

Indications for Left-to-Right Shunt Closure

How closure of ASD, VSD, PFO, and PDA may lead to prevention or improvement of symptoms.

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Communications between the right- and left-sided chambers of the heart result in left-to-right shunting due to the relatively low intracardiac pressures on the right side. In atrial septal defects (ASDs), the volume load affects the right ventricle, leading to enlargement and elevated pulmonary pressures. In ventricular septal defects (VSDs), as well as patent ductus, shunting leads to left ventricular overload.¹ All of these defects can lead to increased pulmonary flow resulting in severe or irreversible pulmonary hypertension and, consequently, reversal of the shunt. The mechanism of the murmur is not due to the size of the communication but rather to increased pulmonary flow.

The degree and direction of shunting are determined by the compliance of the right and left ventricles. The right ventricle is initially more compliant and predominately shunts flow left to right early in the disease process. As right ventricular volume overload and enlargement ensues, pulmonary pressures rise and the left ventricular compliance exceeds the right-sided compliance, leading to right-to-left shunting. The effects of shunts include arrhythmias, endocarditis, right and left ventricular dysfunction, paradoxical emboli, and pulmonary hypertension.² Closure of these defects may lead to prevention or improvement of these conditions.

ASD

ASDs are the most common congenital cardiac malformation, accounting for 10% of those recognized at birth and 40% of anomalies in adults older than 40 years.³

They are often asymptomatic in infancy and childhood, but when a significant shunt exists, dyspnea on exertion occurs in 30% of patients by the third decade, and in more than 75% of patients by the fifth decade of life.⁴ Right ventricular volume overload leads to right ventricular enlargement and right atrial enlargement, which can eventually result in pulmonary hypertension and right ventricular failure. The degree of shunting may increase with age as a result of increased systolic or diastolic dysfunction, affecting the left and right atrial pressure gradient.

Indications

Right atrial and right ventricular enlargement is a class I recommendation for ASD closure, either surgically or percutaneously due to the morbidity and mortality associated with elevated right-sided pressures and pulmonary hypertension. Paradoxical embolism and documented orthodeoxia-platypnea should also be considered for closure. In patients with left-to-right shunting, if the pulmonary vascular resistance is less than two-thirds of the systemic vascular resistance, closure may be contemplated. It is recommended that specific types of ASD, namely sinus venosus, coronary sinus, or primum ASDs, should be closed surgically. Secundum ASDs, the most common type, are due to defects in the fossa ovalis. Patients with secundum ASD are candidates for percutaneous closure due to their favorable anatomic location and characteristics when the defect is < 38 mm (size of the largest Amplatzer device, AGA Medical Corporation, Plymouth, MN) with appropriate margins (approximately 4 mm). The exception is the deficient anterior rim when the

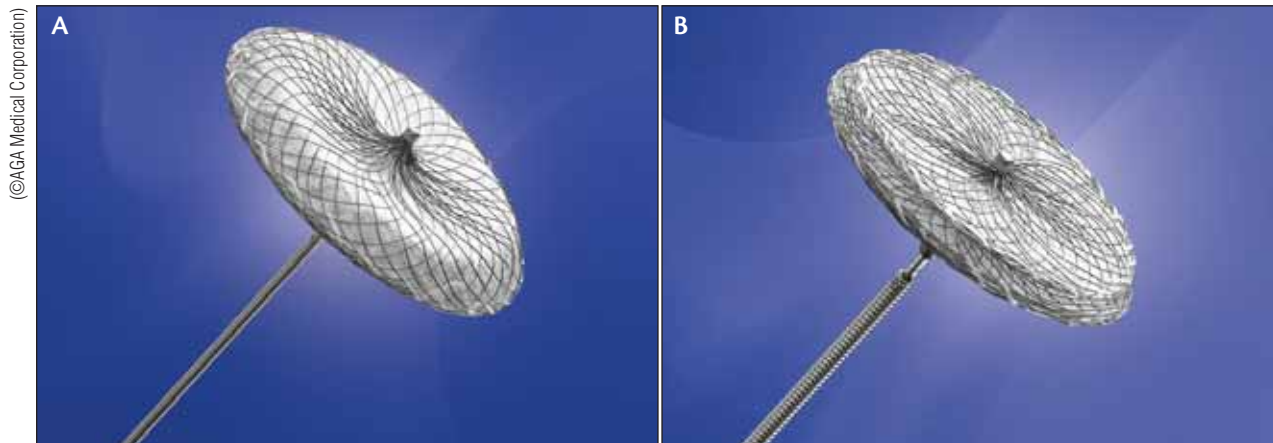


Figure 1. The Amplatzer septal occluder (A) and Amplatzer multifenestrated septal occluder "Cribriform" (B).

device can straddle the aorta.

