Long-term follow-up of children after repair of atrial septal defects

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■ **BACKGROUND** It is critical to repair atrial septal defects during childhood to minimize long-term morbidity and mortality. However, only a few studies have examined factors that predict a favorable outcome.

■ **OBJECTIVE** To examine prognostic variables in the repair of atrial septal defects.

■ **METHODS** Retrospective analysis of children who underwent repair of atrial septal defects between 1957 and 1981.

■ **RESULTS** There were 70 girls and 57 boys with a mean age of 9.3 years at the time of surgery (range 4 months to 20 years). The most common presenting symptoms were fatigue and dyspnea. Before surgery, 74% were in New York Heart Association functional class I, 70% had echocardiographic evidence of right ventricular hypertrophy, and 55% had cardiomegaly on chest radiographs. The average mean pulmonary arterial pressure was 17.1 mm Hg. The only factor significantly related to poor outcome was pulmonary hypertension. Age at surgery did not influence long-term results. Ninety-four percent of patients were in functional class I at follow-up.

■ **CONCLUSIONS** Repair of atrial septal defects is safe before age 21, but it should be done as early as possible in order to minimize the long-term complications of chronic left-to-right shunting.

■ INDEX TERMS: HEART SEPTAL DEFECTS, ATRIAL; RISK FACTORS; FOLLOW-UP STUDIES; AGE FACTORS ■ CLEVE CLIN J MED 1994; 61:29–33

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ATRIAL SEPTAL DEFECTS are the most common congenital defects of the heart, accounting for 7% of them.[1] If not surgically corrected, these lesions lead to increased morbidity and mortality,[2,3] most notably due to the development of fixed pulmonary hypertension, supraventricular arrhythmias, and ventricular failure due to the persistent left-to-right shunt.[4] Surgical repair improves survival, but in an age-dependent fashion: the mortality rate is 1.7% if surgery is performed before 40 years of age, compared with 7% if it is done after age 40.[1] Therefore, the prevalent strategy is to repair these defects during childhood.

Is there an optimal age during childhood to operate, and are there any factors that predict ultimate outcome? To address these questions, we performed a retrospective analysis and long-term follow-up of all patients age 20 and younger at the time of surgery who underwent repair of an ostium secundum atrial septal defect at The Cleveland Clinic Foundation between 1957 and 1981.

TABLE 1
PREOPERATIVE SYMPTOMS IN 127 PATIENTS WHO UNDERWENT SURGERY FOR ATRIAL SEPTAL DEFECTS

Symptoms	Number of patients (%) Age < 5 (n=25)	Age 5-10 (n=40)	Age 10-20 (n=62)
None	16 (64)	30 (75)	35 (56)
Fatigue	8 (32)	7 (18)	17 (27)
Dyspnea on exertion	5 (20)	4 (10)	17 (27)
Angina	1 (4)	1 (2)	11 (18)
Palpitations	0	1 (2)	3 (5)

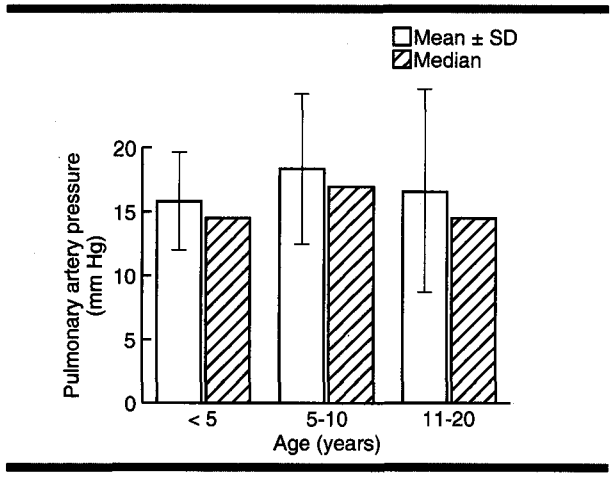

FIGURE 1. Preoperative pulmonary arterial pressure in 111 patients under age 21 who underwent surgery for atrial septal defects.

