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Original Article

Optimal surgical approach for repair of aortopulmonary window

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Abstract *Background:* This is a review of the experience over 26 year in a single institution with surgical repair of aortopulmonary window. *Methods:* Between July 1973 and March 1999, 38 patients underwent surgery for aortopulmonary window at a median age of 5 weeks, and with a median weight of 3.9 kg. Median follow-up was 6.6 years, with a range from 0.8 to 26 years. Additional defects were present in 25 (65%) patients, including interruption of the aortic arch in 7, tetralogy of Fallot in 7, ventricular septal defect in 5, functionally univentricular anatomy in 3, aortic coarctation in 2, and anomalous origin of a coronary artery in 1. We approached via an aortotomy in 45%, an incision through the defect in 31%, and using a pulmonary arteriotomy in 24% of patients. Closure was achieved using a single patch in 30 patients (79%). *Results:* There were 3 (7.9%) in-hospital deaths. Actuarial patient survival was 88% at 10 years. Three patients required reinterventions for stenoses of the great arteries. Freedom from any reintervention was 70% at 10 years. By multivariate analysis, the approach through a pulmonary arteriotomy was shown to result in a higher need for reintervention ($p = 0.01$). *Conclusions:* Repair of aortopulmonary window can be done with excellent results. A pulmonary arteriotomy should be avoided.

Keywords: Aortopulmonary fenestration; surgical repair; congenital heart disease

Aortopulmonary window is a rare lesion which occurs in approximately 0.15% of children with congenital cardiac malformations.[1] It consists of a communication between the ascending aorta and the pulmonary trunk in the presence of two normally formed arterial valves. It can be found as an isolated defect, but is commonly associated with other complex cardiac anomalies, such as interruption of the aortic arch or tetralogy of Fallot.[2] If uncorrected surgically, two-fifths of affected children will die during the first year of life, and a large number will succumb to sequels of congestive heart failure or pulmonary vascular disease in childhood.[3] Pulmonary vascular disease is also the major cause of mortality in older children undergoing repair of aortopulmonary window.[4,5] Mortality after early closure has also been correlated with the presence of other complex malformations.[5,6] We describe here our entire experience with this entity since the first on-pump repair of aortopulmonary window at this institution. We focussed on identifying risk factors for early and late mortality, as well as reinterventions.

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Material and methods

A review was made of the charts from all patients undergoing repair of aortopulmonary window at The Children's Hospital, Boston, between July 1973 and March 1999. We had identified 38 patients through the cardiac database, and they form the basis for this study. Data were obtained from review of the charts, as well as, if possible, written communication with the patient's physician. The most

recent echocardiogram available was reviewed. All data procurement was carried out after institutional approval according to guidelines established by the committee on clinical investigation.

Definitions

A simple aortopulmonary window has no associated cardiac defects, or only simple defects requiring minor intervention such as patency of the arterial duct or atrial septal defect, while a complex aortopulmonary window has one or more associated complex cardiac malformations.

Statistical analysis

Actuarial rates of survival and reintervention were calculated by the Kaplan-Meier product-limit method with 95% confidence intervals derived by Greenwood's formula. Multivariable analysis was performed using Cox proportional-hazards regression model with hazard ratios used as the measure of risk. Two-tailed p values less than 0.05 were considered statistically significant. Variables included in the analysis were age, gender, weight, surgical approach, technique of repair, diagnosis, and need for reintervention treated as time-dependent covariate (Table 1).

Results

Demographics and follow-up

There were 21 females (55%) and 17 male patients. The median age at operation was 5 weeks, with a range of 3 days to 7 years. The mean weight at surgery was 3.9 ± 3.3 kg, with a range from 0.7 to 18 kg. The follow-up ranges from 6 months to 26 years, with a median follow-up of 6.6 years. Three patients were lost to follow-up early after surgery (Table 2).

