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Partial Atrioventricular Canal Defect: Long-Term Follow-Up After Initial Repair in Patients ≥40 Years Old

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Objectives. **This study was undertaken to determine the results of repair of partial atrioventricular (AV) canal in patients ≥40 years old.**

Background. **Although postoperative outcomes in younger patients have been well documented, the fate of older patients with repaired partial AV canal is less clear.**

Methods. **From 1958 to 1990, 31 patients 40 to 71 years old (mean age 51) had repair of partial AV canal. Twenty-three patients had repair of the cleft mitral valve; two had mitral valve replacements; and six needed no mitral valve operation.**

Results. **Early mortality was 6%. One patient was lost to follow-up. Nine of the early survivors are known to have died. There is a small but significant development over the long term of atrial arrhythmias, complete heart block, subaortic stenosis, recurrent mitral regurgitation and, rarely, mitral stenosis. Three of the 28 patients available for follow-up had mitral valve reoperation, and subaortic stenosis developed in 2. Nineteen patients were alive in 1991 (mean follow-up 14 years). Seven patients were in New York Heart Association functional class I, eight were in class II, and four were in class III. Fifteen of the 19 patients reported sustained postoperative improvement.**

Conclusions. **Patients ≥40 years old can have partial AV canal repair with low risk. Long-term survival is good, with subjective improvement in symptoms. Late complications occur but are uncommon, suggesting that long-term follow-up is warranted.**

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Partial atrioventricular (AV) canal represents part of the spectrum of AV septal defects, including an ostium primum atrial septal defect but with two separate AV rings, no significant interventricular communication and usually a cleft in the anterior mitral valve leaflet. The natural history of secundum atrial septal defect has been well described (1–4), with an estimated annual mortality rate of 5% to 10% for medically treated patients >40 years old and a slightly worse outcome for those with primum defects. The efficacy of surgical repair in older patients with secundum atrial septal defects has been well described (5–8), and postoperative outcomes in younger patients with partial AV canal have been well documented. However, the fate of older patients is less clear. The present study was undertaken to examine the determinants of morbidity and mortality and long-term outcome of patients having initial surgical repair of partial AV canal at ≥40 years of age. This age criterion was selected because of the tendency for the cleft mitral valve to thicken by the third and fourth decades and to determine whether mitral valve repair is still feasible in this age group.

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Methods

The records were studied for all 31 patients ≥40 years who underwent repair of partial AV canal at the Mayo Clinic between March 1958 and December 1990. Patient follow-up was from 10 to 401 months (33 years) until October 1991. Data were obtained from the initial clinical charts, and follow-up status was determined from several sources: clinical examination by one of the authors (C.A.W.), Mayo Clinic records, death certificates, patient questionnaires and the patient's current physician. Preoperative chest radiograph, electrocardiogram (ECG) and clinical information were available for all patients. Comparisons used the Fisher exact test; $p < 0.05$ was considered significant.

Results

Preoperative findings. The patient profile is shown in Table 1. There were 31 patients (21 women, 10 men). Age at operation ranged from 40 to 71 years (mean 51). Three patients also had trisomy 21. Preoperatively, patients were variably symptomatic and were assessed according to New York Heart Association functional classification (Table 1). All 31 patients had increased pulmonary vascularity on chest radiograph as well as enlarged central pulmonary arteries and cardiomegaly, consistent with left-to-right shunting. The ECG showed sinus rhythm in 26 of the 31 patients and atrial fibrillation in 5. There was the typical pattern of left-axis deviation and right bundle branch block in 21 patients (68%)