METHODS

Data gathering

Preoperative data. We recorded demographic and historical information, signs and symptoms at presentation, and any contributory laboratory, radiographic, and electrocardiographic data. If cardiac catheterization had been performed, we recorded data on hemodynamic parameters, arterial and venous saturation, vascular resistance, and degree of left-to-right shunting. Echocardiographic data on ventricular function, septal motion, and lesion type were available. Each patient's functional class was determined, based on New York Heart Association (NYHA) standards. In-hospital deaths were recorded and their cause investigated.

Operative data. The technique of operative repair for each patient was determined. All records were examined for postoperative complications, length of hospital stay, outcome, and discharge medications.

Postoperative data. We attempted to contact all patients, either by telephone or by mail, and we asked them to provide information regarding their condition since the time of hospital discharge. Eighty-four patients or their families responded. We asked them about their level of function and about any late complications or symptoms. We obtained the results of follow-up electrocardiograms, if these were available, and we inquired about any long-term cardiac medications and subsequent cardiac catheterizations or operative interventions. We recorded any late deaths and investigated their causes.

Data analysis

Only patients with ostium secundum defects with no other associated cardiac anomalies were included in the analysis. We used Fisher's exact test to determine if age, sex, or subjective and objective symptoms were related to poor postoperative outcome. Poor outcome was defined as a NYHA functional class other than I, chronic postoperative symptoms or complications, or death. The Wilcoxon rank-sum test was used to test for differences between outcome groups. Total and event-free survival were calculated for the follow-up group.

RESULTS

Presentation

Between 1957 and 1981, 127 patients underwent surgery for an atrial septal defect at The Cleveland Clinic Foundation. Their ages ranged from 4 months to 20 years, with a mean age of 9.3 years. Seventy were girls and 57 were boys. Twenty-five patients were younger than 5 years at the time of surgery, 40 were age 5 to 10, and 62 were older than age 10.

History

Patients presented with a variety of subjective complaints, including fatigue (26.0%), dyspnea (19.7%), and angina (9.4%) (*Table 1*). Eighty-one patients had no complaints at presentation.

Physical examination

On initial physical examination, 124 patients (97.6%) had murmurs and 98 (77.2%) had a fixed split second heart sound. Preoperative chest radiographs showed cardiomegaly in 71 patients (55.9%) and increased pulmonary vascular markings in 81

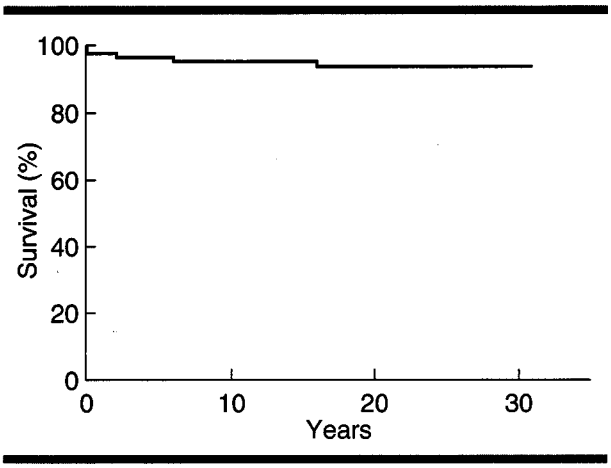

FIGURE 2. Overall survival in 84 patients who underwent surgery for atrial septal defects.

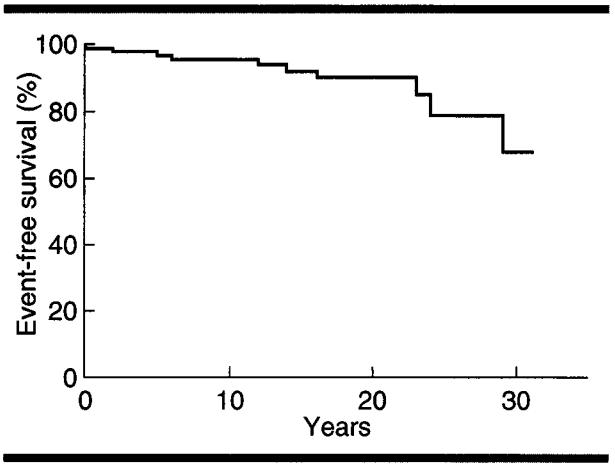

FIGURE 3. Event-free survival in 84 patients who underwent surgery for atrial septal defects.

patients (63.8%). Electrocardiograms were obtained for all patients; 123 patients (96.9%) had normal sinus rhythm.

