Obstructive lesions in the aortic arch in children: A one year experience from a tertiary centre from western India

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Abstract

Background: Obstructive lesions in the aortic arch form a substantial disease burden in emergency care, outpatients and hospitalized children with congenital structural heart disease.

Objectives: To determine the clinical findings, imaging abnormalities and short term (<6 months) post-intervention outcomes in patients presenting with obstructive lesions of the aortic arch.

Method: A prospective observational study was conducted in children (less than 18 years of age) admitted with structural aortic arch obstructive lesions at Bai Jerbai Wadia Hospital for Children, Mumbai from 1st of January to 31st of December, 2019.

Results: There were 25 children admitted with structural aortic arch obstructive lesions during the study period; 14 (56%) were diagnosed as coarctation of aorta (COA), 8 (32%) as hypoplastic aortic arch and 3 (12%) as interrupted aortic arch. Eight (32%) admitted patients underwent catheter-based intervention and 17 (68%) patients required surgery. Re-coarctation was more common in patients with catheter-based intervention.

Conclusions: In the 25 children admitted with structural aortic arch obstructive lesions, 56% had COA, 32% hypoplastic arch and 12% interrupted aortic arch. Whilst 32% underwent catheter-based intervention, 68% required surgery.

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Introduction

Obstructive lesions of the aortic arch are classified into 4 categories viz. discrete coarctation of the aorta (COA), segmental hypoplasia of the aortic arch, interrupted aortic arch and a combination of hypoplastic aortic arch and COA[1]. COA is a common congenital heart defect (CHD) found in approximately 1 per 2900 live births[1-3] and is the seventh common type of CHD[4]. This is probably an underestimate as the diagnosis may be delayed in the paediatric population[4,5]. There is a paucity of epidemiological data on children admitted with aortic arch obstruction in the Indian context.

Objectives

To determine the clinical findings, imaging abnormalities and short term (<6 months) post-intervention outcome in patients presenting with obstructive lesions of the aortic arch.

Method

A prospective observational study was conducted in children (less than 18 years of age) admitted with aortic arch obstructive disease at the Department of Paediatric Cardiology and Cardiac Surgery, Bai Jerbai Wadia Hospital for Children, Mumbai from 1st of January to 31st of December, 2019. The hospital is a tertiary referral centre of western India, serving around 1000 paediatric cardiac cases per month.

Inclusion criteria: Children (below 18 years of age) admitted with COA, hypoplastic aortic arch and interrupted arch illness during the study period were included.

Exclusion criteria: Children admitted outside the study period, children whose parents were not willing to give consent and those who were lost to follow up were excluded.

Ethical issues: Approval was obtained from the Institutional Ethics Committee of Bai Jerbai Wadia Hospital for Children, Mumbai, India (No. IEC-BJWHC/64A/2019). The confidentiality of the patients was maintained and the obtained data remained with investigators and was not shared with any unauthorized persons. Written informed

consent was obtained from the parents of the participating children.

A standardized questionnaire was formulated according to the protocol of an observational study that included data depicting demography (age, sex, gender), clinical parameters (symptoms and signs), age at diagnosis and laboratory parameters like electrocardiographic and echocardiographic findings, catheterisation laboratory data, and treatment related outcome. No previously used data collection tool was used. From the records, clinical and associated medical history, birth weight, age at diagnosis, heart rate, blood pressure and oxygen saturation were collected. From the echocardiographic study, information regarding type of anatomy, nature and gradient across the arch abnormality and left ventricular functions were collected. The data from additional imaging, including cardiac catheterisation or computed tomography (CT) scan (if performed) were collected. The surgical details (type of repair, nature of lesion and age of patient at surgery) and post-operative hospital stay details (intensive care unit stay (days), need for extracorporeal membrane oxygenation (ECMO), ventilation requirement, re-intervention in immediate post-operative period) were collected from the medical records. The follow up data (weight gain, ejection fraction, oxygen saturation, re-intervention after discharge) were collected from the patient's out-patient department records during the follow up visits up to 6 months.

Results

There were 25 children admitted with structural aortic arch obstructive lesions during the study period of whom 16 (64%) were male and 6 (24%) were syndromic. Figure 1 shows the age of presentation. Fourteen (56%) children presented in infancy 6 (24%) being neonates.

