treatment and prognosis of facial palsy on Ramsay Hunt syndrome: results based on a review of the literature. Int Arch Otorhinolaryngol. 2016; 20(4):394–400.
12. Naval Aerospace Medical Institute. 10.7. Peripheral neuropathy. In: U.S. Navy aeromedical reference and waiver guide. Pensacola (FL): Naval Aerospace Medical Institute; 2016. [Accessed 1 Dec. 2016]. Available from http://www.med.navy.mil/sites/nmotc/nami/arwg/Pages/AeromedicalReferenceandWaiverGuide.aspx.
13. Pearson V, Hesselbrock R, Van Syoc D. Bell's palsy (Mar 2015). In: Air Force waiver guide. Wright-Patterson AFB (OH): U.S. Air Force School of Aerospace Medicine; 2016:114–119. [Accessed 1 Dec. 2016], Available from http://www.wpafb.af.mil/afrl/711hpw/USAFSAM.
14. Tiemstra JD, Khatkhate N. Bell's palsy: diagnosis and management. Am Fam Physician. 2007; 76(7):997–1002.
15. U.S. Army Aeromedical Activity. Peripheral neuropathy (ICD9 356.9). In: Flight surgeon's aeromedical checklists. Aeromedical policy letters. Ft. Rucker (AL): U.S. Army Aeromedical Activity; 2014. [Accessed 1 Dec. 2016]. Available from http://glwach.amedd.army.mil/victoryclinic/documents/Army_APLs_28may2014.pdf.
16. Zandian A, Osiro S, Hudson R, Ali IM, Matusz P, et al. The neurologist's dilemma: a comprehensive clinical review of Bell's palsy, with emphasis on current management trends. Med Sci Monit. 2014; 20:83–90.

This article was prepared by Stefanie M. Watkins-Nance, M.D., M.P.H.

You are the flight surgeon preparing to leave your office one Friday afternoon when your boss stops by to see if you wouldn't mind doing him a small favor. Apparently the Wing commander just called and wants his 17-yr-old son checked out before he takes his Flying Class I physical at the U.S. Air Force Academy next week. The General explained that his son is extremely healthy and, in fact, "has never been sick a day in his life." He just wants the exam done as a precautionary measure, since the only thing his son has ever wanted to do was become an Air Force pilot. You graciously agree and a few moments later they arrive.

In walks a strapping young man who looks as healthy as the General walking in behind him had described. There is nothing significant in his medical history and he has no complaints. Vital signs are normal and you begin the physical exam. The head, ears, eyes, nose, and throat exam went well; however, as you auscultate the chest you hear a grade 2/6 systolic ejection murmur with a split S1 along with a split S2 that is wide and fixed.

1. What is the most likely diagnosis at this point?

A. Still's murmur.
B. Patent foramen ovale.
C. Ventral septal defect (VSD).
D. Atrial septal defect (ASD).

ANSWER/DISCUSSION

1. D. Approximately 10–15% of ASDs are diagnosed in adulthood, making it one of the most common congenital heart lesions diagnosed as an adult.[10] A systolic ejection murmur with a fixed split second heart sound that does not vary with respiration is pathognomonic for this lesion. This split is due to the increased blood flow across the pulmonic valve, resulting in delayed closure. There also may be a midsystolic murmur heard best along the upper left sternal border consistent with a right ventricular outflow murmur. A VSD murmur is a grade 2–3/6 harsh holosystolic murmur that is best heard over the left lower sternal border. A Still's murmur is an innocent murmur that is vibratory, sometimes called a "musical" murmur, and is also usually found at the left lower sternal border. It can be easily differentiated from a VSD by auscultating while the patient is supine, as it is louder in that position and diminishes when the patient sits up. Position has no effect on a VSD murmur. A patent foramen ovale usually has no distinguishable murmur associated with it.

"All done Doc?" the General asks. You explain that you hear a heart murmur on exam so you would like to obtain an electrocardiogram (EKG).

2. What classic findings if present on the EKG would help confirm your diagnosis?

A. A secondary R wave (R') in the right precordial leads (V1-3) and a wide, slurred S wave in the lateral leads.
B. ST elevation in all leads except in aVR, V1, and III.
C. Mild right-axis deviation and tall R waves in right ventricular leads with deep S waves in left ventricular leads.
D. Prolonged QRS, prominent QS in V1, and broad R waves in V5, V6, I, and aVL with deep S waves in V1-V2.