Patients without left-to-right shunts or irreversible pulmonary hypertension are contraindicated to undergo closure.⁵ There have been reports, however, that treatment with pulmonary vasodilators may improve pulmonary pressures enough to allow successful ASD closure.⁶ If a bidirectional shunt is present, response to a pulmonary vasodilator and test occlusion of ASDs should be determined. If a favorable response (a decrease in mean pulmonary artery pressure with no decrease in cardiac output and no increase in right atrial pressure) is obtained, ASD closure may commence without reservation. If there is an unfavorable response, pulmonary vasodilators may be initiated, with reassessment in 6 months.⁷

Surgery Versus Percutaneous Closure

Surgical ASD closure has been practiced for more than 50 years and has been shown to be safe and effective, improve survival, and prevent deterioration secondary to heart failure. It is, however, associated with increased morbidity related to sternotomy, cardiopulmonary bypass, and extended intensive care stay. The comparative safety of percutaneous closure was demonstrated by Suchon et al, who showed zero mortality and similar low periprocedural complication rates and early outcomes in a cohort randomized to surgical versus percutaneous closure.⁴ The Amplatzer septal occluders (AGA Medical Corporation) and the Gore Helex occluder (W. L. Gore & Associates, Flagstaff, AZ) (Figures 1 and 2) are available and approved for use in the United States. Amplatzer FDA data have shown lower complications with a similar statistical closure rate compared to surgery.⁸ The Helex device can currently close defects ≤ 16 mm, while the Amplatzer device has sizes up to 38 mm in the United States. Sizing and success of closure has been aided with

the development of test balloon occlusion.

Percutaneous closure leads to decreased right atrial size and normalization of right and left ventricular volumes per MRI evaluation, which has not been established in postsurgical patients.⁹ Studies suggest a transcatheter approach is superior to a surgical approach in terms of improvement in myocardial function. Surgical treatment does not lead to an improvement in myocardial performance index; this has been attributed to the effects of cardiopulmonary bypass.¹⁰ Conversely, after percutaneous closure, right ventricular volume reduction at 6 and 12 months, as well as improvement in right and left ventricular function, results in a decrease in pulmonary artery pressure and improved New York Heart Association functional class.^{11,12}

Complications/Limitations

Surgical complications include infection, bleeding, and atrial arrhythmias. Fever, vomiting, or chest pain should raise suspicion for postpericardiotomy syndrome or tamponade.⁵ Percutaneous closure is associated with a low rate of long-term complications; transient atrial arrhythmias have been reported, but they are rarely clinically significant.¹³ Device erosion is rare (0.05%) and has been reported with the Amplatzer ASD occluder in association with oversizing of the device, as seen in the setting of deficient rims.¹⁴ Although device erosion and migration are uncommon, urgent evaluation is warranted should symptoms occur or if a new effusion is seen on echocardiography.

VSD

Ventricular septal defects may be congenital or result from myocardial infarction or trauma. In VSDs, complicating acute myocardial infarction, the mortality is high

(Courtesy of W. L. Gore & Associates)

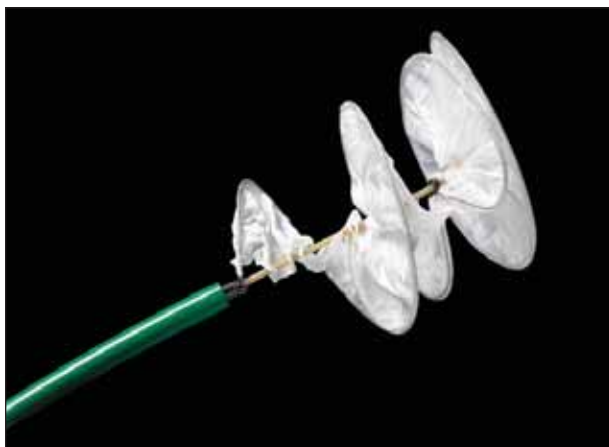


Figure 2. The Gore Helix septal occluder.

with and without closure. Some studies have reported a 2-month mortality rate as high as 90% in patients who did not undergo closure.¹⁵

In congenital patients, VSDs usually spontaneously close in childhood. In one series, up to 80% of VSDs seen at 1 month were closed at follow-up. In adults, spontaneous closure occurs in only 10% of patients. Persistent defects can lead to heart failure, arrhythmias, pulmonary hypertension, endocarditis, and aortic valve regurgitation.¹⁶ The most common VSD is perimembranous, followed by muscular and, rarely, supracristal.¹⁷ At present, only muscular VSDs can be closed percutaneously, although a perimembranous device is being developed. The remainder should be closed surgically, if indicated.