Anatomic descriptions

The defect was located exclusively between the ascending aorta and pulmonary trunk in 27 patients, and these lesions were considered to be proximal windows. It extended to the origin of either the right or the left pulmonary artery in 8 patients, and produced a communication between the right or left pulmonary artery and the ascending aorta, with no communication between the pulmonary trunk and the ascending aorta in 3 patients, these being considered distal aortopulmonary windows.

Associated anomalies

Major complicating lesions were found in 25 patients (65%), including 7 with interruption of the aortic arch, at the isthmus in 6 and between the left common carotid and subclavian arteries in 1, 7 with tetralogy of Fallot, with 4 of these having pulmonary atresia and 3 with pulmonary stenosis, 5 with ventricular septal defects, 3 with double-outlet right ventricle and small left-sided features, 2 with aortic coarctation, and one with an anomalous right coronary artery arising from the pulmonary trunk.

Surgical techniques

Techniques of repair differed across the series. From 1952 to 1973, 13 patients had simple ligation or oversewing of the defect without cardiopulmonary bypass. These patients have not been included in this report. After 1973, all cases employed cardiopulmonary bypass. Deep hypothermic arrest was

Table 1. Multivariable analysis of risk factors for reintervention or long-term survival by Cox proportional-hazards regression analysis. Two-tailed p values less than 0.05 were considered statistically significant.

Predictor of Survival	Variable p value	Predictor of reintervention	Variable p value	Variable hazard ratio	95% CI
Age	0.75	Age (neonate)	0.001	7.4	2.1–30.0
Gender	0.50	Any additional diagnosis	0.04	8.0	2.0–32.1
Weight	0.11	TOF	0.006		
Surgical approach	0.69	Transpulmonary approach	0.01	5.2	1.6–16.8
Additional diagnosis		Gender	0.84	N/A	N/A
IAA	0.98	Weight	0.98	N/A	N/A
TOF	0.93	Closure technique		N/A	N/A
Closure technique		single patch	0.87		
single patch	0.97	double patch	0.86		
double patch	0.96	primary	0.81		
primary	0.93				
Reintervention	0.72				

Abbreviations: IAA: interrupted aortic arch, TOF: tetralogy of Fallot

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Table 2. Details of patients with aortopulmonary window operated at The Children's Hospital, Boston, between 1973 and 1999.