ANSWER/DISCUSSION

2. C. Mild right-axis deviation and tall R waves in right ventricular leads with deep S waves in left ventricular leads are seen with an ASD. A secondary R wave (R') in the right precordial leads (V1-3) and a wide, slurred S wave in the lateral leads is the EKG finding of a right bundle branch block. Stage 1 pericarditis shows ST elevation in all leads except in aVR, V1, and III. Prolonged QRS, prominent QS in V1, and broad R waves in V5, V6, I, and aVL with deep S waves in V1-V2 are indicative of left bundle branch block.

You inform the General that the exam and EKG could be consistent with a diagnosis of an ASD. He is a bit taken aback because no one has

DOI: https://doi.org/10.3357/AMHP.4776.2017

ever heard a murmur before. "Shouldn't this have been discovered earlier by his pediatrician?"

3. You explain to the General

A. ASD is the most common congenital heart disease diagnosed in adulthood because the symptoms are minimal during the first few decades of life.
B. All congenital heart diseases have been present since birth, and no defect should go undiagnosed beyond the first decade of life.
C. His pediatrician should have picked this up shortly after birth and failure to do so is grounds for malpractice.
D. Because this defect went undiagnosed for so long, many treatment options are no longer available.

ANSWER/DISCUSSION

3. A. The reason an ASD is one of the most common congenital disorders diagnosed in adulthood is because symptoms and physical findings, if present, are often subtle at best. Moderate and even large-sized ASDs may have a delayed diagnosis. Hemodynamically, significant lesions tend to be detected earlier in life, as symptoms develop earlier. The vast majority, however, are minimally symptomatic during the first three decades of life. By the fifth decade, however, over 70% are evident as impairment develops.[4]

A pilot himself, the General is concerned that his son may be disqualified from becoming a pilot. You inform him that to better answer that question you would need to obtain another study.

4. What is the next procedure of choice for determining and assessing an ASD?

A. Cardiac catheterization.
B. Transesophageal echocardiogram.
C. Transthoracic echocardiogram.
D. Exercise stress test.

ANSWER/DISCUSSION

4. C. The preferred primary method for the confident, noninvasive diagnosis and categorization of ASDs is the transthoracic two-dimensional echocardiographic examination, with the atrial septum usually best viewed in the subcostal approach.[7] A transesophageal echocardiogram can be done if transthoracic echocardiogram is of poor quality or nondiagnostic.

You are fortunate that the echocardiogram technician in the cardiac lab next door has also stayed late. This study identifies the most common type of ASD is present.

5. Which of the following is the most common type of ASD?

A. Ostium primum.
B. Ostium secundum.
C. Sinus venosus.
D. Septum primum.

ANSWER/DISCUSSION

5. B. Ostium primum, ostium secundum, and sinus venosus are the three types of ASDs; 70–75% of ASDs are the ostium secundum type, making it the most common. This type occurs where the septum primum does not form normally and subsequently fails to cover the fossa ovalis. Ostium primum is the next most common type and occurs approximately 15% of the time. The ostium primum is created when the underdeveloped endocardial cushion is unable to close this defect. For this reason, it is usually associated with a cleft anterior mitral leaflet and is often associated with Down's syndrome. Sinus venosus is a sinoseptal defect caused by abnormal embryonic evolution and is the least common type of ASD.[8]

You are excited to tell the General that this defect, although hemodynamically significant, can be successfully repaired. Although the General is grateful that his son has a correctable condition, he reluctantly asks if this means his dreams of becoming a pilot are over.

6. What is this potential flier's aeromedical disposition?

A. He is disqualified for entry into flight training.
B. He is disqualified for entry into flight training but is likely eligible for a waiver.
C. He may be considered for entry into flight training.
D. He may be considered for entry into flight training but will be restricted to multipiloted aircraft.

