Case Report

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A large congenital atrial septal defect in an adult with delayed therapy

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Abstract

Patients with a large congenital atrial septal defect (ASD) traditionally have the ASD repaired at the preschool age. Unfortunately, insufficient education of patients regarding medical science and clinical recommendations can lead to delayed therapy, resulting in complications during adulthood. We report a rare case of a large congenital ASD in a 20-year-old man. Echocardiography showed a 67-mm ostium secundum defect and moderate mitral and tricuspid regurgitation. The patient underwent transthoracic ASD repair along with mitral and tricuspid valvuloplasty. This report emphasizes the importance of educating patients about congenital malformations and potential interventions in developing countries, particularly in rural communities.

Keywords

Congenital heart disease, atrial septal defect, education, adult, ostium secundum, surgery

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Introduction

Adults with untreated congenital heart disease (CHD) are at risk for long-term complications from underlying CHD, although these individuals often experience minimal symptoms.[1] Small atrial septal defects (ASDs) without obvious right heart volume overload are routinely followed without surgical closure, but increased shunting may occur later in life and surgery can become necessary.[2] Once a hemodynamically

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significant ASD is diagnosed, it should be closed electively. Closure of ASD is indicated in the presence of any hemodynamically significant shunt causing enlargement of right heart structures, irrespective of the presence of symptoms.[3] Transcatheter closure of ASD is safe and improves symptoms and longevity.[4] Even if a patient has a considerable ASD, use of a large device to cover the entire defect is recommended if the defect has sufficient rims.[5] There is no lower age limit for defect closure, but referral for surgery before school age is usual practice.[2] If closure of a hemodynamically significant ASD is delayed, a percutaneous repair may no longer be feasible and open heart surgery is required instead. This is because of the additional repair necessary of an enlarged right atrium and regurgitant mitral and tricuspid valves. Interestingly, a study in a developed country reported that, despite recommendations for life-long monitoring, individuals with CHDs may cease follow-up with cardiology providers as early as 6 years old.[6] Therefore, expanded and comprehensive education of potential complications in patients with progressive CHD is vital for improving survival, care, and quality of life for the population in developing and developed countries.[6,7]

We present a rare case of a large congenital ASD in a 20-year-old man. We emphasize the importance of educating patients about congenital malformations and potential interventions in developing countries, particularly in rural communities.

Case report

A-20-year-old man was referred to our outpatient clinic for a heart murmur that was detected while he was hospitalized for treatment of a left scapular fracture 1 month before presentation. No cyanosis was observed and the level of skin oxygen saturation in the limbs ranged from 97% to 98% under resting conditions. Apart from an accentuated second heart sound and grade 3/6 systolic murmur over the third left intercostal space, laboratory tests and a physical examination were unremarkable. Electrocardiography showed sinus rhythm with incomplete right bundle branch block. Transthoracic echocardiography demonstrated normal ventricular function, right atrial dilation, and a large secundum ASD with a diameter of 67 mm (Figure 1a). The margins of the defect were 6 to 14 mm. The right atrium and tricuspid annuli were enlarged with diameters of 66 mm and 55 mm, respectively. The diameter ratio of the aorta to pulmonary artery was 21/49 mm. The diameters of the right ventricle, left atrium, and left ventricle were 65, 44, and 50 mm, respectively. The functional parameters of the ejection fraction, fractional shortening, cardiac output, and left ventricular end-diastolic volume were 75%, 44, 7 L/minute, and 122 mL, respectively. Pulmonary hypertension with a systolic pressure of 76 mmHg (normal mean pulmonary artery pressure is 45 mmHg), moderate

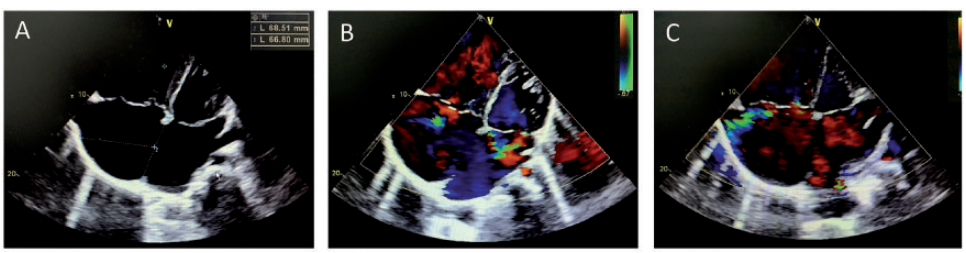

Figure 1. Preoperative echocardiogram shows an interruption in the atrial septa with a diameter of 67 mm (a, line 1), an enlarged right atrium (66 × 68 mm) (a, line 2), moderate mitral regurgitation (b), and moderate to severe tricuspid regurgitation (c).

mitral regurgitation, and moderate to severe tricuspid regurgitation with mild prolapse were observed.