Indications

The major indications for VSD closure in adults are left-to-right shunt leading to right and left ventricular overload, prevention of endocarditis, heart failure, and aortic regurgitation. Endocarditis is associated with a 10% mortality rate in patients with a congenital VSD.¹⁶ Closure of a VSD is indicated when there is clinical evidence of left ventricular volume overload or Qp/Qs (pulmonary-to-systemic blood flow ratio) of 2.0 or more. A history of infective endocarditis is another class I indication.⁵ As recommended for ASD closure, if the pulmonary vascular resistance is less than two-thirds of the systemic vascular resistance, closure may be contemplated. Device closure should only be considered if the VSD is remote from the tricuspid valve and aorta (Figure 3). Patients with irreversible pulmonary hypertension are contraindicated to undergo closure.⁵

Surgical Versus Percutaneous Closure

Surgical treatment is still the gold standard for VSDs resulting from myocardial infarction; the mortality can



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Figure 3. The Amplatzer muscular VSD occluder.

vary from 20% to 80%.¹⁷ Surgeons often recommend a delay consisting of days to weeks to allow for formation of necrotic tissue, which provides a better anchor for closure. Postsurgical shunts are not uncommon and may lead to reintervention.¹⁵ Surgical closure does not improve intracardiac dimensions.¹⁶ Thiele et al observed the outcomes of a group of patients undergoing immediate primary transcatheter closure of postinfarct VSDs. The benefits of percutaneous closure over surgical closure included earlier treatment time, decreased mortality, and the ability to close inferior defects. Long-term follow-up of these patients revealed a mortality rate of 37% at 730 days.¹⁷ Percutaneous closure is feasible in postsurgical and post-traumatic patients. In patients with congenital muscular VSDs, the mortality and complication rates associated with percutaneous closure are very low.¹⁶ To date, a perimembranous closure device has not been approved.

Complications/Limitations

Surgery remains the traditional treatment but has increased morbidity and mortality resulting from scarring and sternotomy. Complete heart block is the main complication.¹⁸

Percutaneous closure involves a rigid sheath that may enlarge the VSD or cause rupture, with resultant dislocation into the right ventricle. Another limitation is small device sizes that are not adequate for large VSDs. Late embolization or dislocation has been described after percutaneous closure due to late healing and enlargement of the defect.¹⁷ The rates of complete heart block are similar to surgical closure and have led to pacemaker placement and, rarely, closure device removal with referral to surgery.¹⁸ Infection and arrhythmias are reported but are less common.¹⁷ To date, the Amplatzer postmyocardial infarction device is available only for compassionate use.



Figure 4. The Amplatzer PDA occluder device.

PDA

The ductus arteriosus connects the main pulmonary artery to the descending aorta. It normally closes spontaneously within 24 to 48 hours after birth. The incidence is 5% to 10% of congenital cardiac malformations and the defect is two times more common in female patients.¹⁹ Untreated mortality in adults is approximately 1.8% per year.²⁰ Cardiac catheterization provides important hemodynamic information, including pulmonary vascular resistance and degree of shunting.²⁰

Ten percent of congenital heart disease patients have the diagnosis of patent ductus arteriosus (PDA).¹⁹ PDA commonly escapes diagnosis until adulthood when it is usually found incidentally by auscultation of a murmur or as an incidental finding on transthoracic echocardiography. Asymptomatic patients may have silent pulmonary hypertension and left ventricular dysfunction.²¹

Indications

Endocarditis is responsible for almost 50% of deaths in patients with untreated PDA; patients with a history of endarteritis are strongly recommended to undergo surgical or percutaneous closure. Left atrial or left ventricular enlargement indicating left ventricular volume overload is a class I indication.²⁰ Other class I recommendations include pulmonary arterial hypertension or the presence of left-to-right shunting.²² If the PDA is calcified, is too large for device closure, or has distorted anatomy, surgical closure is recommended.²² Asymptomatic patients with a PDA are also candidates for closure with a transcatheter device. As with ASD and VSD, irreversible pulmonary hypertension with net right-to-left shunt is a contraindication for closure.