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Table 1. Patient Profile

Pt No./ Gender	Age at Op (yr)	Yr of Op	FU (mo)	FU Age (yr)	PAs	Preop MR	Preop FC	MV Op	Reop, yr	FU MR	FU FC
1/F	48	1958	401	82	40	Mild	II	Repair		Severe	III
2/F	40	1958	216	58	33	Mild	II	Repair		*	*
3/M	48	1958	126	58	38	Mild	III	Repair	TVR, 1968	*	*
4/F	43	1960	356	73	34	Mild	III	Repair		*	*
5/F	50	1961	240	70	41	Mod	III	Repair	MVR, 1969	*	*
6/M	48	1963	332	76	30	Mild	III	None	CABG, 1979	Mod	II
7/F	47	1963	339	76	31	None	II	Repair	MVR, 1989	Mild	II
8/F	41	1963	338	69	25	Mild	II	Repair (DO)		Mod	I
9/F	51	1964	223	68	51	Mod	III	Repair		*	*
10/M	53	1967		53	24	Mild	II	Repair		†	†
11/M	48	1968	55	52	45	None	III	None		ND	ND
12/F	50	1972	226	69	24	Mild	I	Repair		None	I
13/M	52	1972	1	52	80	Severe	IV	MVR+AVR		*	*
14/F	71	1974	209	89	53	Mod	IV	MVR		Severe	III
15/F	60	1974	87	68	50	Mod	IV	Repair+CABG		*	*
16/M	56	1975	201	73	63	Mild	II	None (DO)		Mod	I
17/F	40	1976	181	55	43	Mild	II	Repair		Mild	III
18/F	50	1977	174	64	38	None	III	None		ND	II
19/F	68	1979	143	80	66	Mild	II	Repair		Mild	II
20/F	64	1981	126	75	25	Mild	III	Repair		*	*
21/F	42	1981		42	68	Mild	III	Repair		†	†
22/F	45	1983	104	53	35	Mild	II	Repair		ND	II
23/F	44	1984	79	52	31	Mild	II	Repair		Mild	I
24/F	40	1984	72	48	44	Mild	II	Repair	MVR/AVR, myectomy, 1991	None	II
25/F	65	1985	76	72	46	Severe	II	Repair/An		ND	I
26/M	66	1986	65	71	65	Mod	III	Repair/An		Mod	III
27/F	44	1986	62	50	45	Mild	I	None (DO)		Severe	II
28/M	53	1987	7	54	39	Mod	III	Repair+CABG		*	*
29/F	40	1987	55	44	46	Mild	I	None		ND	I
30/M	62	1989	30	65	60	Severe	II	Repair/An		Mild	I
31/M	57	1990	10	58	42	Severe	I	Repair/An+CABG		Mild	II

*Late death. †Early postoperative death. An = annuloplasty; AVR = aortic valve replacement; CABG = coronary artery bypass graft surgery; DO = double orifice; F = female; FC = New York Heart Association functional class; FU = follow-up; M = male; Mod = moderate; MR = mitral regurgitation; MV = mitral valve; MVR = mitral valve replacement; ND = no data; Op = operation; PAs = pulmonary artery systolic pressure (mm Hg); Preop = preoperative; Pt = patient; Reop = reoperation; TVR = tricuspid valve replacement.

and first-degree AV block in 16 of the 26 patients in sinus rhythm (9). Fourteen patients underwent echocardiography, 27 had cardiac catheterization, and 11 had both tests. The degree of preoperative mitral regurgitation was determined from several sources, including data from cardiac catheterization, echocardiography and double-sampling dye curves at the time of the surgical procedure. The overall degree of mitral regurgitation was mild in 18 patients, moderate in 6, severe in 4 and absent in 3.

Data for pulmonary artery pressures were obtained from echocardiographic or catheterization laboratory data when available; in two patients data were obtained intraoperatively. Pulmonary artery systolic pressures in all patients ranged from 24 to 80 mm Hg (mean 44). Nine patients had pulmonary artery systolic pressure ≥50 mm Hg. Preoperative left ventricular ejection fraction in 10 of the 31 patients ranged between 47% and 70% (mean 59%). Shunt ratios of pulmonary/systemic blood flow were obtained in 25 patients and ranged from 1.8 to 7.1 (mean 3.3). Pulmonary arteriolar resistance index was obtained in 16 patients and ranged from 0.5 to 8.5 units·m^2 (mean 2.9). Three patients had significant coronary artery disease at initial operation.

Twenty-eight patients had an isolated cleft in the anterior mitral valve leaflet. Three patients (Patients 8, 16 and 27) had a double-orifice mitral valve in which each papillary muscle receives the respective chordae of two small valve orifices. One of the three (Patient 16) had no mitral valve cleft (Table 1). There were several associated lesions (Table 2). A membranous ventricular septal aneurysm was present in six patients, including all three with trisomy 21, one of whom (Patient 24) also had subaortic stenosis, bicuspid aortic valve and secundum atrial septal defect.