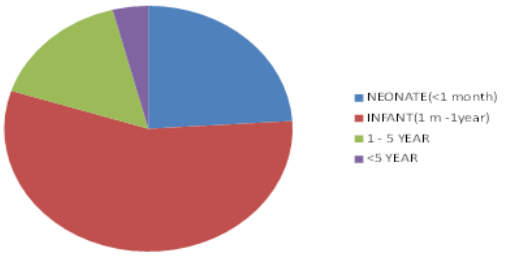

Figure 1: ***Age of presentation***

Figure 2 shows the birth weights. The mean birth weight was 2.3 kg with 3 (12%) babies being below 2kg.

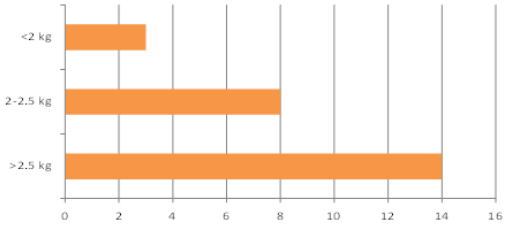

Figure 2: ***Birth weight***

Figure 3 shows the presenting symptoms. Ten (40%) presented with an asymptomatic murmur and decreased lower limb pulsation whilst 7 (28%) presented with features of heart failure.

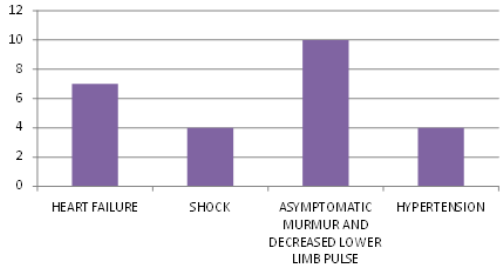

Figure 3: ***Presenting symptoms***

Figure 4 shows the presenting symptoms in relation to age. Majority of neonates presented with shock whereas young children presented mainly with hypertension.

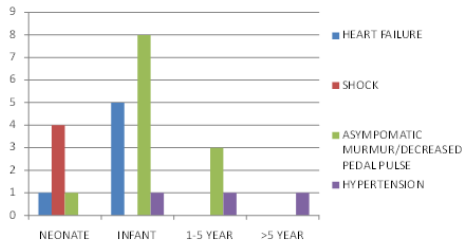

Figure 4: ***Presenting symptoms in relation to age***

Figure 5 shows the associated structural cardiac anomalies. The most common associated cardiac anomaly was patent foramen ovale / atrial septal defect. This was followed by patent ductus arteriosus and bicuspid aortic valve. Interrupted aortic arch was mostly associated with ventricular septal defect.

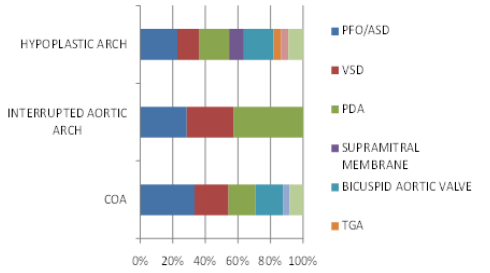

Figure 5: ***Associated structural cardiac anomalies***

Figure 6 shows the non-cardiac issues. Congenital kidney anomalies (multicystic dysplastic kidneys, ectopic fussed kidneys), liver disorders (cholestatic jaundice) and brain anomalies (periventricular leukomalacia, cortical cysts) were the associated non-cardiac issues.

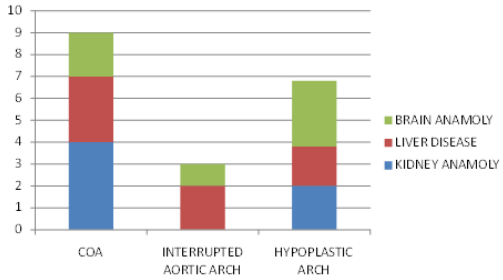

Figure 6: ***Non-cardiac issues***

Figures 7, 8 and 9 show the heart rate, oxygen saturation and blood pressure at presentation. On presentation, the mean heart rate was 133.2/min, mean oxygen saturation (Sp02) was 96.6% and mean blood pressure was 70/38 mmHg.