Year of operation	Age at surgery	Location of defect	Associated anomalies	Exposure	Type of repair	Immediate outcome	Reintervention	Status at F/U
1973	2 years	Proximal	TOF/PA	Pulm	Si. patch	Lg trm vent	Cdt chg	Alive
1975	6	Proximal	VSD	Pulm	Si. patch	Good	None	Dead
1976	1 year	Proximal	TOF/PA	Pulm	Primary	Good	RVOT augment	Alive
1976	7	Proximal	None	Aortic	Si. patch	Lg trm vent	None	Alive
1979	12	Proximal	TOF	Pulm	Si. patch	Good	RVOT augment	Alive
1979	72	Proximal	VSD	Aortic	Si. patch	Good	None	Alive
1980	71	Distal	DORV, MS	Window	Narrowing	Good	Glenn, AP Shunt	Dead
1981	19	Proximal	VSD	Pulm	Si. patch	Good	None	Alive
1981	1	Proximal	DORV, small LV	Aortic	Si. patch	Lg trm vent	Fontan	Alive
1984	6	Proximal	None	Aortic	Si. patch	Dead	N/A	Dead
1984	7	Proximal	Coarctation	Aortic	Si. patch	Good	None	Alive
1984	7	Proximal	DORV, small LV	Aortic	Si. patch	ARF	N/A	Lost F/U
1986	10	Distal	Coarctation	Aortic	Si. patch	Card arrest	None	Alive
1987	18	Proximal	VSD	Aortic	Si. patch	Good	N/A	Lost F/U
1989	2	Proximal	None	Aortic	Si. patch	Good	None	Alive
1989	11	Proximal	None	Aortic	Si. patch	Good	N/A	Lost F/U
1989	3	Distal	None	Aortic	Si. patch	Good	None	Alive
1989	11	Proximal	IAA (B), DORV	Aortic	Si. patch	Good	None	Alive
1991	3	Distal	IAA(A)	Window	Si. patch	Good	RPA dilation	Alive
1991	1 year	Proximal	TOF/PA	Aortic	Primary	Good	None	Alive
1992	2	Proximal	None	Window	Primary	Good	None	Alive
1993	1	Distal	IAA(A)	Pulm	Si. patch	Good	RPA/AA dilation	Alive
1993	3	Proximal	None	Aortic	Do. patch	Good	None	Alive
1993	1	Proximal	ARCAPA	Aortic	Si. patch	Dead	N/A	Dead
1994	1	Distal	IAA(A)	Window	Si. patch	Good	None	Alive
1994	5	Proximal	TOF	Window	Si. patch	Mediastinitis	None	Alive
1994	1	Proximal	TOF	Aortic	Si. patch	Good	RVOT augment.	Alive
1995	3	Proximal	IAA(A)	Aortic	Si. patch	Good	None	Alive
1996	1	Distal	IAA(A)	Window	Si. patch	Good	None	Alive
1996	7	Proximal	None	Window	Si. Patch	Good	None	Alive
1996	6	Distal	ASD, valvar PS	Window	Do. patch	Good	None	Alive
1997	33	Distal	VSD	Window	Si. patch	RSV/ECMO	None	Alive
1997	3	Proximal	None	Pulm	Si. patch	Good	PA dilation	Alive
1997	4 years	Proximal	None	Window	Si. patch	Good	None	Alive
1998	2	Distal	IAA(A)	Pulm	Si. patch	Good	None	Alive
1998	6	Proximal	TOF/PA	Transpulm	Primary	Good	Cdt dilation	Alive
1998	7 years	Distal	None	Window	Do. patch	Dead	N/A	N/A
1999	1	Proximal	None	Window	Si. patch	Good	None	Alive

Age at surgery is in weeks, unless marked as years. A proximal defect involves only the ascending aorta and the pulmonary trunk.

Abbreviations: AA: aortic arch, AP shunt: aorto-pulmonary shunt, ARCAPA: anomalous right coronary artery arising from the pulmonary trunk, ARF: acute renal failure, cdt chg: conduit change, DORV, MS: double-outlet right ventricle, mitral stenosis, F/U: follow-up, ECMO: extracorporeal membrane oxygenation, IAA: interrupted aortic arch (type in parenthesis), LV: left ventricle, RPA: right pulmonary artery, RSV: respiratory syncytial virus, RVOT: right ventricular outflow tract, TOF/PA: tetralogy of Fallot with pulmonary atresia, VSD: ventricular septal defect

used in 16 cases (42%). The mean bypass time, including the period of circulatory arrest, was 108 ± 35 minutes, with a range from 41 to 228 minutes. The mean cross clamp time was 51 ± 21 minutes, with a range from 14 to 98 minutes, and the mean circulatory arrest time was 34 ± 20 minutes, with a range from 6 to 64 minutes.

The approach to the aortopulmonary window was through an aortotomy in 17 cases (45%), an incision through the defect itself in 12 (31%), and through a pulmonary arteriotomy in 9 cases (24%). The defect was closed using a single patch in 30 patients (79%), direct closure was used in 4 patients, and a double patch was used in 3 patients. One patient with a functionally univentricular heart had only narrowing of his aortopulmonary window along with an atrial septectomy. Except for patients with functionally univentricular repairs, all associated anomalies were corrected concomitantly.

Early morbidity and mortality

Notable complications occurred in 7 patients: long-term ventilation in 3, and respiratory syncytial viral

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infection requiring extracorporeal membrane oxygenation, cardiac arrest, acute renal failure requiring peritoneal dialysis and tracheostomy, and severe deep mediastinal wound infection in one patient each.