Surgical procedures. All patients had repair of the primum atrial septal defect, 2 with suture closure and 29 with patch closure. Prosthetic patch material was used in 24 patients, including three repairs with Ivalon sponge. The remaining five repairs were done with pericardium. The mitral valve was treated in several ways (Table 1). Twenty-three patients had

Table 2. Additional Congenital Cardiac Lesions in 31 Patients With Partial Atrioventricular Canal

Associated Congenital Lesions	No. of Patients
Membranous ventricular septal aneurysm	6
Secundum atrial septal defect	4
Coronary sinus atrial septal defect	1
Patent foramen ovale	4
Double-orifice mitral valve	3
Triple-orifice tricuspid valve	1
Other tricuspid valve abnormalities	5
Subaortic stenosis	1
Bicuspid aortic valve	1
Persistent left superior vena cava	3

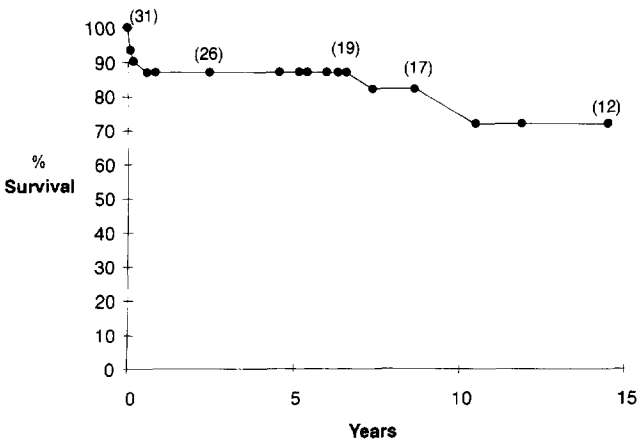


Figure 1. Kaplan-Meier survival curve for 31 patients with repair of partial atrioventricular canal. Actuarial survival at 5 years was 87%, and that for 10 years was 72%. **Numbers in parentheses** = number of patient survivors.

suture repair of the anterior mitral valve leaflet with apposition-ing of the cleft edges. Of the 19 patients with mitral valve repair alone (no annuloplasty), 14 had mild regurgitation, 4 had moderate regurgitation, and 1 had no valvular leak. Of four patients with both mitral cleft suture repair and annuloplasty, one had moderate and three had severe mitral regurgitation. Six patients with mild or no mitral regurgitation had no mitral valve repair, including one (Patient 16) with a double-orifice valve without a cleft.

Two patients had mitral valve replacement. One 52-year old patient (Patient 13) had been referred with a diagnosis of rheumatic heart disease with mild aortic stenosis and regurgitation and severe mitral regurgitation. Before operation in 1972, the diagnosis of partial AV canal had not been considered, and mitral and aortic valve replacements were performed. The second patient (Patient 14, 71 years old) was the oldest referred in this series for surgical repair. At operation, the mitral valve was calcified and was therefore replaced.

Two of the three patients with double-orifice mitral valve had an anterior mitral valve leaflet cleft, one of which was closed by suture (Patient 8). Two patients (Patients 15 and 26) had severe tricuspid regurgitation and required tricuspid valve annuloplasty. The patient with subaortic stenosis (Patient 24) had a myotomy and subaortic resection. Three patients (Patients 15, 28 and 31) had coronary artery bypass graft surgery at initial repair, one of whom (Patient 28) had had a previous left ventricular aneurysmectomy and coronary artery bypass surgery 12 years earlier and needed excision of a calcified false aneurysm at the site of the previous suture line as well as a right coronary artery bypass graft.

Early postoperative follow-up. There were no intraoperative deaths. The postoperative hospital stay ranged from 7 to 47 days (mean 14). Seven patients had a hospital stay >14 days (Table 3). Three patients (Patients 5, 13 and 25) had atrial arrhythmias that were associated with a mild stroke in one. Patient 3 had complete heart block but was asymptomatic. The year was 1958, and he did not receive a pacemaker.

