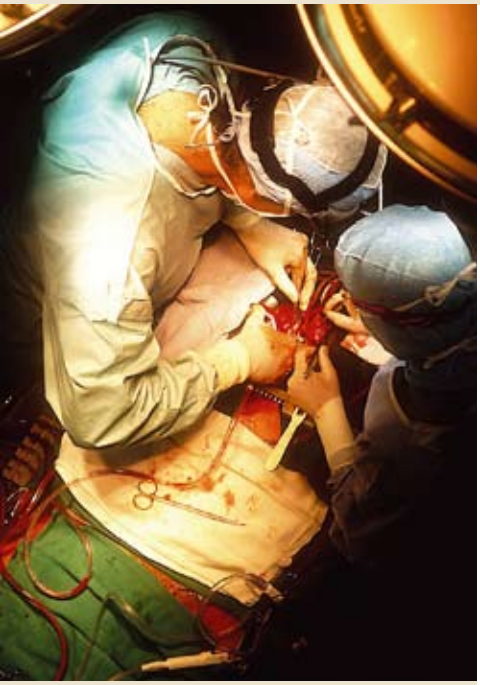
Fifteen years of experience in repair of aortopulmonary window in children



Nader Givtaj MD.,Mohammad Sadegh Pour Abbasi MD.* Shahyad Salehi MD.,Ramin Baghaee MD.

Abstract

Introduction:

Aorto-pulmonary window is a very rare Malformation which accounts for about 0.15% of all cardiac anomalies. There is no tendency for AP windows to close spontaneously. The natural history of infants with large AP windows is as unfavorable rarely they survive to childhood and those who survive beyond early life have important pulmonary vascular disease (1). The Richardson classification system for aorto-pulmonary window includes simple defects between the ascending aorta and pulmonary trunk (type I), defects extending distally to include the origin of the right main pulmonary artery (type II), and anomalous origin of the right main pulmonary artery from the ascending aorta with no other aorto-pulmonary communication (type III).

Material and Methods:

We reviewed our cases of aorto-pulmonary window who underwent surgical repair From 1992 to 2007 at Sahid Rajee Heat center, Tehran, Iran. There were 30 children with male to female ratio of 2:1 .We evaluated demographic information of the patients.We used different operative techniques . The approach for AP Window repair was ligation without CPB in two cases, division and suturing using CPB in one patient, trans-window in 17 (PTFE patch in16; Dacron in 1), trans-aortic in 9 (in 8 PTFE patch, in one simple suturing) and trans-pulmonary in 2 (both with PTFE)

Among 15 patients with associated cardiac anomalies, 13 (87%) underwent single stage repair with the IAA repair was the most common (2 cases) .Factors such as cross-clamp and bypass time, mortality, early and late morbidity, ICU stay, hospital stay, duration of ventilator support were compared between groups with various methods of repair using “SPSS 16”.

Resullts:

In among 30 patients male to female ratio was 2:1 .Mean age of the opatients was 28± 9 months ;range 2-90 months,weight 8.6±4.6;range 2-17 kg . Most patients (73%) had sub-systemic or systemic pulmonary hypertension (23% and 50% respectively) .Morphology of Ap windows was type I (87%, n=26), type II (10%, n=3) distal and one type III (3%), This agree with other reportsres. 63% of patients were symptomatic, most commonly dyspnea and 43% were in heart failure on admission. Preoperative EF was 0.66 ± 0.07 which increased to 0.75 ± 0.07 post-operatively apparently duo to elimination of shunt and resultant increase in forward flow of aorta. Preoperative pulmonary artery pressure was 63 ± 13 mm Hg. 19 (63%) of patients had associated cardiac anomalies most frequently various forms of Aortic stenosis (23%) fallowed by interrupted aortic arch (IAA), coronary anomalies and VSD (each about 7%). In most other series IAA has been most common associated anomaly (6), (7). The overall in-hospital mortality was 10% (3 patients), two of whom had associated anomaly, one had undergone arterial switch operation for TGA who couldn’t be separated from CPB, and the other one had simultaneous repair for interrupted aortic arch. The mortality was no different among patients with or without associated anomaly (10% each). Among the 27 survivors, the mean ICU stay was 4.4 days (range 3 - 12 days) and the mean post-operative hospital stay was 10.7 days (range 7 - 16 days). Early complications were bleeding (two cases), pneumonia (one) and CVA (one). Mean follow-up was 49 months (range 2 – 280 months) and there was no re-operation or late death. There were 4 cases of residual AP Window detected by echocardiography; none of them required re-intervention. Among patients with residual AP Window

*Corresponding Author Rajaee Heart center Iran University of Medical sciences Tehran Iran E mail:drmpourabbasi@yahoo.com

two cases were seen with banding technique (100%) one with trans-aortic patch repair (11%) and one case with trans-window patch repair (5%).

