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Research Paper

Surgical Management of Aortopulmonary Window: 7 years of Experience of a Single Cardiac Centre.

Mohammad Rokonujjaman[1] , Shaheedul Islam[2], Nusrat Ghafoor[3] , Syed Tanvir Ahmad[4], Ibrahim Khalilullah [5],Abdullah Al Shoyeb[6], Atiqur Rahman[7], Golam Saklayeen[8]

1. *Associate Professor & Consultant, Dept. of Cardiac surgery, Ibrahim Cardiac Hospital & Research Institute.*
2. *Associate Professor & Consultant, Dept. of Cardiology, Ibrahim Cardiac Hospital & Research Institute.*
3. *Associate Professor & Consultant, Dept. of Radiology, Ibrahim Cardiac Hospital & Research Institute.*
4. *Assistant Professor & Associate Consultant, Dept. of Cardiac surgery, Ibrahim Cardiac Hospital & Research Institute.*
5. *Assistant Professor & Associate Consultant, Dept. of Cardiac Anesthesiology, Ibrahim Cardiac Hospital & Research Institute.*
6. *Registrar & Specialist, Dept. of Cardiac surgery, Ibrahim Cardiac Hospital & Research Institute*
7. *Registrar & Specialist, Dept. of Cardiac surgery, Ibrahim Cardiac Hospital & Research Institute*
8. *8.Assistant registrar, Dept. of Cardiac surgery, Ibrahim Cardiac Hospital & Research Institute*

Address of correspondence: *Dr. Mohammad Rokonujjaman.*
MBBS, MS(CV&TS), MRCPS(Glasgow).Associate Professor & Consultant, Dept. of Cardiac surgery, Ibrahim Cardiac Hospital & Research Institute,

ABSTRACT
BACKGROUND: *Aortopulmonary window (APW) is a rare congenital anomaly and it represents 0.2–0.3% of all congenital heart lesions. Increasing pulmonary arterial hypertension and its consequences are the main concern of this disease.*
OBJECTIVE: *As a small scale centre, we tried to share our 7years of surgical experience in managing a very small series of 5 cases of aortopulmonary window.*
METHODS: *This retrospective study was done from December 2013 to January 2021. 5 patients with aortopulmonary window who underwent successful surgical repair are the study subjects. Their age ranged from 4 months to 6 years (mean 2.26±1.6) yrs. An initial diagnosis was obtained from 2D echocardiography, which showed echo dropout in the parasternal short-axis view. There were no associated cardiac anomalies. CT aortopulmonary angiographies were performed in all patients. All the patients had type 1 variety of AP window and surgical management was done by opening the aorta.*
RESULTS: *There were no early or late deaths. There was no pulmonary hypertensive crisis. All patients underwent echocardiography before discharge; none showed a residual shunt. Every case received afterload reducing agent and diuretics on follow-up. All patients were followed up at intervals of 3 months, with the longest follow-up being 3 years. At follow-up, all patients were NYHA I. The mean RVSP on echocardiography was 32 mmHg at 3 months.*
CONCLUSIONS: *Operative repair should be offered as soon as the diagnosis of AP window has been established, regardless of the patient's age. Various surgical techniques can be applied depending on the size of the communication. Early and long-term outcomes after surgical correction are excellent regardless of age or pulmonary vascular resistance.*
KEYWORDS: *Aortopulmonary window • Pulmonary arterial hypertension*

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I. INTRODUCTION

Aortopulmonary window (APW) is a rare, surgically correctable congenital cardiac anomaly. This lesion represents between 0.2% and 0.3% of all congenital cardiac lesions.[1,2] It was first described by Eliotson in 1830.[3]

Subsequent classification was done by Mori in 1978[4] and by Richardson in 1979.[5] This lesion is associated with rapid progression of pulmonary arterial hypertension (PAH) unless it is corrected surgically. We would like to share 7 years of surgical experience in managing a spectrum of 5 cases.