Surgery Versus Percutaneous Closure

Surgical closure has been the conventional treatment and has a negligible mortality rate but an increased morbidity rate.¹⁹ Percutaneous closure was first described in 1998;²³ since that time there have been many new generations of devices, including improved sheaths and sizes. The Amplatzer PDA occluder (AGA Medical Corporation) (Figure 4) can close defects 4 to 12 mm in size, but smaller residual defects can be occluded with coils. New advances have also provided allowance of confirmation before device release.²¹ Temporary test occlusion with a balloon catheter can help with sizing and predict the degree of success.²⁰ Percutaneous transcatheter closure is a safe alternative to surgery. It decreases endocarditis, arrhythmias, and development of pulmonary hypertension.

Complications/Limitations

Complications and limitations include residual left to right shunt, incomplete closure, hemolysis, distal embolization, and endocarditis.²¹

PFO

Patent foramen ovale (PFO) has an overall prevalence of 27%, as described in one autopsy series. The interatrial tunnel usually fuses within 2 years. In one series, it was found to be patent in 35% of children. Cryptogenic stroke is defined as an ischemic cerebrovascular episode in the absence of identifiable cause. This accounts for 30% to 40% of patients affected who are younger than 55 years.²⁴ PFO is more prevalent in patients with cryptogenic stroke than in patients with known cause of stroke (20% vs 40%–50%).²⁴

Indications

Factors suggesting need for PFO closure include younger patients (younger than 50 years), no other causes for stroke, large PFO, coexisting atrial fibrillation, recurrent events, failure of anticoagulation to prevent recurrence, intolerance of anticoagulation, and a history of pulmonary embolism or deep vein thrombosis.²⁵

In cryptogenic stroke, conventional therapy is anticoagulation and/or antiplatelet medications. There have not been any studies that substantially show one of these treatments to be superior to the other. Twenty-five percent of these patients have repeat recurrences of cerebral vascular accident (CVA) or transient ischemic attack in 4 years despite medical treatment.²⁴ Thus, medical therapy has not been shown to prevent recurrence but has been associated with increased risk of bleeding.¹³ Surgical closure increases morbidity, usually manifested as atrial fibrillation, infection, bleeding and tamponade. Percutaneous PFO closure secondary to cryptogenic stroke has a success

rate of 86% to 100%, with a recurrent stroke rate of 0% to 4.9%. Percutaneous closure is superior to conservative treatment in patients who achieve complete closure or have more than one stroke.¹³ Mas et al have shown that the recurrence rate of stroke was increased in patients with an atrial septal aneurysm that coexisted with a PFO, leading some physicians to perform closure in the setting of atrial septal aneurysm.¹³

Migraines are not currently an indication for PFO closure. The MIST trial examined this indication but the primary endpoint was not reached.²⁶ Of note, these patients had no history of transient ischemic attack or CVA. In registry data of patients with a history of transient ischemic attack/CVA plus migraine, 50% to 70% of patients had a reduction or elimination of their migraine. This population still has to be studied. There are insufficient data to make a recommendation about PFO closure in patients with a first stroke and a PFO. PFO closure may be considered for patients with recurrent cryptogenic stroke despite medical therapy.²²

Complications/Limitations

Surgical treatment leads to decreased recurrence of transient ischemic attacks and CVA, but increased morbidity due to atrial fibrillation, bleeding, infection, and tamponade.²⁶ Rates of atrial fibrillation are very low (< 2.5%), and arrhythmia is usually transient. Hazards of PFO closure include thrombus formation on the device (incidence of 2%), seen mostly with the CardioSeal device (NMT Medical, Boston, MA), as well as embolization and erosion, which are very rare.²⁵

CONCLUSION

Cardiac shunts are associated with heart failure, pulmonary hypertension, infection, and arrhythmias. Closure of these lesions may prevent or reverse these complications. Surgical closure is safe and well-validated. Percutaneous closure devices have comparable safety and efficacy in specific situations and may be associated with less morbidity due to a less invasive procedure. Surgical closure has been the conventional management for many years, but with the advent of percutaneous devices and continued advances in technology, percutaneous closure of cardiac shunts may become more prevalent. ■