Discussion:

An aorto-pulmonary window is a communication between the pulmonary artery (PA) and the ascending aorta in the presence of two separate semilunar valves. There is no tendency for AP Windows to close spontaneously [8] Since the early 1990s, diagnosis has relied exclusively on two-dimensional echocardiography [9] but type II and III lesions are difficult to differentiate from PDA [10]. Cardiac catheterization and cineangiography with retrograde aortography is done in infants 6 months of age for evaluating the presence of irreversible pulmonary vascular disease and in case of complex APW for delineating the
exact morphology. In our series in 27% (8) of patients the diagnosis was made only through echocardiography, remainder of the cases was diagnosed via both echocardiography and angiography.Using multivariate analysis, we assessed the effects of (1) patient related factors (age, sex, weight and type of aorto-pulmonary window) and (2) procedural factors (type of approach) on post-operative course (ICU stay, Post-operative hospital stay, duration of ventilator support and post-operative EF). Age ,sex and weight had no clear impact on post operative course. As mentioned earlier the overall in-hospital mortality was 10% (3 patients). The reported mortality among other series ranged from 7.6% to 27%).The mortality was no different among patients with or without associated anomaly (3% each). Also there was no difference among various methods of repair in respect of morbidity, ICU stay; ventilator support and post operative EF .

Conclusion:

In our view Trans aortic or transwindow repair is the procedure of choice for APW repair and, simple ligation without CPB should be avoided due to the possibility of residual APW and distortion of pulmonary artery.

Key words: Aortopulmonary window,Transaortic repair,Cardiopulmonary bypass

Introduction:

An aorto-pulmonary window is a communication between the pulmonary artery (PA) and the ascending aorta in the presence of two separate semilunar valves, the later characteristic distinguishes it from persistent truncus arteriosus. It is also called aorto-pulmonary fistula, aortic septal defect, aorticopulmonary septal defect, and aorto-pulmonary fenestration. APW accounts for 0.2% to 0.6% of all cases of congenital heart diseases (1). Elliotson (1830) was first to describe a patient with AP window.

The current classification for APW proposed by members of the STS-Congenital Heart Surgery Database Committee and representatives from the European Association for Cardiothoracic Surgery involves the terms proximal, distal, total, and intermediate defects (2) figure 1.

- **AP window, NOS**
  An AP window not further described (not otherwise specified [NOS]).
- **AP window and interrupted aortic arch**
  An AP window in association with an interrupted aortic arch 3, 4. The AP window and interrupted aortic arch may then be coded separately to further specify the individual AP window and interrupted aortic arch types.
- **AP window, type 1 proximal defect**
  An AP window located just above the sinus of Valsalva, a few millimeters above the semilunar valves, with a superior rim but little inferior rim separating the AP window from the semilunar valves.
- **AP window, type 2 distal defect**
  An AP window located in the uppermost portion of the ascending aorta, with a well-formed inferior rim but little superior rim.
- **AP window, type 3 total defect**
  An AP window involving the majority of the ascending aorta, with little superior and inferior rims.
- **AP window, intermediate type**

An AP window similar to the total defect but with adequate superior and inferior rims.

A simple APW is defined as having no associated cardiac defects, or simple defects requiring no or only minor intervention, such as patency of the ductus arteriosus, right aortic arch, or atrial septal defect. A complex APW, on the other hand, has one or more associated complex cardiac malformations or anomalies requiring more complex repair, such as interrupted aortic arch or ventricular septal defect.

There is no tendency for AP Windows to close spontaneously (5). The defect is variable in size, but all defects result in a large, generally continuous left-to-right shunt when the pulmonary vascular resistance falls, similar to other interarterial communications such as patent ductus arteriosus or truncus arteriosus. Without corrective surgery, irreversible obstructive changes in the pulmonary vascular bed develop early, followed by death in the second decade, although patients surviving into the fourth decade have been reported (6). Even there is an interesting report of a patient with Eisenmenger syndrome secondary to a large unrepaired AP window who gave birth to three children in her thirties, survived into her fifties with relatively preserved quality of life, and died at age 60 (7).