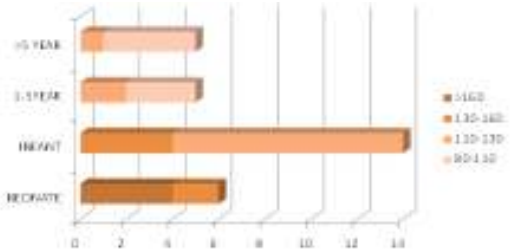

Figure 7: ***Heart rate at presentation***

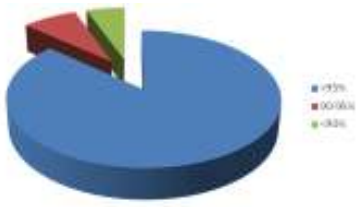

Figure 8: ***Oxygen saturation at presentation***

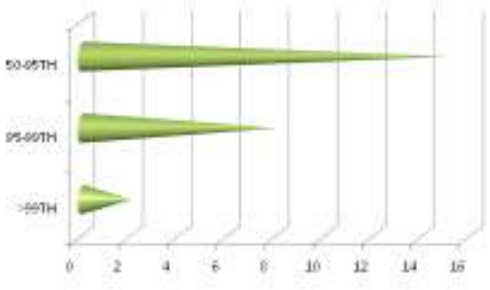

Figure 9: ***Blood pressure at presentation***

On investigation, a 12-Lead electrocardiogram (ECG) revealed left ventricular hypertrophy (by voltage criteria) in 11 (44%) patients. Echocardiographic evaluation showed that 14 (56%) affected children had discrete COA, 8 (32%) had hypoplastic arch and 3 (12%) had interrupted aortic arch. Both COA and hypoplastic arch were present in 6 children. The site of the pathology in isolated COA was post-subclavian in 14 (56%) and pre-subclavian in 11 (44%). Transverse arch hypoplasia was found in 16 (64%) patients with hypoplastic arch followed by uniformly small arch and distal arch hypoplasia (Figure 10).

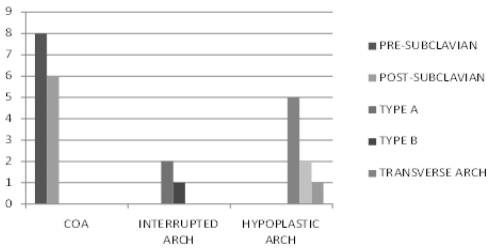

Figure 10: ***Echocardiographic measurements***

In case of interrupted aortic arch there was predominantly Type A interruption. In cases of isolated COA, the initial pressure gradient was >60 mm Hg in 7 (28%) cases, between 40-60mm Hg in 11 (44%) cases and <40mm Hg in 7 (28%) cases. Most patients with hypoplastic arch had a pressure gradient of <40mm Hg (Figure 11).

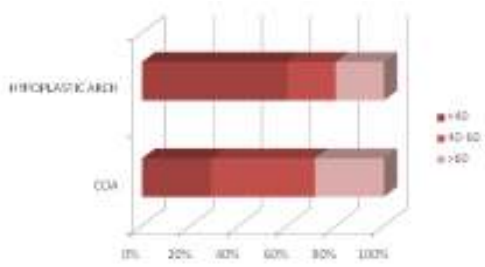

Figure 11: ***Initial pressure gradient***

The Z-score lay between -2 to -3 in most cases but extreme form of hypoplasia was also found in 2 patients (Figure 12).

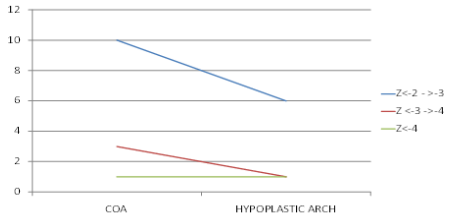

Figure 12: ***Echocardiography measurement: Z-score***

Cardiac CT also revealed similar results with the majority of patients having post-subclavian coarctation although hypoplastic arch were readily diagnosed by CT (48%) compared to echocardiography [fig 13].

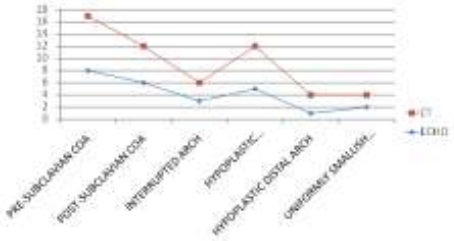

Figure 13: ***Comparison of Echo and CT findings regarding site***

Regarding cardiac function, majority of the left ventricular dysfunction was found in isolated coarctation cases [Figure 14].

