AORTIC OBSTRUCTION SECONDARY TO NON-SPECIFIC AORTOARTERITIS IN CHILDREN MASQUERADING AS DILATED CARDIOMYOPATHY

Poster Contributions
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Background: Nonspecific aortoarteritis (NSAA) affects the aorta and its major branches. We hereby report a series of children with NSAA related aortic obstruction causing severe left ventricular (LV) dysfunction and heart failure. This presentation is not uncommon, yet often misdiagnosed in clinical practice.

Methods: Out of a total of 160 children between 3 and 15 years of age who presented with LV dysfunction over the last 4 years, 19 (11.8%) were diagnosed to have significant aortic obstruction secondary to NSAA and formed the study group. The data of these patients has been analysed retrospectively for the clinical details, management strategies and follow up.

Results: The mean age was 9.89 years (range 3-15 years), 14 were females. LV dysfunction was severe i.e. LV ejection fraction (EF) <20% in 16. Nine of 19 children (47.4%) did not have either hypertension or discrepancy in peripheral pulses at presentation. Rest of 10 patients had only mild hypertension. Importantly, none of the patients had an established diagnosis of NSAA and 15 of them were being treated as idiopathic dilated cardiomyopathy. Twelve children presented with acute heart failure and all these underwent emergent or urgent balloon dilatation for aortic obstruction. Echocardiographic trivial to mild aortic regurgitation was present in five (26%) patients. All patients underwent successful balloon dilatation of stenotic segment of aorta. Symptomatic improvement was observed in all immediately. During the follow up (2 months to 4 years), five (26%) patients underwent balloon dilatation twice while two (10.5 %) patients had the procedure repeated thrice. Stent was implanted in four (21%) patients for persistent gradient and/or severe LV dysfunction. LVEF normalised in 5 (26%) while it improved significantly in the rest.

Conclusion: NSAA related aortic obstruction is rare but an important cause of LV dysfunction in children. The typical presentation with hypertension and discrepant peripheral pulses may not be seen in a significant number. The presence of unexplained AR in the setting of LV dysfunction may provide a clue. A high index of suspicion is needed for the recognition as the management is rewarding.