Cardiac catheterization

Cardiac catheterization was performed in 111 patients (87.4%). All patients with associated defects apart from a secundum atrial septal defect were excluded from the study. The mean and median pulmonary arterial pressures are represented in *Figure 1*. Mitral insufficiency was present in 17 patients; it was mild in 14 (12.5%) and moderate in 3 (2.6%). No patients had severe mitral insufficiency.

Functional class before surgery

Preoperatively, 95 patients (74.8%) were in class I, and 31 (24.4%) were in NYHA class II. Data were not available for one patient. No patients were found to be in classes III or IV.

Operative technique

Patients underwent operative repair with hypothermic and bypass techniques. One hundred four patients (81.9%) underwent primary closure and 23 patients (18.1%) required patch graft placement to close their defects. There were no operative deaths.

In-hospital deaths

Two patients (1.6%) died in the hospital after surgery: one 13-year-old patient died of uncontrollable hemorrhage, and an 18-year-old died of a nonreversible low-output event.

Follow-up

We were able to obtain follow-up data for 84 (66.4%) of the 127 patients who underwent surgery. The period of follow-up ranged from 7 to 31.5 years, with a mean time of 15 years. Their ages at the time of contact ranged from 7 to 48 years, with a mean age of 25 years.

Survival

Of the 84 patients for whom follow-up data were available, 81 (96.4%) were alive. (*Figure 2*). Of the three patients who had died, one had died following a motor vehicle accident, and two had died of unknown causes. These events occurred 2, 6, and 16 years after surgery.

Symptoms

The patients' quality of life was expressed in terms of event-free survival, based on the absence of change in NYHA functional class and the absence of new or persistent cardiac symptoms. Event-free survival was 96% at 10 years after surgery, 92% at 20 years, and 73% at 30 years (*Figure 3*). At the time of follow-up, 8 patients complained of being easily fatigued, 5 had palpitations, and 3 had dyspnea on exertion. Two patients had experienced documented episodes of atrial fibrillation, and one patient had suffered from postcardiotomy syndrome (*Table 2*).

Functional class on follow-up

Seventy-six patients (93.8%) were in functional

TABLE 2
POSTOPERATIVE SYMPTOMS IN 84 PATIENTS FOR WHOM FOLLOW-UP WAS AVAILABLE

Symptoms	Number of patients (%) Age < 5 (n=25)	Age 5-10 (n=40)	Age 10-20 (n=62)
None	14 (88)	25 (89)	33 (82)
Fatigue	1 (6)	3 (11)	4 (10)
Palpitation	1 (6)	1 (4)	3 (8)
Dyspnea on exertion	1 (6)	1 (4)	1 (2)
Atrial fibrillation	0	0	2 (5)

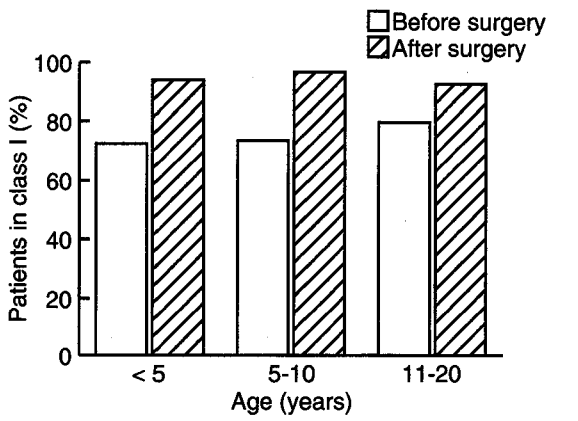

FIGURE 4. Percentage of patients in New York Heart Association functional class I before and after surgery for atrial septal defects.

class I and five were in class II on follow-up. There were no patients in class III or class IV (*Figure 4*).

Analysis

Preoperative and postoperative factors were analyzed to see if any of them were prognostic of a poor outcome. For the entire patient group, none of the factors examined were significantly related to death or to a functional class other than I. The only factor that was significantly related to chronic postoperative symptoms was a mean pulmonary arterial pressure greater than 30 mm Hg ($P < .05$). However, there was no difference in mean pulmonary arterial pressure between age groups (*Figure 1*).