There were two early (≤30 days postoperatively) deaths, giving a perioperative mortality of 6%. Patient 10 had a low cardiac output postoperatively and pulmonary edema associated with atrial fibrillation. On day 9, he died of pulmonary embolus.

Patient 21 had the highest calculated pulmonary arteriolar resistance in the group (8.5 units·m^2). Her postoperative hospital stay was short and was complicated only by atrial fibrillation. She died in the night at 24 days postoperatively. Autopsy was refused, and death was presumed to be due to arrhythmia.

Late mortality. The Kaplan-Meier actuarial survival curve is shown in Figure 1. There were nine late deaths (i.e., >30 days postoperatively, range 43 days to 30 years) (Table 4). The

Table 3. Prolonged Postoperative Course

Pt No./ Gender	Age at Op (yr)	Preop MR	Preop FC	MV Op	Postop Stay (days)	Postop Complication
3/M	48	Mild	III	Repair	23	CHB (asymptomatic), not paced
5/F	50	Mod	III	Repair	23	Atrial arrhythmias, residual MR, pericardiotomy syndrome
8/F	41	Mild	II	Repair (DO)	47	Pleural effusion, decortication of lung postop day 24
13/M	52	Severe	IV	MVR+AVR	15	Atrial arrhythmias, stroke
18/F	50	None	III	None	16	Reop secondary to bleeding
25/F	65	Severe	II	Repair/An	17	Atrial arrhythmias
28/M	53	Mod	III	Repair+CABG	42	Atrial arrhythmias, CHF, wound dehiscence

CHB = complete heart block; CHF = congestive heart failure; Postop = postoperative; other abbreviations as in Table 1.

Table 4. Late Mortality

Pt No./ Gender	Age at Op (yr)	Preop MR	Preop FC	MV Op	Postop Time to Death	Postop Complications and Course
2/F	40	Mild	II	Repair	18 yr	Traffic accident
3/M	48	Mild	III	Repair	10 yr	Paced 9 yr postop; TVR 10 yr postop; fell, and cerebral hemorrhage developed
4/F	43	Mild	III	Repair	30 yr	ND
5/F	50	Mod	III	Repair	20 yr	MVR 8 yr postop, ventricular tachycardia, myocardial infarction
9/F	51	Mod	III	Repair	16 yr	CAD, CHF, diabetes
13/M	52	Severe	IV	MVR+AVR	43 days	Stroke, hemolysis, CHB (long bypass)
15/F	60	Mod	IV	Repair+CABG	8 yr	Myocardial infarction
20/F	64	Mild	III	Repair	11 yr	CAD, CHF, LVEF 32%, mod MR
28/M	53	Mod	III	Repair+CABG+aneurysmectomy	7 mo	LV dysfunction, LVEF 19%, CHB

CAD = coronary artery disease; EF = ejection fraction; LV = left ventricular; Mod = moderate; MR = mitral regurgitation; other abbreviations as in Table 1 and 3.

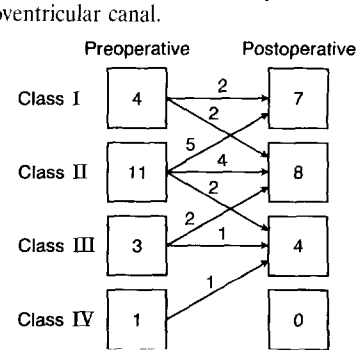
earliest death was in Patient 13, who was in functional class IV preoperatively. He had aortic and mitral valve replacements with a long bypass time and died of stroke with multiorgan failure. Five other deaths (Patients 5, 9, 15, 20 and 28) were related to coronary artery disease and left ventricular dysfunction, complicated in one patient (Patient 9) by diabetes mellitus.

Survivors. There were 29 early survivors and 9 late deaths. This yielded 20 known survivors, with follow-up data to October 1991 obtained for 19; 1 patient (Patient 11) was lost to follow-up after 4 years. All 19 survivors have had a follow-up period ranging from 10 to 401 months (mean 158); age at latest follow-up ranged from 44 to 89 years (mean 65).