ANSWER/DISCUSSION

6. B. Aeromedical disposition for entrance into the Air Force depends on the type of ASD and the characteristics of any shunting, if present. Pulmonic to systemic flow ratios greater than 1.5 are considered to be hemodynamically significant. Pulmonic to systemic flow ratios less than 1.5 are insignificant and, therefore, may be considered for entry into flying training or continued unrestricted flying in a trained asset. Persons who have pulmonic to systemic flow ratios greater than 1.5, have right ventricular hypertrophy, or are experiencing symptoms need repair of the defect. Those under 25 yr of age who are successfully repaired without residual abnormalities may also be considered for entry into flying training or continued unrestricted flying, but will need a waiver. The best prognosis after repair is for ostium secundum ASDs, which is fortunate, as they are the most common type. Repairs performed after the age of 25 will be considered by the Air Force on a case-by-case basis, with waiver very likely if hemodynamically normal and no residual cardiac abnormalities exist.[11]

Navy initial candidates with this condition are considered disqualified and waivers will not be recommended. Rated and nonrated aviation personnel (all classes) will also be considered disqualified; however, if the ASD is hemodynamically stable, waiver will be recommended. Hemodynamically stable is defined as being asymptomatic, no right ventricular enlargement on echocardiogram, no fixed splitting of S2, normal EKG, and normal chest X-ray. Cases not meeting stable criteria are considered disqualifying and waivers will be considered on a case-by-case basis on designated personnel.[5]

For initial applicants in the Army (all classes), an exception to policy or waiver for initial aviation candidates is probable if surgically

corrected in childhood and without any sequelae noted after a full cardiologic assessment. For rated and nonrated Army aviation personnel (all classes), waivers are usually granted provided a complete cardiology work-up is normal and without sequelae, or postoperatively with normal recovery and postoperative cardiologic assessment. If permanent pacing is required, the aviator will be disqualified.[9]

Atrial septal defect is not specifically listed in the Federal Aviation Administration Guide for Aviation Medical Examiners, but would fall under other cardiac conditions. The guide states that any "congenital heart disease accompanied by cardiac enlargement, ECG abnormality, or evidence of inadequate oxygenation" must be deferred to the Federal Aviation Administration. Special issuance would most likely be granted once these clinically significant parameters, which may require surgical repair, were corrected.[3]

7. What are the aeromedical concerns with an unrepaired ASD?

A. Potential of right-to-left shunting of blood clots.
B. Pulmonary edema.
C. Arrhythmias.
D. All of the above.

ANSWER/DISCUSSION

7. D. Since pressure in the left atrium is higher than that in the right atrium, shunting of blood flow in ASDs usually flows from the left to right atrium, at least initially. This causes increases in right-sided volume, resulting in both right atrium and right ventricular enlargement. Shunts greater than a 1.5 pulmonary to systemic flow ratio result in significant volume overload, which may present symptomatically as easy fatigability, dyspnea secondary to pulmonary edema, or arrhythmias, including atrial fibrillation.

In the flight environment, Valsalva, anti-G straining maneuvers, and positive pressure breathing are typically a part of the aviator's everyday life. These maneuvers may cause the reversal of blood flow from right to left. This reversal could serve as a conduit for embolic material and cause disastrous results. It is for these reasons that unrepaired ASDs will not be waived.[11]

8. Which of the following refers to the process whereby hemodynamic changes occur that can result in blood flow through this ASD shifting from a left-to-right shunt to a right-to-left?

A. Ebstein's anomaly.
B. Holt-Oram syndrome.
C. Eisenmenger's syndrome.
D. Williams syndrome.

ANSWER/DISCUSSION

8. C. An increase in pulmonary blood flow as a result of the shunt often leads to the development of pulmonary vascular disease, which causes increased pulmonary vascular resistance. This increased resistance may cause an initial left-to-right shunt to then reverse and become right-to-left.[12] This is known as Eisenmenger's syndrome. Right-to-left shunts allow deoxygenated blood to enter into the arterial system; therefore, patients inflicted with this condition tend to be cyanotic. Ebstein's anomaly is characterized primarily by abnormalities of the tricuspid valve and right ventricle. It is a congenital malformation and is more prevalent in infants whose mothers took lithium frequently toward the beginning of their pregnancy.[1] Holt-Oram syndrome is characterized by upper limb abnormalities, usually involving the radius, carpal bones, and/or thumbs, along with a cardiac septal defect, most commonly being an ostium secundum ASD.[2] Williams syndrome is a genetic disorder that involves many systems, including the heart, as it is associated with supravalvular aortic stenosis.[6]