DISCUSSION

The question of the optimal age for repair of atrial septal defects is an important one. The blood flow from the left to the right atrium can lead to a number of complications: pulmonary hypertension and increased pulmonary vascular resistance, supraventricular tachycardias, Eisenmenger's complex, and decreased life span.[4] Repair of these lesions is indicated to ameliorate or prevent these sequelae and, thereby, to minimize long-term morbidity and mortality. We examined several variables to see if they were significantly related to long-term morbidity and mortality in an attempt to determine the optimal age to undergo surgery.

Operative repairs of atrial septal defects have been performed for 35 years.[5] As techniques have evolved, perioperative mortality has fallen. From 1977 through 1990, several investigators have reported perioperative mortality rates between 1.7% and 3.3%.[5–7] Our results were comparable, with an in-hospital mortality rate of 1.6%. These figures indicate that repair techniques are exceedingly safe and have little impact on long-term survival.

Our study included only pediatric patients (< age 21), thus making the age at surgery an important variable to consider. Murphy et al[7] recently found that survival in patients under age 24 was not significantly different from that of age-matched controls, and age was an independent predictor of long-term survival ($P < .001$). Others recommend repair during childhood because it is safer[8] and leads to a decreased incidence of atrial dysrhythmias,[6] as well as to decreased manifestations of volume overload in the right side of the heart.[9,10] Even in patients over age 60 who have pulmonary hypertension, repair decreases pulmonary arterial pressure. However, pulmonary arterial resistance almost always remains elevated in older patients.[11] Our results corroborate these findings: in a purely pediatric population, age at the time of surgery was not significantly related to poor outcome.

Forty-three of our 127 patients were lost to follow-up at up to 30 years after surgery, and our results and conclusions must be considered in this context. Although the potential for bias is present, these data still have value, because few studies have followed up as many patients for such an extended period.

Another important variable to consider is the presence or absence of pulmonary hypertension. In one study, pulmonary hypertension occurred in 1.4% of 513 patients, all older than 27 years.[12] Only

5% to 8% of children with atrial septal defects have systolic pulmonary arterial pressures greater than 50 mm Hg.[5] When pulmonary hypertension is present, surgical risk is increased.[13,14] However, surgical repair is still preferred to medical therapy in this group; long-term survival is better, and patients enjoy better function.[15] In our study, increased mean pulmonary arterial pressure was significantly related to poor outcome ($P < .05$) for the entire patient population. However, there was no difference in mean pulmonary arterial pressure between age groups within this population. This would indicate that the incidence of pulmonary hypertension does not appreciably increase in the first two decades of life.

The presence of atrial dysrhythmias has also been studied with respect to long-term outcome. Most studies demonstrate that atrial dysrhythmias occur more frequently in adult patients with atrial septal defects than in children.[6,12] Further, the older the patient, the greater the tendency for atrial dysrhythmias to persist after surgery.[4] Our results revealed a low incidence of supraventricular arrhythmias after surgery in children, with only two documented cases (3.7%) at follow-up, and no arrhythmias preoperatively.

As part of our long-term follow-up, we assessed the patients' quality of life as reflected by their NYHA functional class and how it changed after surgery. In all three age groups, the percentage of patients in functional class I increased after repair of their defects. Reybrouck et al[16] recently showed that repair of atrial septal defects before age 5 led to improved exercise tolerance that was not significantly different from age- and sex-matched controls. However, those who underwent repair after age 5 had significantly less exercise tolerance, compared not only with controls but also with patients who underwent repair before age 5.[16] Our data, on the other hand, show improvement in all pediatric age groups. Considered together, both studies support repair before age 5, but repair after age 5 is not contraindicated.

The question remains: When should these defects be repaired? We have established that the operative risks are negligible, even in the very young, and long-term survival is enhanced by repair in childhood. If the increased volume load in the right side of the heart is not allowed to persist, the incidence of pulmonary hypertension and atrial dysrhythmias is low. When pulmonary hypertension does occur, however, it is associated with poor outcome. In addition, our data and those of others show that function and exercise tolerance approach normal levels when repair is performed early. Therefore, we advocate repair of atrial septal defects as early as possible, and definitely before the patient is 3 to 5 years old.

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