Patients and Methods:

From 1992 to 2007, 30 patients have undergone APW repair at Saheed Rajee Heat center, Tehran, Iran. Male to female ratio was 2:1. Age at operation ranged from 2 months to 7.5 years (median 1.8 years). Follow up ranged from 2 months to 12 years (median 2.4 years). Mean preoperative pulmonary artery pressure was ranged from 40 to 90 mmHg. 10 patients (30%) had complex Aorto-pulmonary window. Morphology of APW was Proximal (90%, n=27), distal (10%, n=3). 63% of patients were symptomatic, most commonly dyspnea and 43% were in heart failure on admission. Surgical approaches were trans-window in 17 (56.7%), trans-aortic in 9 (30%) and trans-pulmonary in 2 (6.6%) patients. Two patients underwent simple banding of APW without CPB.

The technique for AP Window repair was division and primary suturing without patch using CPB in one patient, trans-window (sandwich patch) in 17 (Poly Tetra Fleur Carbon "PTFE" patch in 16; Dacron in 1), trans-aortic in 9 and trans-pulmonary in 2 (both with PTFE patch).

Associated cardiac anomalies are shown in table 1 and compared with data from children`s memorial hospital. Among 12 patients with associated cardiac anomalies, 10 (83.3%) underwent single stage repair with the Interrupted Aortic arch (IAA) repair was the most common (2 cases) table 2.

Table 2: *simultaneous repair of the associated lesions in patients with aortopulmonary window. (VSD: Ventricular Septal Defect, PDA: Patent Ductus Arteriosus, IAA: Interrupted Aortic arch, ASD: Arterial Septal Defect, AV Aortic Valve, RPA: Right Pulmonary Artery, LPA Pulmonary Artery, PFO: Patent Foramen Ovale, RCA Right Coronary Artery)*

	Frequency	Percent
IAA Repair	2	6.7
Arterial switch Repair	1	3.3
AV Comissurotomy	1	3.3
LPA Repair	1	3.3
PFO Closure	1	3.3
RCA bypass graft	1	3.3
RCA Transfer	1	3.3
RPA Transfer	1	3.3
VSD Closure	1	3.3
Total	10	3.3

Results:

The overall in-hospital mortality was 10% (3 patients), two of whom had associated anomaly, one had undergone arterial switch operation for TGA who couldn't be separated from CPB, and the other one had simultaneous repair for interrupted aortic arch. Neither age at operation (p=0.21) nor weight (p=0.22) had impact on mortality. The mortality although higher in patients with complex lesions (1 of 20 or 5% in simple APW, 2 of 10 or 12.5 % in Complex APW, P= 0.21) the difference was not statistically significant. Among the 27 survivors, the mean ICU stay was 4.4 days (range 3 - 12 days) and the mean post-operative hospital stay was 10.7 days (range 7 - 16 days). Preoperative pulmonary arterial pressure, age and weight of the patients ha no effects on post operative hospital stay or ICU stay (p> 0.1). Also we compared our results in terms of cardiopulmonary bypass time, cross clamp time; age and weight of our patients with the best results belong to European congenital data base (table 3). As it is seen our patients are significantly older (mean 27.7 months) than the reference database (mean 7.5 months). Our pump times (mean 67.7 vs. 88.7 minutes) and cross clamp times (mean 28.9 vs. 39 minutes) were some what lower. More detailed individual data are shown in table 4.

Early complications were bleeding (two cases), pneumonia (one) and CVA (one). There were 4 cases of residual AP Window detected by echocardiography; Among patients with residual APW two cases were seen with banding technique without Cardio-pulmonary Bypass (CPB) (100%), while only 7.1% of cases (2 of 28) who underwent repair using Cardio-pulmonary Bypass had residual defect; P=0.001).

Discussion:

Current diagnostic techniques emphasize noninvasive echocardiographic assessment (figure 1) and in most patients there is no longer a need for cardiac catheterization [8] but type II and III lesions are difficult to differentiate from PDA (figure 2) [9]. In Infants 6 months of age cardiac catheterization is done for evaluating the presence of irreversible pulmonary vascular disease and in case of complex APW for delineating the exact morphology.

All children are now repaired as neonates. This has improved the results with regard to complications from pulmonary hypertension.

