Case Report

Trans- catheter Closure of Large Tubular PDA Followed by Post Device Closure Syndrome: A case study

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ABSTRACT

Patent ductus arteriosus (PDA) is the commonest congenital heart disease. PDA can be closed by trans-catheter device implantation or surgical closure. Pediatric cardiologists have been closing the PDA with device with tremendous success. Although large PDA device closure followed by post device closure syndrome is rare, this clinical condition can be managed by pre device implantation evaluation of predictor factors. Our patient had large tubular PDA which was properly evaluated before ADO I device implantation and managed his post device closure syndrome evidenced by left ventricular dysfunction and sudden rise of systemic blood pressure. His post device closure syndrome and para device leak were properly managed during hospital stay and close follow up.

Keywords: *Large tubular PDA, Post device closure syndrome, Left ventricular dysfunction, Para device leak*

CASE SUMMARY

A 2-years 1-month-old baby boy weighing 7.8 kg (<3rd centile) was admitted in our Pediatric Intensive Care Unit (PICU) with the diagnosis of large tubular patent ductus arteriosus (PDA, 8 mm), L-R shunt, PDA gradient max PG 45/6 mmHg with severe pulmonary arterial hypertension, and severe pneumonia with heart failure. He was diagnosed as a case of PDA at the age of 5 months in our OPD and was planned for device closure of PDA. But due to COVID-19 pandemic situation he could not come to us. However, he had history of repeated lower respiratory tract infection (LRTI) for which he was admitted in local hospital and managed by intravenous antibiotics. Pediatrician excluded tuberculosis and infective endocarditis as well. He had history of not growing well in comparison with other peers for last few months. However, he was on diuretics, digoxin, ACEI and tab sildenafil with poor compliance of medication.

On admission in our PICU, he was conscious, cooperative, afebrile, his weight was 7.8 kg (<3rd centile), height 80 cm (<3rd centile), febrile (990F), with tachycardia (heart rate 160 b/m), tachypnea (60 b/m), blood pressure 90/37 mmHg, SPO2 98% in room air. Examination of cardiovascular system (CVS) revealed hyperdynamic precordium with apex beat on left 5th ICS,1st and 2nd heart sounds were audible in all 4 areas with continuous machinery murmur at left 2nd ICS, grade 4/6. He had vesicular breath sound with fine crepts on both lungs with hepatomegaly 3 cm at mid clavicular line from right costal margin.

Chest X ray revealed cardiomegaly with plethoric lung fields with persistent prominent opacity in right hilar region. CT scan of chest with contrast was done which showed a large PDA with plethoric lungs with atelectasis. We did all preprocedural investigations which were normal except low hemoglobin (9 gm/dl), leukocytosis (18 x 109/L) and increased C-reactive protein (CRP) (3 mg/dl). He was admitted in hospital and was treated with inj. ceftriaxone, inj. fimoxyclav inj. furosemide, inj. lasix, tab enalapril, tab digoxin. After one week of ICU treatment, he was fit for procedure with normal total WBC count (11 x109/L) and CRP (0.3 mg/dl).

His angiogram suggested large PDA (10 mm) with moderate pulmonary hypertension (PASP 40 mmHg) with PVR 3.99, PVR/SVR 0.31). PDA