Figure 14: ***Left ventricular dysfunction***

In terms of management, 14 (56%) of COA underwent balloon coarctoplasty and the rest of the cases associated with arch hypoplasia required surgical correction. Catheterisation data showed that the initial pressure gradient was mostly in the range of 20-40 mm Hg whereas post-intervention, it reduced to <20 mm Hg. All the patients with interrupted arch and hypoplastic arch underwent surgical correction. The post-operative intensive care unit (ICU) stay was <7 days in 19 (75%) patients but 6 (24%) patients required prolonged ICU stay >7 days. One patient was put on extracorporeal membrane oxygenation (ECMO) support and eventually died. In follow up, 19 (75%) patients who had balloon dilatation and 4 (17%) with surgical correction had re-coarctation that required re-intervention (Figure 15).

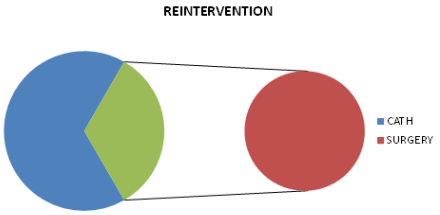

Figure 15: ***Re-intervention required after initial procedure***

Overall follow-up data revealed that the majority of children had normal left ventricular ejection fraction >60% and blood pressure within 50-95th centile (Figures 16 and 17).

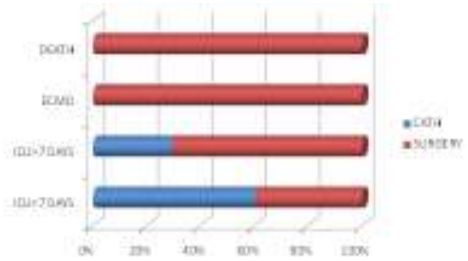

Figure 16: ***Outcome following intervention***

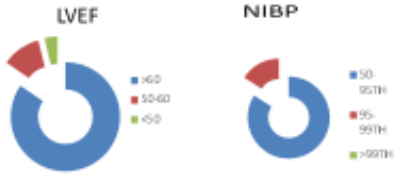

Figure 17: ***Follow-up data***

Discussion

Obstructive lesions of the aortic arch include COA, interrupted aortic arch and hypoplastic arch[1]. These abnormalities in systemic blood flow occur with varying severity ranging from minimal haemodynamic compromise to profound haemodynamic derangements[2]. Most often, the obstruction at aortic arch is identified because of a murmur or hypertension detected on routine examination. Echocardiography is the examination of choice for assessment of aortic arch anomalies in children. Cardiac CT provides the exact anatomy of the aortic arch and collateral circulation and is useful to establish surgical strategy. Significant hypertension, shock or congestive heart failure are indications for intervention. Surgical relief of the aortic obstruction and catheter interventional techniques (balloon angioplasty and stents) are available alternatives. These patients continue to require lifelong follow-up for management of associated problems including arterial hypertension, atherosclerotic disease, re-coarctation and aneurysm formation. Similarly, preliminary data of this study showed that there is a significant disease burden of obstructive lesions of aortic arch in the paediatric population among which 8 (32%) admitted patients required catheter-based interventions and 17 (68%) required surgery.

Amongst obstructive lesions in the aortic arch, CoA was found in 1 per 2900 live births[1-3]. CoA was found in 18% of patients with Turner syndrome in the study by Cramer JW, *et al*[6] and in 10% of patients with Williams syndrome by Pham PP, *et al*[7]. Similar results were revealed in our study where CoA was the predominant entity amongst aortic arch obstruction followed by hypoplastic arch and interrupted arch. Amongst the total affected children, 6 (24%) were syndromic. Majority of the children (56%) presented in infancy with asymptomatic murmur and decreased lower limb pulsation. Neonates commonly presented with shock whereas young children presented mainly with hypertension. The most common associated cardiac anomaly was patent foramen ovale and patent ductus arteriosus followed by bicuspid aortic valve. Similar results were obtained in studies by Roos-Hesselink JW, *et al*[8], Kappetein AP, *et al*[9]and Oliver JM, *et al*[10]. Interrupted aortic arch was mostly associated with ventricular septal defect. Non-cardiac issues like congenital kidney anomalies (multicystic dysplastic kidneys, ectopic fused kidneys), liver disease (cholestatic jaundice) and brain anomalies (periventricular leucomalacia, cortical cysts) were associated mostly with cases of interrupted arch (65%) and discrete coarctation (35%) in our study whereas Curtis SL *et al*[11] had shown that prevalence of intracranial aneurysms was 10% in patients with CoA in their study.