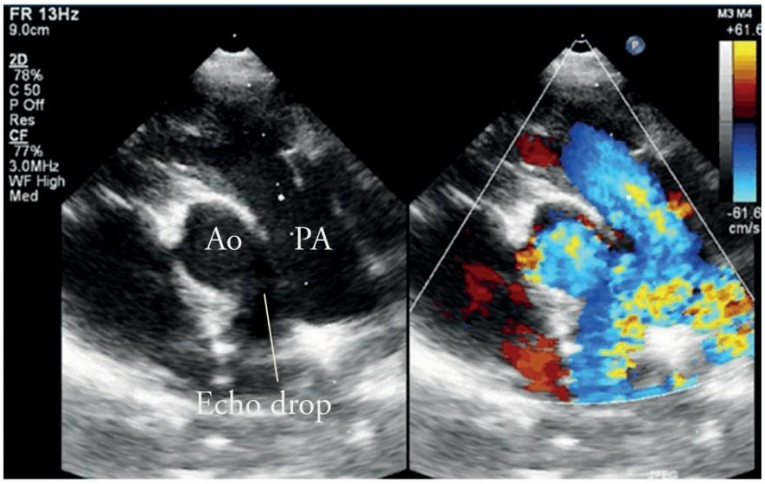
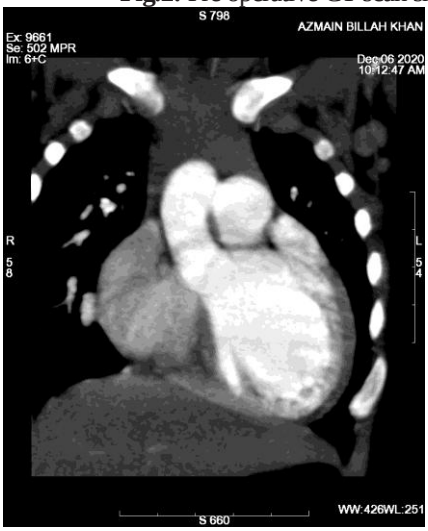


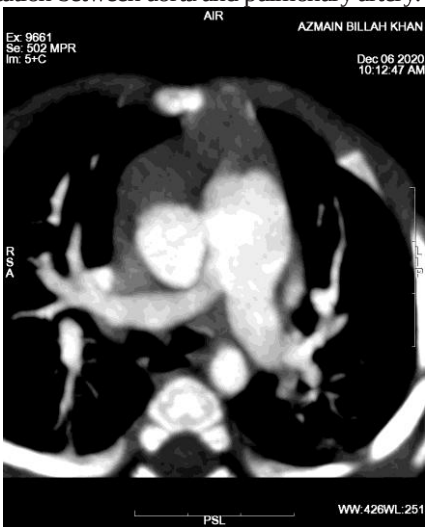
Figure 1: 2D echocardiography in modified short-axis view shows echo dropout between aorta and pulmonary artery. Ao: aorta; PA: pulmonary artery.

II. METHODS

This retrospective study of 5 patients with APW who underwent successful surgical repair was carried out from December 2013 to January 2021. Ages ranged from 4 months to 6 years (mean of 2.26±1.6 years). There were 3 male subjects and 2

Fig.2: Pre operative CT scan showing communication between aorta and pulmonary artery.



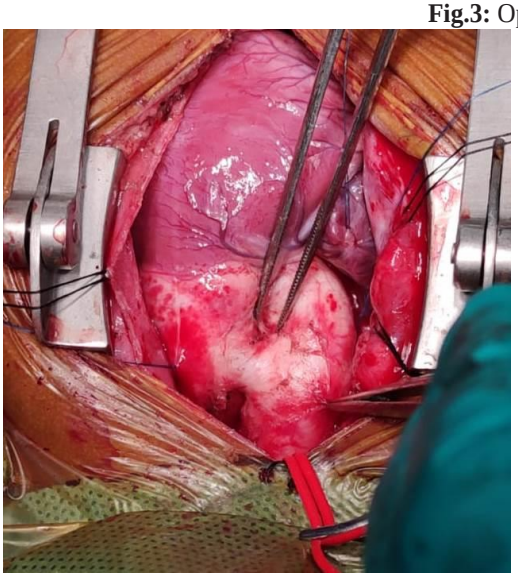


female subjects with a male:female ratio of 3:2. The mean weight at operation was 9.4±3.4 kg. Clinical presentation varied from recurrent respiratory tract infection, dyspnoea on exertion, palpitations, and failure to thrive. There were no associated cardiac anomalies except patent foramen ovale in two cases. An initial diagnosis was obtained by 2D echocardiography, which showed echo dropout in the parasternal short-axis view (Fig.1). CT aoto-pulmonary angiography was performed in all 5 patients and cardiac catheterization in 2 patients. All the patients had Type I defect. The mean right ventricular systolic pressure (RVSP) was 94 ± 2 mmHg.