Although his ASD is disqualifying, once the defect is surgically corrected, this potential aviator will most likely be eligible for a waiver. According to Air Force guidance, those under 25 yr of age who are successfully repaired without residual abnormalities may be considered for entry into flying training.[11]

Watkins-Nance SM. ***You're the flight surgeon: atrial septal defect.*** **Aerosp Med Hum Perform. 2017; 88(6):604–607.**

ACKNOWLEDGMENTS

The author would like to thank Lt. Col. Eddie Davenport, Chief Cardiology Consultant, Aeromedical Consultation Service, for his professional review. The views expressed in this article are those of the author and do not necessarily reflect the official policy or position of the Air Force, the Department of Defense of the U.S. Government.

REFERENCES

1. Allan LD, Desai G, Tynan MJ. Prenatal echocardiographic screening for Ebstein's anomaly for mothers on lithium therapy. Lancet. 1982; 320(8303):875–876.
2. Basson CT, Cowley GS, Solomon SD, Weissman B, Poznanski AK, et al. The clinical and genetic spectrum of the Holt-Oram syndrome (heart-hand syndrome). N Engl J Med. 1994; 330(13):885–891.
3. Federal Aviation Administration. Item 36. Heart. In: Guide for aviation medical examiners. Washington (DC): Federal Aviation Administration; 2016:72–79. [Accessed 20 June 2016]. Available from http://www.faa.gov/about/office_org/headquarters_offices/avs/offices/aam/ame/guide/.
4. Marelli AJ. Congenital heart disease in adults. In: Goldman L, Schafer AI, editors. Goldman's Cecil medicine, 24th ed. Philadelphia (PA): Saunders Elsevier; 2011:397–408.
5. Naval Aerospace Medical Institute. 3.10 Atrial septal defect (ASD)/patent foramen ovale. In U.S. Navy aeromedical reference and waiver guide. Pensacola (FL): Naval Aerospace Medical Institute; 2016. [Accessed 20 June 2016]. Available from http://www.med.navy.mil/sites/nmotc/nami/arwg/Pages/AeromedicalReferenceandWaiverGuide.aspx.
6. Pober BR, Johnson M, Urban Z. Mechanisms and treatment of cardiovascular disease in Williams-Beuren syndrome. J Clin Invest. 2008; 118(5):1606–1615.
7. Shub C, Dimopoulos IN, Seward JB, Callahan JA, Tancredi RG, et al. Sensitivity of two-dimensional echocardiography in the direct visualization of atrial septal defect utilizing the subcostal approach: experience with 154 patients. J Am Coll Cardiol. 1983; 2(1):127–135.
8. Strader JR Jr, Gray GW, Kruyer WB. Clinical aerospace cardiovascular medicine. In: Davis JR, Johnson R, Stepanek J, Fogarty JA, editors. Fundamentals of aerospace medicine, 4th ed. Philadelphia (PA): Lippincott Williams & Wilkins; 2008:338–343.
9. U.S. Army Aeromedical Activity. Atrial septal defect (ASD) (ICD9 745.5). In: Flight surgeon's aeromedical checklists. Ft. Rucker (AL): U.S.

Army Aeromedical Activity; 2014. [Accessed 20 June 2016]. Available from http://glwach.amedd.army.mil/victoryclinic/documents/Army_APLs_28may2014.pdf.
10. van der Linde D, Konings EE, Slager MA, Witsenburg M, Helbing WA, et al. Birth prevalence of congenital heart disease worldwide: a systematic review and meta-analysis. J Am Coll Cardiol. 2011; 58(21):2241–2247.
11. Van Syoc D, Gore S, Davenport E. Congenital heart disease (Feb. 15). In: Air Force waiver guide. Wright-Patterson AFB (OH): U.S. Air Force School of Aerospace Medicine; Updated March 1, 2016:228. [Accessed 20 June 2016]. Available from http://www.wpafb.af.mil/afrl/711hpw/usafsam.aspx.
12. Vongpatanasin W, Brickner ME, Hillis LD, Lange RA. The Eisenmenger syndrome in adults. Ann Intern Med. 1998; 128(9):745–755.