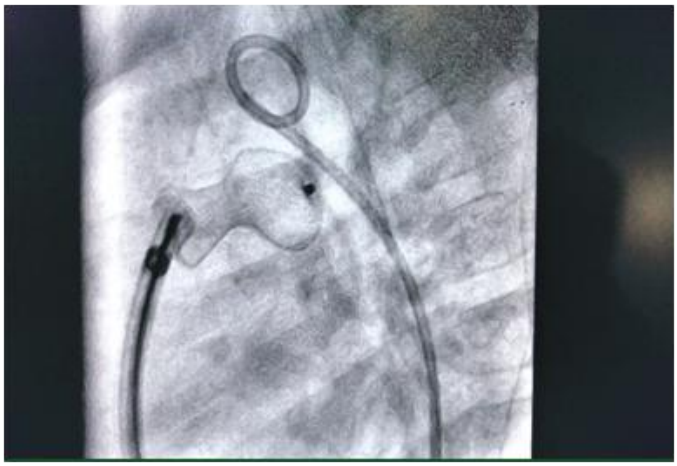

Figure 1: Large tubular PDA (10 mm) was closed by ADO I 14/12

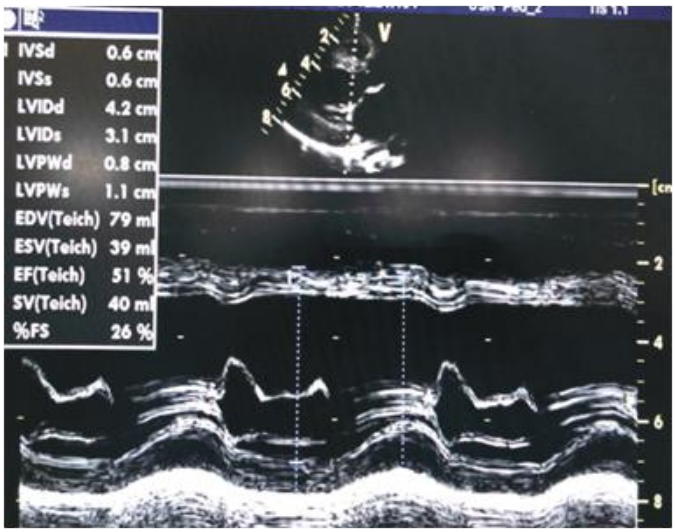

Figure 2: Post procedure left ventricular dysfunction revealed by echocardiography (LVEF 53%)

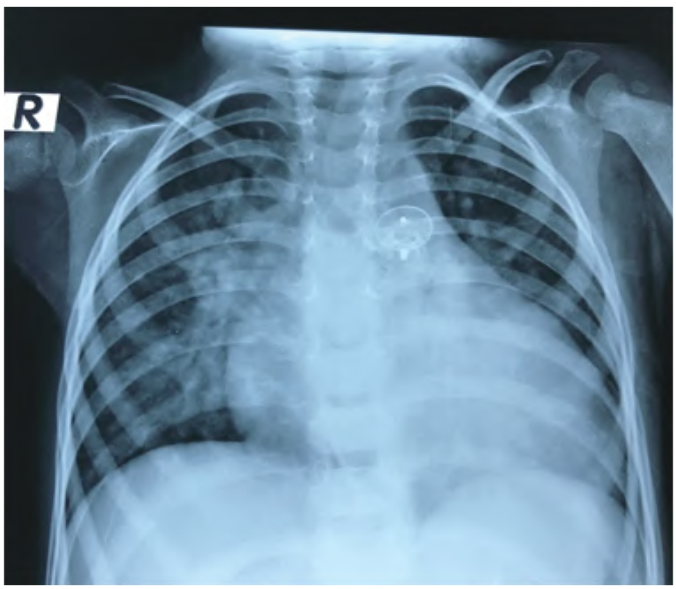

Figure 3: Post device closure chest X-ray showed PDA device in situ, cardiomegaly with plethoric lung field

device closure was done successfully by Amplatzer PDA device 14/12 (Figure.1). Device implantation was done by slight pulling of the device inside the PDA making as mashroom head. Before deploying the device oxygen was given for ten minutes, PASP was reduced to 30 mmHg with reduced PVR 1.36 and PVR/SVR 0.14.

After procedure he was shifted to PICU. Post procedure echo revealed PDA device in situ with mild intradevice leak with partial para device leak, mild AR, moderate to severe MR, mild pulmonary arterial hypertension, dilated left atrium and left ventricle (LV). No left pulmonary artery or no descending aorta obstruction, no pericardial or pleural effusion with fair LV function (LVEF 53%) (Figure. 2). Just after procedure his vital signs were within normal range (BP: 95/50 mmHg, HR:125 beat/min, RR: 25 b/min). In PICU we started inf. dobutamine (5microgram/kg/min) to support cardiac function. Inf. dobutamine act as inotrope and afterload reducing agent. But after 5 hours of procedure, he developed sudden spike of blood pressure (160/65mmhg, above 95th centile). Immediately infusion lasix was started since post device closure chest X-ray also revealed cardiomegaly with plethoric lung field (Figure 3). As the blood pressure remained high, inf. milrinone was added followed by ACEI while NPO was over. But still his blood pressure was high. Tab nifedipine was also added and other cardiac medications were going on. Gradually child's left ventricular function improved with stable vital signs within 4 days after procedure. However, he was under treatment of infusion dobutamine, inj.lasix followed by tab ACEI and oral diuretics. During hospital stay we excluded hemolysis and hematuria. On 5th day post procedure, the child was discharged with medications and advice. On 7th day post procedure, child came for follow up and echo revealed PDA device in situ, tiny residual PDA with intra device leak (improving), moderate MR, mild PAH, GBVF (LVEF 55%). He was on oral diuretics and ACEI for 7 months and next 5 months he was on oral diuretics.

After one year (on 29/10/2022) follow up echocardiography showed PDA device in situ, no

residual PDA, no intra or para device leakage, unobstructed flow through the descending aorta and LPA, no MR, no PAH, GBVF. (LVEF 60 %).