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1. Julian DG, Campbell-Cowen J, McLennan JM. Cardiology, 8th ed. Elsevier Limited, Philadelphia, PA: 2005.
2. Gatzoulis MA, Swan L, Therrien J, Pantely GA. Adult Congenital Heart Disease: A Practical Guide. Blackwell Publishing Ltd, Hoboken, NJ: 2005.
3. Borow K, Karp R. Atrial septal defect. Lessons from the past, directions for the future. N Engl J Med. 1990;13:1698-700.
4. Suchon E, Pieculewicz M, Tracz W, et al. Transcatheter closure as an alternative and equivalent method to the surgical treatment of atrial septal defect in adults. Med Sci Monit. 2009;15:CR612-617.
5. ACC/AHA 2008 guidelines for the management of adults with congenital heart disease: executive summary. A report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Develop Guidelines for the Management of Adults With Congenital Heart Disease). J Am Coll Cardiol. 2008;52:1890-1947.
6. Schwerzmann M, Zafar M, McLaughlin PR, et al. Atrial septal defect closure in a patient with "irreversible" pulmonary hypertensive arteriopathy. Int J Cardiol. 2006;110:104-107.
7. Frost AE, Quiñones MA, Zoghbi WA, Noon GP. Reversal of pulmonary hypertension and subsequent repair of atrial septal defect after treatment with continuous intravenous epoprostenol. J Heart Lung Transplant. 2005;24:501-503.
8. The Amplatzer Septal Occluder System. Summary of Safety and Effectiveness Data: www.accessdata.fda.gov/cdrh_docs/pdf/P000039b.pdf. Accessed August 29, 2010.
9. Teo KS, Dundon B. Percutaneous closure of atrial septal defects leads to normalization of atrial and ventricular volumes. J Cardiovasc Magn Reson. 2008;10:55.
10. Shaheen J, Alper L, Rosenmann D. Effect of surgical repair of secundum-type atrial septal defect on right atrial, right ventricular, and left ventricular volumes in adults. Am J Cardiol. 2000;86:1395-1397.
11. Dhillon R, Josen M, Henein M, et al. Transcatheter closure of atrial septal defect preserves right ventricular function. Heart. 2002;87:461-465.
12. Schoen SP, Kittner T, Bohl S, et al. Transcatheter closure of atrial septal defects improves right ventricular volume, mass, function, pulmonary pressure, and functional class: a magnetic resonance imaging study. Heart. 2006;92:821-826.
13. Dardas PS, Ninios VN, Mezilis NE. Percutaneous closure of atrial septal defects: immediate and mid-term results. Hellenic J Cardiol. 2010;51:104-112.
14. Cheatham J. Frequency, causes, and consequences of PFO device thrombosis and erosion. Presented at the 2009 TCT annual meeting in San Francisco, CA.
15. Pedra CA, Pontes SC Jr. Percutaneous closure of postoperative and post-traumatic ventricular septal defects. J Invas Cardiol. 2007;19:491-495.
16. Bol Raap G, Meijboom FJ. Long-term follow-up and quality of life after closure of ventricular septal defect in adults. Eur J Cardiothorac Surg. 2007;32:215-219.
17. Thiele H, Kaulfersch C. Immediate primary transcatheter closure of postinfarction ventricular septal defects. Eur Heart J. 2009;30:81-88.
18. Carminati M, Butera G, investigators of the European VSD Registry. Transcatheter closure of congenital ventricular septal defects: results of the European Registry. Eur Heart J. 2007;28:2361-2368.
19. Choi DY, Kim NY, Jung MJ, Kim SH. The results of transcatheter occlusion of patent ductus arteriosus: success rate and complications over 12 years in a single center. Korean Circ J. 2010;40:230-234.
20. Wiyono SA, Witsenburg M. Patent ductus arteriosus in adults: case report and review illustrating the spectrum of the disease. Neth Heart J. 2008;16:255-259.
21. Lee CH, Leung YL. Transcatheter closure of the patent ductus arteriosus using an Amplatzer duct occluder in adults. Jpn Heart J. 2001;42:533-537.
22. Adams RJ, Albers G, Alberts MJ, et al. American Heart Association, American Stroke Association. Update to the AHA/ASA recommendations for the prevention of stroke in patients with stroke and transient ischemic attack. Stroke. 2008;39:1647-1652.
23. Masura J, Walsh KP, Thanopoulos B et al. Catheter closure of moderate- to large-sized patent ductus arteriosus using the new amplatzer duct occluder: immediate and short-term results. J Am Coll Cardiol. 1998;31:878-882.
24. Slottow TL, Steinberg DH, Waksman R. Overview of the 2007 Food and Drug Administration Circulatory System Devices Panel meeting on patent foramen ovale closure devices. Circulation. 2007;116:677-682.
25. Wu LA, Malouf JF, Dearani JA. Patent foramen ovale in cryptogenic stroke: current understanding and management options. Arch Intern Med. 2004;164:950-956.
26. Maredu RK, Shah MS. Percutaneous closure of patent foramen ovale: a case series and literature review. Clin Med Res. 2007;5:218-226.