Over time the surgical repair of APW witnessed continuous evolution of techniques that reflect the advances in pediatric cardiac surgery; major points are listed below:

1952: (Gross) closed approach (left thoracotomy) (10)

1957: (Cooley) division of the APW between clamps using cardiopulmonary bypass (11) while the patient was on CPB.

1964: (Bjork) trans-pulmonary using patch

1968: (Wright et al) trans-aortic approach by direct suture.

1969: (Deverall et al) the first trans-aortic closure with a patch (12).

1978: (Johansson et al) Sandwich type closure

1991: (kitagawa et al) rerouting the pulmonary trunk for distal aorto-pulmonary window defects. (13)

1994: (messmer) pulmonary artery flap to close the window and pericardial patch to close the pulmonary trunk (14)

1998: (Di Bella & Gladstone) only pulmonary artery flap to close the window (15)

Our approaches were included trans-aortic (n=9), trans-window technique division and patch closure using a 'sandwich' technique (n=17) and trans-pulmonary (n=2) with equally good results, Most centers now recommend trans-aortic patch closure for most patients with an APW [16, 17, 18]. Earlier in our series we had two cases of banding of window, both had residual shunt on fallow up, so it can not be recommended. Other techniques have been described including division using the PA to patch the aorta, reconstructing the PA with pericardium [21, 22]. There is a known association between interrupted aortic (IAA) arch an APW. 10 to 20% of patients with the diagnosis of APW have associated IAA. Conversely about 4% of patients with the diagnosis of IAA have APW (19). The type of IAA nearly always is type A (19) , (9). In our series we had 2 cases of IAA (7%). It is now recommended to repair both condition in the same time preferably at the neonatal period.

Conclusion:

Echocardiography is sufficient for the diagnosis and planning the operation for APW. Complete correction of aortopulmonary window in neonatal period is now recommended to avoid complications related to pulmonary hypertension. In the presence of interrupted aortic arch both lesion is best repaired at the same occasion. The preferred technique is trans aortic patch closure to avoid injury to the coronary arteries, aortic valve leaflets, and PA orifices and to allow for adequate growth of both the aorta and PA structures.

Figures & Tables:

***Figure 1**: Echocardiographic view of an aortopulmonary window (asc = ascending aorta, Ao-pulm window = aortopulmonary window, pulm artery = pulmonary artery, desc. Ao = descending aorta)*

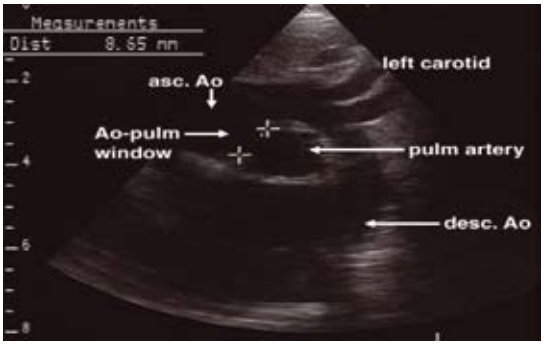


***Figure 2** types of aortopulmonary window*

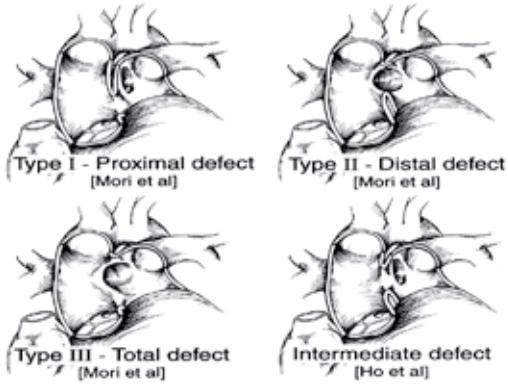


***Table 1**: Associated lesions in children with aorto-pulmonary window, comparison between two centers.*

Associated Anomalies	Shahid Rajaee Heart Center (1992-2007)	Children`s Memorial Hospital (1961-2001)
Interrupted aortic arch	2	4
Ventricular septal defect	2	3
Patent ductus arteriosus	2	-
Coronary Anomaly	2	-
Right pulmonary from Aorta	1	4
Arterial septal defect	1	1
Transposition of great arteries	1	1
Left pulmonary Stenosis	1	-
Tetralogy of fallot	-	1
Total	12 (of 30)	14 (of 22)

***Table 3:** comparison between Values for the 5 best hospitals in the selected group (parenthesis) and our series. NA: not available.*