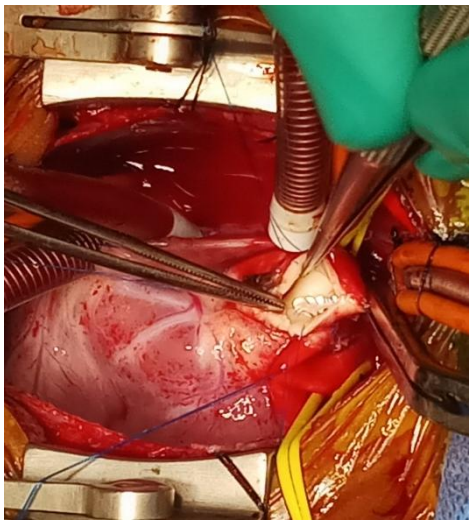
SURGICAL TECHNIQUE:

Patients were operated through a median sternotomy. After the anatomical features were visualized, a decision regarding the surgical approach was made. Aortotomy was performed to close the defect with 0.4 mm PTFE patch from aortic side in all patients. Standard CPB was used to close the defect. Temperature reduced to 32^0C. Antegrade cold blood Delnido Cardioplegia solution used in all cases. No patients required circulatory arrest. PFO kept open in two cases.

Fig.3: Operative picture of APW



Per operative view of AP window



AP window after patch closure

The mean cross-clamp time was 46 ± 0.29min, and the mean pump time was 76 ± 0.78 min. Inotropic support used for 48 h. None of the patients developed a pulmonary hypertensive crisis. The mean duration of mechanical ventilation was 16 ± 0.2 h. The mean intensive care unit stay was 2 ± 0.16 days and the total post- operative hospital stay was 7 ± 0.38 days.

Written informed consent from all patients and approval from our hospital ethics committee were obtained prior to preparing this manuscript.

III. RESULTS

There were no early or late deaths. All patients underwent echocardiography before discharge; no patient had evidence of a residual shunt. Mild left ventricular dysfunction was seen in 2 patients that regressed with afterload reduction and diuretics on follow-up. All patients were followed up at intervals of 3 months with the longest follow-up being 3 years. At follow-up, all patients were in NYHA class 1. The mean RVSP on 2D echocardiography was 32mmHg at 3 months follow-up.

IV. DISCUSSION

APW is an uncommon congenital cardiac anomaly. It has haemodynamic features that are similar to those of a large PDA, even more so to a common truncus arteriosus. Unlike a common truncus arteriosus, APW has well defined aortic and pulmonary valve apparatuses. Embryologically, APW is explained by incomplete fusion or malformation of the right or left conotruncal rings between the fifth and eighth weeks of intrauterine life. Normally, conotruncal septation completes between

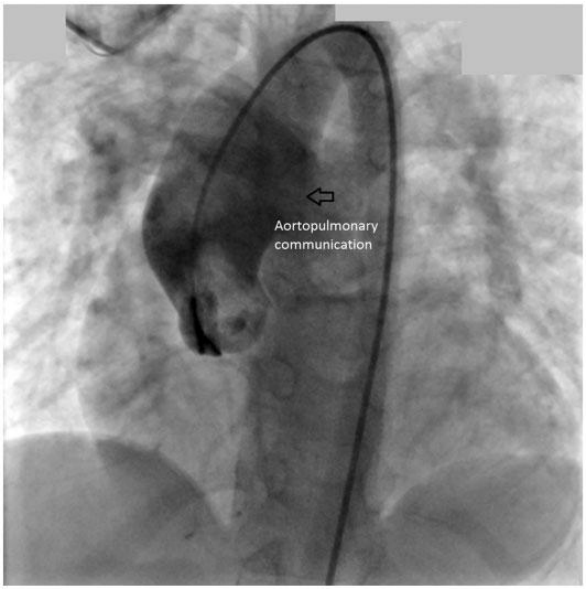


Figure 4: Catheterization study (catheter course from descending aorta to arch and ascending aorta) showing aortopulmonary communication.