We decided to treat the patient's ASD by surgery. A standard median sternotomy incision was performed. On opening the pericardium, a massive right atrium was visualized (Figure 2). The aorta was then cannulated. Separate cannulas were placed in the superior vena cava and inferior vena cava (IVC). After full heparinization, cardiopulmonary bypass was routinely applied, the aorta was cross-clamped, and cold Del Nido cardioplegic solution was instilled via the aortic root to arrest the heart. Following opening of the right atrium, careful exploration of the abnormities was completed. Mitral valvuloplasty (De Vega technique), repair of the ASD with a suitable bovine pericardial patch, tricuspid valvuloplasty with implantation of a prosthetic ring (size 32, Sorin Sovering band; Sorin Group, Saluggia, Italia), and right atrioplasty with partial excision were sequentially performed. During the operation, care was taken to check that the lower end of the superior vena cava was large enough to accommodate the superior vena cava and pulmonary venous return being separated from the left atrium. Alternatively, two separate channels may be created to ensure that these two sources of venous return are unobstructed. Similarly, the upper end of the IVC was carefully checked and sutured to avoid obstruction and a residual shunt after patch implantation. After careful hemostasis and closing of the wound in layers, the patient was carefully transferred to the intensive care unit in a stable condition. Early postoperative management included continuous arterial blood pressure monitoring and ventilation to stabilize the circulation. Transthoracic echocardiography and an electrocardiogram were performed to identify any hint of a residual shunt, pulmonary hypertension, or arrhythmia, especially atrioventricular block. The patient recovered with sinus rhythm and without further complications. Postoperative transthoracic echocardiography (Figure 3) showed no detectable flow between the two atria or valvular regurgitation, a right atrial diameter of 31 mm, and pulmonary pressure was reduced to 28 mmHg. The patient was discharged uneventfully on postoperative day 9 with recommendation for follow-up at 3 years.

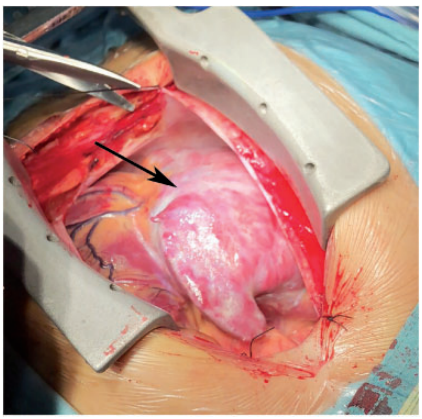

Figure 2. Intraoperative view of the enlarged right atrium (arrow).

Discussion

During the past decade, progress has been made in determining the natural history of

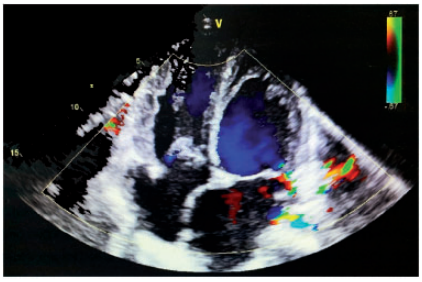

Figure 3. Postoperative echocardiogram shows no detectable interruption between the two atria and right atrial diameter of 31 mm.

ASD in adults and advances in detection and treatment have improved patients' outcomes. However, ASD remains the most underdiagnosed CHD in adults,[8] with an incidence of 22%.[9] The natural history of patients with ASD compared with that in healthy subjects is shown in Figure 4.[8,10] Interestingly, Shah et al. compared the long-term outcomes of patients with ASD with or without surgical closure in a historical prospective study.[11] These authors reported that there was no difference in survival or clinical symptoms between the two groups. Additionally, they found no difference in the incidence of new arrhythmia, stroke, or other embolic phenomena, or cardiac failure. No patient in either group developed progressive pulmonary vascular disease.[11] However, for patients with hemodynamically significant ASD, the defect is recommended to be closed electively.[2,3] There is no lower age limit for defect closure, but usual practice is to refer patients for surgery before school age. Some surgeons may offer elective closure as early as 2 years old and preferably before 5 years old.[2] The majority of secundum defects can be closed surgically with a minimally invasive procedure or percutaneously using a catheter delivery mechanism. When using surgical repair of the ASD following the gold standard approach (median sternotomy), the cardiopulmonary bypass time is typically shorter using this technique compared with the traditional technique under cardiopulmonary bypass.[2]

Recent reports have shown that operators tend to have more experience in large ASD closure using the transcatheter device approach.[12] An increasing amount of evidence has shown that patients with large ASDs (usually >38 mm) and defects with deficient rims are usually referred for surgical closure rather than transcatheter closure.[12–14]