DISCUSSION

Patent ductus arteriosus (PDA) is the commonest congenital heart disease with prevalence 5-10% of all congenital heart disease, excluding premature infants[1]and is estimated to occur approximately 1 in 2000 live births.[2] In 1971 Portmann described the first successful transcatheter closure of PDA and thereafter the procedure became the standard of care and widespread in the 1980[2]. Transcatheter closure of PDA has become the state of art for most cases, reserving only the surgical options for very few cases[3]. With large technical advances in the devices used for pediatric cardiac interventions, even large PDAs are now amenable for transcatheter closure[4].

Large PDA with hemodynamically significant left to right shunt causes pulmonary over-flooding that result in left ventricle (LV) volume over burden and remodeling, and it compensates by expanding stroke volume. Growth failure, repeated history of lower respiratory tract infection and congestive cardiac failure (CCF) occurs in cases of large PDA with greater shunts.[4,5] Our patient had the similar clinical presentation which was described in various studies. Furthermore, he had history of noncompliance of cardiac medication which deteriorated the status of overburdened LV with moderate to severe MR, mild AR and pulmonary hypertension. It is documented that large PDA with severe pulmonary hypertension causes cardiopulmonary dysregulation. Hence, large PDA should be closed surgically or by PDA device. Percutaneous PDA closure has proved to be safe and effective with short- and long-term results comparable to surgical closure.[5] As the size and shape of the PDA of our patient was suitable for device closure, we closed the PDA by ADO[1]. But immediately after closure of large PDA shunt caused sudden reduction of left ventricular preload and increase of afterload and left ventricular dysfunction with instability of vital signs.

This significant sudden reduction of left ventricular preload and increase of afterload lead to left ventricular dysfunction and sudden rise of blood pressure is documented as “Post PDA closure syndrome” in different research. Moreover, that firmly established ventricular dysfunction following PDA ligation through surgical or percutaneous device closure has been reported by various studies elsewhere.[6-8]

Post PDA device closure syndrome is the cardiorespiratory instability that may occur almost within the first few hours to 12 hours after device closure or ligation of large PDA. It happens due to LV dysfunction and vascular tone dysregulation. The recent studies reported a decrease in fraction shortening (FS) and ejection fraction (EF) associated with an increase systemic vascular resistance evidenced by abrupt rise of systolic blood pressure and a sudden reduction in preload.[5,6,9] Our child also had the same clinical findings such as sudden rise of systolic pressure and reduced LV ejection fraction (LVEF) with severe MR which were documented by 2 D and color doppler echocardiography.In a Saudi Arabian study, Galal et al described the immediate deterioration of LV EF and LV FS after closure of large PDA in children which required few months to recover.[7]

In recent studies it was documented that long term large PDA causes hemodynamic changes of an older child and there was high rate of occurrence of post-operative left ventricular systolic dysfunction.[6] Presence of preprocedural associated cardiac lesions such as severe MR that our patient had, would have been the predictor factor of post device closure left ventricular dysfunction or post device closure syndrome.[6]

Gupta SK et al described how LV dysfunction happens after percutaneous PDA device closure which is reversible. According to their study, large PDA causes increase preload of left ventricle which actually causes left ventricular volume overload and remodeling evidenced by alteration of systolic and diastolic dysfunction[8]. Pre device closure of large PDA according to Frank Sterling law, increased preload causes increased stretching of left ventricular muscle and contractility to overcome significant left to right shunt. Sudden reduction in preload and simultaneous relative increase in

afterload due to removal of low resistance pulmonary circulation leads to 'afterload mismatch'. Sudden changes in loading conditions, preexisting LV volume overload and chronic compensation by Frank Starling mechanism explains immediate post closure LV dysfunction. Therefore, increased LVEDd and increased LV FS% of children with large PDA shunt are foremost data to follow up after device closure of PDA as these have been changed drastically post procedure. During follow up there was reduction of LVEDd, LVEDV, LVESd LVESV, while LVEF improved.

Our patient had improvement of LV dysfunction after few days of device closure although his moderate to severe MR fully resolved after one year. The child was on ant failure medication as ACEI and diuretics till thento compensate the increased afterload and LV dysfunction.[5]

CONCLUSION

Percutaneous closure of hemodynamically noteworthy PDA causes post device closure syndrome which is evidenced by sudden rise of systolic blood pressure and reversible LV dysfunction within 12 hours to 24 hours of the procedure. Patient requires ant failure medications for long time to revert the remodeling of LV. The large size of PDA, older age of patient, associated cardiac lesion, preprocedural LVEDd would be suspected as the predictor factors of acute decrease in LV systolic function. To control post PDA device closure syndrome, afterload reducing agent (ACEI) has significant role till normalization of LV volume overload and LV dysfunction.

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