	No of cases	Minimum	Maximum	Mean	Std. Deviation
Age (Month)	30 (74)	2 (0.1)	90 (96)	27.7 (7.5)	23.7 (17.3)
Weight (kg)	30 (73)	2 (1)	17 (20)	8.6 (4.7)	4.6 (3.4)
Aortic X Time (min)	27 (61)	12 (9)	55 (140)	28.9 (39)	8.6 (22.4)
CPB Time (min)	27 (65)	32 (19)	99 (268)	67.7 (88.7)	19.7 (48.9)
Vent Support (day)	29 (NA)	1 (NA)	7 (NA)	2.2 (NA)	1.6 (NA)
ICU Stay (day)	29 (NA)	3 (NA)	12 (NA)	4.4 (NA)	2.2 (NA)
Hosp Stay (day)	27 (NA)	15 (NA)	35 (NA)	20.8 (NA)	3.8 (NA)

No	Age	Sex	Weight	CoAnomaly Type	XTime	CPBTime	Repair	Approach	repair Type	ICUStay	Vent Support	Mortality	Morbidity Type	Follow
1	2	Female	4	LPA Stenosis	20	40	PTFE	Trans Aortic	LPA Repair	7	3	No	No Morbidity	2
2	3	Male	4		25	60	PTFE	Trans Window		3	1	No	No Morbidity	9
3	3	Female	4		26	70	PTFE	Trans Pulmonary		3	3	Yes	Bleeding	-
4	4	Male	5	ASD PDA	28	60	PTFE	Trans Window	PFO Closure	7	2	No	No Morbidity	18
5	4	Male	5	RPA from Aorta	35	85	Simple Closure	Trans Aortic	RPA Transfer	3	1	No	No Morbidity	5
6	4	Male	5	IAA			PTFE	Trans Aortic	IAA Repair			Yes	No Morbidity	-
7	5	Male	4	Coronary Anomaly	55	88	PTFE	Trans Window	RCA bypass graft	4	1	No	Pneumonia	3
8	7	Female	6		25	80	PTFE	Trans Pulmonary		7	3	No	Bleeding	120
9	8	Male	5		35	63	PTFE	Trans Aortic		3	1	No	No Morbidity	36
10	10	Male	6	AS	24	40	PTFE	Trans Window		3	2	No	No Morbidity	.
11	11	Male	6		18	44	PTFE	Trans Window		3	1	No	No Morbidity	4
12	12	Male	6	TGA	25	90	PTFE	Trans Window	Arterial switch	3	3	Yes	No Morbidity	-
13	17	Female	4	AS, PDA	25	84	PTFE	Trans Window		3	1	No	No Morbidity	12
14	18	Female	7	AS	43	61	PTFE	Trans Window		6	2	No	No Morbidity	11
15	18	Female	5	VSD	28	89	PTFE	Trans Window		4	2	No	No Morbidity	38
16	24	Male	12	Coronary Anomaly	43	72	PTFE	Trans Window	RCA Transfer	5	2	No	No Morbidity	72
17	30	Male	8		30	90	PTFE	Trans Aortic		3	1	No	No Morbidity	90
18	30	Male	10		20	70	PTFE	Trans Aortic		5	5	No	No Morbidity	126
19	36	Male	10	AS			Banding	Trans Window		4	2	No	No Morbidity	3
20	36	Male	10				Banding	Trans Window		3	1	No	No Morbidity	2
21	36	Male	15	PDA	12	40	PTFE	Trans Window		4	2	No	No Morbidity	.
22	36	Male	2	AS	27	59	PTFE	Trans Aortic		3	2	No	No Morbidity	60
23	44	Male	12	IAA	35	58	PTFE	Trans Window		4	1	No	No Morbidity	72
24	44	Male	12	AS	32	99	PTFE	Trans Aortic		12	7	No	CVA, Arrest	18
25	48	Male	10		30	60	PTFE	Trans Window		4	1	No	No Morbidity	108
26	60	Female	16		25	92	PTFE	Trans Window		3	2	No	No Morbidity	38
27	60	Female	16	AS	30	92	PTFE	Trans Window		3	2	No	No Morbidity	36
28	60	Female	14	AS	22	32	Dacron	Trans Window		9	7	No	No Morbidity	.
29	72	Male	17	VSD	31	41	PTFE	Trans Window	AV Comissurotomy	3	1	No	No Morbidity	34
30	90	Female	17		30	70	PTFE	Trans Aortic	VSD Closure	4	2	No	No Morbidity	24

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