The clinical presentation of COA differs significantly in paediatric patients in comparison with adults. Although infants with severe COA may present with signs and symptoms of heart failure and cardiogenic shock as the ductus closes, most adults present with systemic arterial hypertension. On physical examination, femoral arterial pulses are diminished and usually delayed[12]. In this study, the mean heart rate was 133.2/min, mean Sp02 was 96.6% and mean NIBP was 70/38 mm Hg. A 12-Lead ECG revealed left ventricular hypertrophy (by voltage criteria) in 11 (44%) patients.

Echocardiographic evaluation of the study population showed that 14 (56%) affected children had discrete COA, 8 (32%) had hypoplastic arch and 3 (12%) had interrupted aortic arch. Both COA and hypoplastic arch were present in 6 children. In the study by Van Praagh R, *et al*[13], in a large series of patients with interruption, 42% were type A, 53% type B and 4% type C whereas predominantly type A interruption was found in our study. The site of the pathology in isolated coarctation was post-subclavian in 16 (56%) of cases and pre-subclavian in 12 (44%) of cases. Pillsbury RC, *et al*[14] suggested that when the involved segment is hypoplastic, it remained as an anatomic entity, although lacking any luminal patency. The persisting presence of the fibrous connection between the patent components of the arch also limits the length of the gap between them. Transverse arch hypoplasia was found in 16 (62%) in our study followed by uniformly small arch and distal arch hypoplasia. We found that most patients with hypoplastic arch had pressure gradient <40mm Hg. In addition, the Z-score lay between -2 to -3 in most cases though the extreme form of hypoplasia was found in 2 patients.

Cardiac CT demonstrates visualization of the aortic arch with precise characterization of the location and extent of coarctation, and assessment of the presence and extent of collateral vessels. CT angiography is also used to assess concomitant coronary anomalies. Cardiac CT provides exceptional visualization of the aortic arch and detection of post-repair complications including arch "kinking" and pseudoaneurysm[12]. In our study, we found that cardiac CT findings were corroborative with the echocardiographic findings although hypoplastic arch was more readily diagnosed by CT (48%) compared to echocardiography.

Regarding treatment, our study revealed some interesting aspects about current practices of interventions. Warnes CA, *et al*[15] suggested that in patients with a native CoA or re-coarctation, a measured peak-to-peak gradient greater than or equal to 20 mm Hg by cardiac catheterisation is an

indication for intervention, either by transcatheter or surgical approach, whereas patients with extensive collaterals should undergo intervention even if the peak-to-peak gradient is less than 20 mm Hg. Our catheterisation data showed that the initial pressure gradients were mostly in the range of 20-40 mm Hg, whereas post-intervention it was reduced to <20 mm Hg. Accordingly, 14 (56%) coarctation underwent balloon coarctoplasty and the rest required surgical correction. The post-operative ICU stay was <7 days in 19 (76%) patients but 6 (24%) patients required prolonged ICU stay >7 days. One patient was put on ECMO support and eventually died.

On follow up, our data revealed that 19 (75%) of the patients who had balloon dilatation had re-coarctation that required re-intervention. Adams EE, *et al*[16], Cohen M, *et al*[17] and Pandey R, *et al*[18] have suggested that patients with repaired coarctation are at risk of late re-coarctation and aneurysm development. The study by Brown ML, *et al*[19] opined that a younger age at the time of surgery was associated with a higher risk of re-stenosis. Our study results revealed re-coarctation in 17% patients with surgical correction and this required re-intervention. However, overall follow-up data revealed that the majority of the children had normal left ventricular ejection fraction >60% and blood pressure within 50-95th centile.

The strengths of our study are that it was prospective in design and probably the first such epidemiological study from Western India. The current practices have been better reflected by the study. Moreover, conclusions from the final analysis might assist policy formulation. However, there are some limitations to our study. Firstly, it was a tertiary centre-based study and hence may not be representative of disease pattern in the community. Also, the number of patients were relatively small and that might result in the deviation of statistical assumption.

Conclusions

In the 25 children admitted with structural aortic arch obstructive lesions, 56% had COA, 32% hypoplastic aortic arch and 12% interrupted aortic arch. Echocardiographic findings were corroborative of CT scan findings. Thirty two percent underwent catheter-based intervention and 68% required surgery. Re-coarctation occurred mainly with catheter-based interventions.

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