When early diagnosis is missed or patients' follow-up is lacking, progression to a hemodynamically significant ASD can occur, resulting in further problems. Embolization through a persistent ASD may cause problems and is associated with an increased risk of stroke, especially in patients with large and long-standing defects.[15] With age-related changes in compliance, a large ASD can result in increased left to right shunting, increased pulmonary

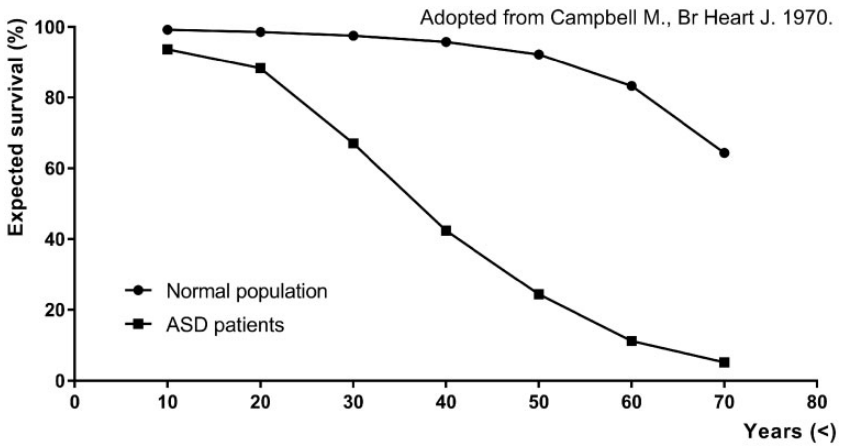

Figure 4. Graph showing expected survival of patients with ASD compared with healthy subjects (historical data).[10]
ASD, atrial septal defect.

blood flow, and progressive right-sided diastolic overload. Considerable enlargement of the right atrium or the tricuspid valve then requires surgical repair of the ASD together with the right atrium or the mitral and tricuspid valve. In the present case, we performed mitral valvuloplasty with the De Vega technique, mainly because it permitted normal anatomical growth of the valve and it is believed to be an excellent method of correcting mitral insufficiency in younger patients.[16] Our patient was 20 years old, but his body weight was not normal (42 kg). We believe that the De Vega technique would be beneficial for growth of the valve to some extent. In serious cases of ASD, once pulmonary hypertension and finally Eisenmenger syndrome has developed, repair is no longer possible. In these cases, complete heart and lung transplant or a lung transplant with repair of the ASD may be necessary. Furthermore, adult patients with an unrepaired ASD have an increased risk of psychiatric disorders compared with the general population.[17]

Complications associated with a delayed surgical repair of ASD because of the patient's choice may allow patients to become free from a shunt, but not from advanced arrhythmias, pulmonary vasculopathy, right ventricular remodeling, or even heart failure.[18] Potential complications after ASD closure in adulthood include tachyarrhythmia, bradyarrhythmia, stroke, residual shunt, right heart failure or progressive pulmonary arterial hypertension, left atrioventricular valve regurgitation and subaortic stenosis, left atrial hypertension, and pulmonary venous congestion.[19] Furthermore, there is growing evidence that outcomes for patients with late postoperative pulmonary arterial hypertension after repair of ASD are considerably worse compared with those with moderate to severe pulmonary vasculopathy, who are left unoperated.[18] Based on these difficulties, there is currently no evidence of benefit with the so called "treat-and-repair" strategy in management of patients with CHD and pulmonary arterial hypertension.[20] Most adults with ASD are currently safely treated using available techniques of surgical, individualized, diagnostic evaluation and case-by-case discussion.[18]

Many adult patients who are diagnosed with CHD are lost to cardiac follow-up and are associated with undesired outcomes.[21] For patients who are never diagnosed with CHD and may still be lost to care, the key to improving outcomes is education of the general public regarding symptoms and treatment to enhance patients' health behavior.[22] More than 80% of patients with CHD do not receive a specialist follow-up, and as a result, have poor outcomes.[23] Additionally, government and public support is critical for additional resource allocation. If patients with CHD who are lost to follow-up become aware of the noninvasive therapies, disease sequelae, and the benefits of life-long follow-up, there is great potential that they would seek expert advice. Patients with CHD can only enjoy their full life potential through educated professionals, educated adult patients with CHD, and an educated public.[23]

Conclusion

Elective surgical repair of large ASDs should be indicated once they become hemodynamically significant. Additionally, broad education of ASD-related medical science is vital and should be emphasized in developing countries, particularly in rural communities.

Ethics statement

This study protocol was approved by the Ethics Committee of the Second Xiangya Hospital of Central South University, Changsha, China. Written consent for publication of information and images was provided by the patient's parents.

Declaration of conflicting interest

The authors declare that there is no conflict of interest.

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Author contributions

All authors contributed to the conception and design of the work, and drafting and revision of the manuscript. All authors read and approved the final manuscript.

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