Early mortality was defined as the in-hospital mortality or death occurring within 30 days of surgery. There were 3 early deaths (7.9%). Two had simple aortopulmonary window, with one death due to a severe pulmonary hypertensive crisis occurring early postoperatively in a 7 year-old, and the other occurring in a premature neonate weighing 700 grams with severe intraparenchymal pulmonary hemorrhage after cardiopulmonary bypass. The patient with a complex aortopulmonary window was a premature newborn with anomalous origin of the right coronary artery from the pulmonary trunk who died of hyaline membrane disease several weeks after repair.

No significant univariate or multivariate risk factors could be identified for early mortality or morbidity.

Long-term survival

There were 2 late deaths: One patient whose aortopulmonary window was closed via the pulmonary trunk using a single patch concomitantly with closure of the ventricular septal defect in 1975 died 2 months after surgery of congestive heart failure. Autopsy showed that the left pulmonary artery was completely occluded by the suture line. A second patient with aortopulmonary window, double-outlet right ventricle, mitral stenosis, and hypoplastic left ventricle, died of a ventricular arrhythmia 17 years after his initial operation. His procedures included narrowing of the aortopulmonary window and atrial septectomy, followed by a classic Glenn shunt, and a Blalock-Taussig shunt.

Overall actuarial survival at 1, 5 and 10 years was 88% (95% confidence intervals from 78 to 98%) (Fig. 1). No significant differences were found for any of the variables studied between the simple and complex anatomical variants. Cox proportional-hazards multivariable regression analysis did not reveal any significant predictors of survival.

Reinterventions

Ten patients required a total of 11 reinterventions, defined as either catheter-based or surgical interventions. Reinterventions related to repair of the aortopulmonary window were necessary in 3 patients. One patient with interruption of the aortic arch had percutaneous dilation of stenotic areas in the ascending aorta and one of the pulmonary arteries 6 months

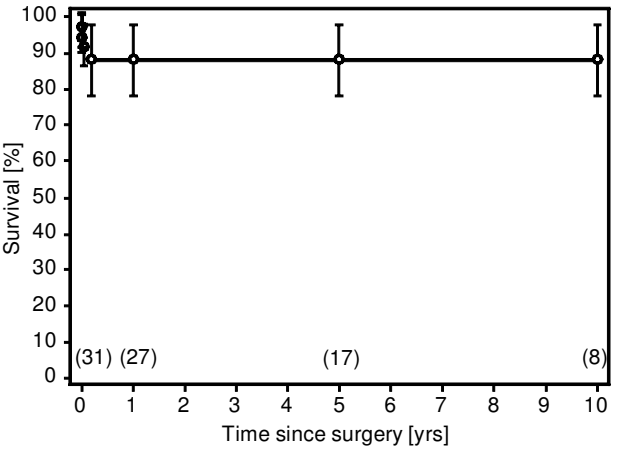

Figure 1.
Kaplan-Meier actuarial survival for all 35 patients with aortopulmonary window after hospital discharge. Error bars indicate 95% confidence intervals based on Greenwood's formula. The estimated cumulative survival at ten years was 88%.

postoperatively, and two patients with repair of simple aortopulmonary window through a pulmonary arteriotomy needed dilation of supravalvar pulmonary stenosis, and of right pulmonary arterial stenosis, at 2 years and 4 years postoperatively, respectively. Reinterventions for associated diagnoses other than aortopulmonary window included right ventricular outflow tract augmentation for tetralogy of Fallot with pulmonary stenosis in 3 patients, conduit changes for tetralogy of Fallot with pulmonary atresia in 2, as well as one Fontan procedure, one Glenn shunt, and one aortopulmonary shunt for double outlet right ventricle with small left ventricle. Freedom from reintervention at 1, 5, and 10 years was 84%, 70% and 70%, respectively (Fig. 2).

By multivariable analysis, aortopulmonary windows with complex associated malformations had a higher likelihood of reintervention ($p = 0.04$), particularly with tetralogy of Fallot ($p = 0.006$). Surgical approach through a pulmonary arteriotomy was also associated with a higher rate of reintervention ($p = 0.01$).