5th to 8th weeks of intrauterine life.[8] APW can result from incomplete fusion (Type I), poor alignment (Type II) or total absence (Type III) of the right and left conotruncal cushions. The clinical presentation is an excessive left-to-right shunt, and most patient presents early in life. Development of pulmonary hypertension and pulmonary vascular resistance is usually rapid. The initial diagnosis can generally be easily achieved by transthoracic echocardiography, where one sees echo dropout in the parasternal short-axis view. Early diagnosis and surgical intervention in all patients are essential because they could develop PAH rapidly. CT angiography usually performed in all patients. Of the untreated patients, 40% die within the first year of life.[9] Untreated patients rapidly develop pulmonary vascular obstructive disease. An operation should be offered to all patients as soon as the diagnosis is confirmed. APW is frequently found as an isolated lesion but can be associated with various other congenital cardiac anomalies.[10-12] In our study, 2 (40%) patients had associated patent foramen ovale (PFO) only, with isolated APW in the rest. Surgical options are tailored to the anatomy of the APW.

Commonly, the defect is large, with a significant left- to-right shunt, resulting in congestive heart failure, pulmonary hypertension and early development of pulmonary vascular obstructive disease. In some patients, if the anatomy is suitable, the operation can be done through an anterolateral thoracotomy without the use of cardiopulmonary bypass even if the repair has to be combined with ligation of a PDA, a pulmonary artery banding or correction of an interrupted aortic arch (IAA).[9] In our institution, the preferred approach was a median sternotomy. Sandwitch patch closure technique (Johansson *et AL.*) was used in all cases.[15-18] PFO left open in both the cases. Two patients had pre-operative mild to moderate mitral regurgitation which improved to trivial in one year follow-up. Internationally published studies have correlated IAA, severe PAH and low-cardiac output as factors associated with mortality.[20-22] But in our series, we had only type 1 variety and resulted in a good result. We did not have any early or late deaths.

Our strategy was to extubate patients as early as possible. The mean period of mechanical ventilation was 16 ± 0.2 hours. The possible occurrence of a pulmonary hypertensive crisis should be kept in mind while managing these children in the postoperative period. In our series, none had a postoperative pulmonary arterial hypertensive crisis. Our other management strategies were adequate pain control with intravenous analgesics in all patients, nitroglycerine infusion for all patients apart from inotropic supports, early mobilization, and diuretics (furosemide 0.5–1mg/kg) and ACEI in the post-operative period.

The mean intensive care unit stay was 2 ± 0.16 days and the hospital stay was 7 ± 0.38 days. Though the published literature is against the late closure of APW (beyond 2 years), in our study we still achieved a death-free series operating on a wide range of patients, even beyond 2 years of life.

All patients underwent echocardiography before discharge. The scans showed no residual shunt in any patient and a reduction in the RVSP. We use RVSP as a measure for calculating PAH postoperatively: RVSP <35 mmHg, marked as no PAH; ≥35–45, marked as mild PAH; ≥45–60 mmHg, marked as moderate PAH and >60 mmHg, marked as severe PAH. One patient had moderate PAH postoperatively and required sildenafil. Our policy was to use sildenafil and ACEI for at least 3 months postoperatively until it becomes normal. Their early and late follow-up examinations showed excellent outcomes with symptomatic improvement and reduction in pulmonary artery pressures. Our study showed that PAH developed irrespective of age at operation and the long-term outcome

following surgery was excellent.

V. CONCLUSION

APW is a rare but well-defined anomaly. This study is one of the largest series from a single institution, collected over a period of 7 years. Unless there is a right-to-left shunt despite oxygen administration, no patients with APW should be denied the operation. The results of surgical treatment of APW demonstrate low risk, even if the APW is associated with major cardiac anomalies. Surgical options are tailored to the anatomy and the size of the defect. Whenever possible, continuity ligation, division and suture should be done with cardiopulmonary bypass on standby. The possible occurrence of a pulmonary hypertensive crisis should be kept in mind when one is managing these children in the postoperative period. In our series, no patient had a postoperative pulmonary arterial hypertensive crisis. The published literature relates IAA, severe PAH and low-cardiac output to the risk of death, but in our series, though we encountered all of these conditions, we did not have early or late deaths. Long-term outcome after operative correction was excellent regardless of age or pulmonary vascular resistance.

Conflicts of interest: none declared.

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