Clinical status. At follow-up evaluation, rhythm was sinus in 11 patients, atrial fibrillation in 6 and paced in 2. Of the 13 late survivors with suture repair at initial operation, data about residual mitral regurgitation were available for 11. Two patients had no regurgitation; mitral regurgitation was mild in six patients, moderate in two and severe in one. Of the six patients with no suture repair of the mitral valve, echocardiographic data from three demonstrated moderate or severe mitral regurgitation.

Seven patients were in functional class I, eight in class II and four in class III (Fig. 2). The three patients with double-orifice mitral valves have survived from 62 months to >28 years postoperatively and are in functional class I or II. Overall, 15 of 19 patients reported an overall improvement in symptoms and functional class postoperatively, ranging from 25% to 100% improvement (mean 73%) (Fig. 2).

Figure 2. Changes in functional class of 19 patients undergoing repair of partial atrioventricular canal.



Four patients reported feeling worse; severe mitral regurgitation developed in two (Patients 1 and 27) 34 and 6 years postoperatively. Patient 17 has functional class III symptoms 15 years postoperatively, with only mild mitral regurgitation but severe emphysema. The fourth patient (Patient 31) reports class II symptoms despite coronary artery bypass grafting.

Reoperation. There were five reoperations (Table 1): three for mitral valve replacement (Patients 5, 7 and 24), one for coronary artery bypass grafting (Patient 6) and one for tricuspid valve replacement (Patient 3). All three patients requiring reoperation for mitral valve replacement had previous mitral valve repair. Predominant mitral stenosis had developed in two patients 6 and 26 years later, and severe mitral regurgitation had developed in the other. Patient 7 was 47 years old at initial operation, and to avoid heart block, the patch was sutured onto the mitral valve. Twenty-six years later moderate mitral regurgitation and stenosis developed. At reoperation, there was endocardial proliferation over the mitral leaflets, so she had a St. Jude mitral valve replacement.

Patient 24 was 40 years old at initial operation, when she had mild mitral regurgitation and subaortic stenosis requiring myectomy. Six years later, moderate mitral stenosis, stenosis of her bicuspid aortic valve and tunnel subaortic stenosis (mean gradient 50 mm Hg) developed, necessitating mitral and aortic valve replacements.

The third patient (Patient 5) had severe mitral regurgitation 8 years postoperatively and a small residual atrial septal defect. At reoperation (at the Mayo Clinic), the mitral valve cleft was bordered by thickened cartilaginous leaflet tissue, necessitating mitral valve replacement with suture closure of the residual atrial septal defect.

Patient 6 required coronary artery bypass grafting 16 years after repair and, at follow-up 28 years postoperatively, was in functional class II. The last patient (Patient 3) had a pace-

maker insertion 9 years postoperatively. During the next year, severe tricuspid regurgitation developed, necessitating valve replacement.

Five patients required permanent pacemaker insertion; complete heart block developed perioperatively in only one (Patient 3). Complete heart block developed spontaneously in one patient (Patient 14) at age 78 years, 7 years after repair. Three other patients needed pacing (Patients 5, 7 and 31), but all had reoperation for either mitral valve replacement or coronary artery bypass grafting at some time previously.

Two patients had recurrent subaortic stenosis. Patient 24 had recurrence 7 years later that required aortic and mitral valve replacement and repeat myectomy. The second patient (Patient 1), an 82-year old woman, had no preoperative cardiac catheterization or echocardiography at age 48 years. At follow-up evaluation, she had severe mitral regurgitation and functional class III symptoms associated with severe discrete subaortic stenosis.

Discussion

To our knowledge, this report represents the largest series of patients who have had initial repair of partial AV canal at age ≥40 years, including the oldest survivor of such a procedure (now 89 years old). Although reports exist describing small numbers of these adult patients, long-term follow-up has not been well described (10–13). Others have described (3,8,14–18) angiographic features, natural history in younger patients, associated defects and methods of corrective operation. Although the present study group represents the biases of a tertiary referral center, follow-up is complete except for one patient lost to follow-up.

The early (30-day) mortality in this series was 6%, with an actuarial 5-year survival of 87% and 10-year survival of 72%. These figures are similar to the 10% perioperative mortality reported in other large series (14,15) of predominantly young patients.