Of the survivors, 25 (76%) had recent echocardiograms performed. These revealed mild stenosis of either the right or left pulmonary arteries in 14, mild stenosis of the aortic arch in 5, mild aortic regurgitation in 2, and mild aortic stenosis in 1.

Discussion

The first successful repair of an aortopulmonary window was performed and reported by Gross in 1952, who used a simple ligature without cardiopulmonary bypass.[7] Since then, surgical management

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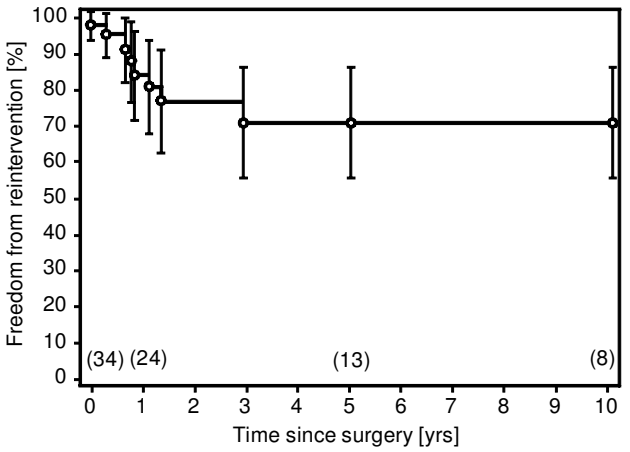

Figure 2.
Kaplan-Meier actuarial freedom from intervention for all 35 patients with aortopulmonary window after hospital discharge. Error bars indicate 95% confidence intervals based on Greenwood's formula. The estimated freedom from reintervention at 10 years was 70%.

has evolved, so that now precise repair is achieved under direct vision using cardiopulmonary bypass. This report, the largest as far as we know published to date, shows that repair of aortopulmonary window can be performed with excellent overall results. As demonstrated in this series, nonetheless, even patients with aortopulmonary window in the absence of major complicating lesions remain at risk if pulmonary vascular disease is present. It is now widely recognized that early repair is essential to prevent gradual development of pulmonary vascular disease,[3,5] and repair of an aortopulmonary window should be performed soon after the diagnosis is established.

Our report also demonstrates that exposure of the defect through a pulmonary arteriotomy should be avoided as it significantly increases the need for reintervention. The ideal type of surgical exposure has been long debated.[3–6] Several authors describe using the transaortic route as their method of choice, but none has been able to show a statistically significant advantage of one technique over another.[3,5,6] The transpulmonary approach was advocated as early as in 1966 in a report from our institution,[8] and was used in one quarter of our patients. Based on the large number of patients included in this series, we were able to show that this approach is an independent risk factor for late reintervention. Narrowing of the pulmonary trunk, or the proximal parts of the right or left pulmonary arteries, was the typical late complication in this and other studies.[6] This can be avoided by exposing the aortopulmonary window through the aorta or the defect itself. When looking at the aortopulmonary window through the aorta, the edges are easily identified. The distance to the orifices of the coronary arteries and the aortic valve is noted, and those structures are carefully avoided.[9] In cases where either the right or left pulmonary artery arises directly from the ascending aorta, a lesion sometimes, albeit erroneously, called hemitruncus, the pulmonary artery has to be detached and reimplanted in the distal pulmonary trunk.

Several authors have described using autologous material only to cover the window. An approach through the defect is usually used, where the flap used to close the defect is created from the "roof" of the defect.[10–13] These authors recommend patching only in special situations, where the defect is more complex or there is an anomalous origin of one coronary artery. Others recommend always to patch the defect.[14] We believe that the ultimate goal is to obliterate the defect while preventing recurrent stenoses or distortion of the great arterial trunks. We would not hesitate, therefore, to use a pericardial or synthetic patch in cases where tension or distortion is likely to occur. We did not find any long-term complications from the use of a patch.

