

Atrial Septal Defect in Adults: Does Repair Always Mean Cure?

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Atrial septal defect (ASD) is the most prevalent congenital cardiac anomaly in adults¹, accounting for ~35% of all congenital heart defects. Late presentation is due to the insidious development of right ventricular remodeling, with enlargement of right cardiac chambers. Patients may seek medical attention with symptoms related to right or global myocardial dysfunction, arrhythmias or thromboembolic events. Until recently, pulmonary vasculopathy was assumed to be a rare complication in ASD patients. However, with progressive knowledge and interest in treating pulmonary arterial hypertension (PAH), numbers are changing. Of 1877 adults with congenital heart disease followed up for five years in the European Union, 896 had ASD, with PAH prevalences of 12% and 34% for repaired and unrepaired defects respectively. Fifteen patients had the full picture of the Eisenmenger syndrome².

The fortunate side of the scenario is that the widespread use of echocardiography has allowed a big number of patients to be diagnosed early. Furthermore, transcatheter closure of secundum defects offers a less invasive alternative of repair to patients who fulfill anatomical and size criteria, and may be an appropriate therapeutic option for subjects with paradoxical embolism³. Also minimally invasive video-assisted surgical techniques have allowed successful closure of the communications, and represent a new and promising therapeutic approach to patients with ASD⁴. The dark side of the story is: with late presentation, treatments may allow patients to become free from the communications, but not from advanced right ventricular remodeling and failure, severe arrhythmias and pulmonary vasculopathy.

In terms of pulmonary vascular abnormalities, a question remains: do all ASD patients successfully subjected to repair become cured²⁵. There are many important points to be considered here. First, although the risk of developing severe PAH is relatively low for patients with ASD in general (< 10%), it is higher for those with *sinus venosus* defects (~14%). Second, there is growing evidence that outcomes for patients with late postoperative PAH after repair of congenital cardiac septal defects are considerably worse when compared to those with moderate to severe pulmonary vasculopathy, who are left unoperated. Third, although there has been enthusiasm with the so called "new drugs" for PAH management, there has not been evidence

that combined medical therapy and surgery are beneficial in patients with congenital heart disease. Actually, the literature on this subject is limited to case reports and small case series, and data on long-term observations have not been provided. Based on these difficulties, a specific taskforce in the 5th World Symposium on Pulmonary Hypertension of the World Health Organization (Nice, France, 2013) came up with the conclusion that currently there is no evidence of benefit with the so called "treat-and-repair" strategy in the management of patients with congenital heart disease and PAH⁶.

On the basis of these assumptions, we would like to propose that adults with ASD who are candidates to repair (surgical or percutaneous) should be carefully evaluated. Although noninvasive diagnostic evaluation suffice in most instances, subjects suspected of having elevated pulmonary vascular resistance must be considered for more sophisticated diagnostic procedures, which include cardiac catheterization. Those presenting with a *sinus venosus* defect, importantly dilated central pulmonary arteries (Figure 1), with peripheral oxygen saturation of < 93% on exertion (in the absence of respiratory disorders) or echocardiographic findings suggestive of unusual pulmonary hemodynamics may be considered as "at risk" for postoperative complications and limited outcomes.

We therefore propose a short list of steps and indices that may be of help for decision making in adults with ASD who are potential candidates for repair. Importantly, this is no guideline. Also, we do not expect all clinicians to follow exactly the steps presented in the list. We just propose that these indices are taken into consideration, especially for "at risk" patients. Parameters listed in the Table 1 should be looked on as characteristics of an ideal candidate for ASD closure. So, we generally take them into account to decide how close or how distant patients are from the ideal condition. The general idea is that while closing an ASD, we must be quite sure that the communication will no longer be required for keeping the patient in stable conditions. That is, ASD closure in adults is associated with a "bidirectional concern". A communication may be necessary not only in patients who are at risk for right heart failure due to pulmonary hypertension or right ventricular restrictive physiology ("right-to-left concern"), but also to prevent an increase in pulmonary capillary pressure (and pulmonary edema) in subjects with relevant left heart disease ("left-to-right concern").

In conclusion, most adults with ASD are now safely treated using available techniques of surgical or percutaneous repair, and become free of complications over the long-term. However, a sophisticated diagnostic armamentarium is available, and should be used to identify a small fraction of patients who are considered as "at risk" for postoperative complications, particularly the persistence of elevated pulmonary vascular resistance and PAH. There is general agreement that ASD closure in this small population may be hazardous, with poor long-term outcomes. Individualized diagnostic evaluation and case-by-case discussion is still the best policy.

Keywords

Heart Defects, Congenital; Heart Septal Defects, Atrial/surgery; Ventricular Dysfunction, Right; Hypertension, Pulmonary; Surgical Procedures Minimally Invasive.

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Table 1 – Ideal parameters for safely assigning ASD patients to repair

1. Size of the defect: non-restrictive if >10 mm in adults
2. Anatomy: secundum type is less frequently associated with PAH
3. Normal oxygen saturation at rest and on exertion
4. Left-to-right shunting on echocardiographic analysis
5. Increased pulmonary-to-systemic blood flow ratio (Qp/Qs, echocardiography) <ul style="list-style-type: none"> • Ideally, Qp/Qs \geq 3.0 in large non-restrictive defects • Attention to patients with large defects and “not so elevated” Qp/Qs ratios (e.g. \leq 2.0)
6. Increased velocity-time integral of flow measured on pulmonary veins (echocardiography, ideally >24 cm)
7. Pulmonary artery systolic pressure not above 70 mmHg (cardiac catheterization)
8. Systolic-to-diastolic pulmonary artery pressure ratio of >2:1 (catheterization). Pulmonary arterial diastolic pressure is low in uncomplicated ASD
9. Pulmonary vascular resistance index of < 6 Wood units•m ² (ideally, <4 Wood units•m ²)
10. Normal pulmonary wedge pressure and left ventricular and diastolic pressure. Absence of significant mitral valve disease and left ventricular diastolic dysfunction
11. Absence of a right ventricular restrictive physiology
12. Absence of advanced myocardial deterioration, severe arrhythmias or relevant comorbidities as additional risk factors



Figure 1 – Enlarged heart (mainly right cardiac chambers) with prominent pulmonary artery and markedly dilated hilar vessels in an adult female with atrial septal defect. All these abnormalities may be seen in advanced pulmonary arterial hypertension. Thus, extended diagnostic evaluation is needed.

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