Clinical features. Only four patients in this series were in functional class I at presentation, suggesting that patients surviving to middle adulthood generally become more symptomatic with advancing age, which supports closing these defects at detection, even in those >40 years old. Preoperatively, atrial fibrillation was present in 3 of the 7 patients >60 years old and in only 2 of the 24 younger patients. All patients in atrial fibrillation were in functional class III or IV. This corresponds to accepted experience of worsening rhythm problems with advancing age as well as clinical deterioration with atrial arrhythmias (4). Only 1 of 18 patients who presented between age 40 and 50 years had moderate or severe mitral regurgitation. Conversely, none of the patients ≥60 years old were without mitral regurgitation, and five of seven had moderate or severe mitral regurgitation ($p = 0.002$, Fisher exact test).

Surgical procedures. All but two patients in the initial series were able to have mitral repair when indicated, suggesting that repair can be achieved in the majority of older patients, even though there is a tendency for cleft mitral valves to become thicker with advancing years and for the mitral annulus to calcify (19). Only the oldest patient (71 years) had a calcified mitral valve and had to have a mitral valve replacement. The second patient underwent operation in 1972 by a surgeon who was not comfortable with the technique of mitral valve repair.

The long-term results of mitral valve repair were also excellent. Only three patients needed mitral valve replacement 6 to 26 years after initial repair; in one patient (Patient 24) this was undoubtedly related to abnormal mitral valve attachments contributing to subaortic stenosis that could not be relieved without sacrificing the mitral valve. Such patients with abnormal mitral valve attachments may represent a different (perhaps more severe) subset of this anomaly with a different natural history (20,21).

Subaortic stenosis developed in two patients; in one it recurred 7 years after resection, and in the other (Patient 1) there was no evidence of it at the initial procedure, confirming reports that this is a progressive, acquired lesion (20).

The three patients with double-orifice mitral valves all presented with minimal symptoms, and all were in functional class I or II at follow-up despite moderate to severe mitral regurgitation. The patient with the cleft double-orifice valve that was not sutured progressed to severe mitral regurgitation, whereas the patient with the sutured cleft double-orifice valve had less significant regurgitation at follow-up. Although this lesion may pose a technical challenge for the surgeon, suture of an associated cleft may improve the long-term results (22,23).

Only the earliest patient in this series had complete heart block perioperatively (incidence of 3%). This is in keeping with our current surgical experience with a low incidence of heart block. In one patient heart block developed 7 years postoperatively, but in the eighth decade of life, when additional degenerative change in the conducting system might also be implicated. Three patients who required pacing years after operation had all had further operation for mitral valve replacement or coronary artery bypass grafting.

Postoperative hospital stay seemed to correlate with outcomes. Those whose stay was >12 days had reduced survival that appeared to be independent of the year of the surgical procedure or of the attending surgeon.

Pulmonary pressures. Six patients had a preoperative pulmonary artery systolic pressure >60 mm Hg. Two patients died, and both were in functional class III or IV preoperatively. Of the three patients who died within 2 months of operation, two had the highest preoperative pulmonary artery pressures recorded in this study. This corresponds to findings of another large series (13) of primum atrial septal defect repair, in which the variable that indicated operative mortality most strongly was pulmonary hypertension. All 4 survivors with an initial pulmonary artery systolic pressure >60 mm Hg were in atrial fibrillation at follow-up, in contrast to only 3 of the 16 survivors in the group with a pulmonary artery systolic pressure <40 mm Hg. This is similar to the findings in patients >50 years old with secundum atrial septal defect (6). There was a

positive correlation between functional class and severity of systolic pulmonary artery pressure and an increase in pulmonary artery pressure with age.

The three patients with trisomy 21 have all survived long term. Although an increased risk of pulmonary hypertension has been reported with this syndrome (24–26), none of our three patients had significantly increased pulmonary pressures, and overall they had a good outcome.

Conclusions. Partial AV canal defect can be repaired safely in the adult population. Operative mortality is low, and survival compares well with secundum atrial septal defects repaired early. Mitral valve repair should be used when possible; it gives excellent results. In keeping with the findings of other series involving younger patients (14–16), functional status usually improves.

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