Whereas most series describe worse outcomes for aortopulmonary windows co-existing with major associated lesions,[6] this was not seen in our study. As compared with those having no or simple associated lesions, repair of aortopulmonary window did not result in worse outcome in association with more complex defects. Patients with more complex associated lesions, however, did have a statistically higher need for late reintervention. This is not surprising, since most reoperations were unavoidable, such as the changing of conduits in tetralogy of Fallot with pulmonary atresia or creation of Glenn shunts for functionally univentricular repairs, or predictable, such as augmentation of the right ventricular outflow tract after repair of tetralogy of Fallot. In particular, the high incidence of late obstruction of the aortic arch after concomitant surgery for repair of interruption of the aortic arch reported in another large study[6] was not seen in our experience. Interruption of the aortic arch at the isthmus has been described as occurring more often with aortopulmonary window than the more common interruption between the left common carotid and subclavian arteries,[2,6] a finding confirmed by our study.

Conclusions

Long-term outcomes after repair of aortopulmonary window are excellent. When aortopulmonary window is not complicated by major associated lesions, surgical mortality is related almost exclusively to preexisting pulmonary vascular disease. Repair through a pulmonary arteriotomy results in a significantly

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higher probability of reintervention. We recommend repair through the aorta or through the defect, with placement of a patch if necessary.

References

1. Rowe RD. Aortopulmonary septal defect. In: Keith JD, Rowe RD, Vlad P, eds. Heart Disease in Infancy and Childhood, 3d edition. McMillan, New York, 1978: 215–224.
2. Kutsche LM, Lodenyk HS, Van Mierop. Anatomy and pathogenesis of aortopulmonary septal defect. Am J Cardiol 1987; 59: 443–447.
3. Tkebuchava M, von Segesser LK, Vogt PR, Bauersfeld U, Jenni R, Kunzli A, Lachat M, Turina M. Congenital aortopulmonary window: diagnosis, surgical technique and long-term results. Eur J Cardiothor Surg 1997; 11: 293–297.
4. Bertolini A, Dalmonte P, Bava GL, Moretti R, Carvo G, Marasini M. Aortopulmonary septal defects: a review of the literature and report of ten cases. J Cardiovas Surg 1994; 35: 207–213.
5. Van Son J, Puga FJ, Danielson GK, Sewrad JB, Mair DD, Schaff HV, Ilstrup DM. Aortopulmonary window: factors associated with early and late success after surgical treatment. Mayo Clin Proc 1993; 68: 128–133.
6. McElhinney DB, Reddy VM, Tworetzky W, Silverman NH, Hanley FL. Early and late results after repair of aortopulmonary septal defect and associated anomalies in infants < 6 months of age. Am J Cardiol 1998; 81: 195–201.
7. Gross RE. Surgical closure of an aortic septal defect. Circulation 1952; 5: 858–863.
8. Putnam TL, Gross RE. Surgical management of aortopulmonary fenestration. Surgery 1966; 59: 727–735.
9. Deverall PB, Lincoln C, Aberdeen E, Bonham-Carter RE, Waterston DJ. Aortopulmonary window. J Thorac Cardiovasc Surg 1969; 57: 479–486.
10. Shatapathi P, Rao KM, Krishnan KV. Closure of aortopulmonary septal defect. J Thorac Cardiovasc Surg 1987; 93: 789–791.
11. Di Bella I, Gladstone DJ. Surgical management of aortopulmonary window. 1998; 65: 768–770.
12. Kitagawa T, Katoh I, Taki H, Wakisaka Y, Egawa Y, Takahashi Y, Akita H, Matsuoka S. New operative method for distal aortopulmonary septal defect. Ann Thorac Surg 1991; 51: 680–682.
13. Johansson L, Michaelsson M, Wasterholm CJ, Aberg T. Aortopulmonary window: a new operative approach. Ann Thorac Surg 1978; 25: 564–567.
14. Tiraboschi R, Salomone G, Crupi G, Manasse E, Salim A, Carminati M, Vanini V, Parenzan L. Aortopulmonary window in the first year of life: report on 11 surgical cases. Ann Thorac Surg 1988; 46: 438–441.

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