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Association of mitral regurgitation with anomalous left coronary artery from the pulmonary artery (ALCAPA) in children.

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Objective: To evaluate the frequency and association of isolated mitral regurgitation with ALCAPA syndrome in children presenting with left ventricular dysfunction or dilated left ventricle.

Methods: 20 year review of ALCAPA syndrome from two paediatric cardiac centres in United Kingdom was undertaken. Patients with abnormal coronary artery origins apart from ALCAPA syndrome were excluded.

Results: Eighteen consecutive patients were identified. Twelve of them were females (66%). Age at presentation varied from one month to seventeen years. The age from first presentation to suspect the diagnosis of ALCAPA varied from one week to eight years. Only 3 patients had the diagnosis made at presentation (16%). The commonest reasons for referral were features of cardiac failure in 9 (50%), and respiratory distress or asthma in 8 (44%) patients. 5 (27%) patients were investigated for failure to thrive. The common initial diagnosis was dilated cardiomyopathy in 12 (66%). The echocardiographic features at presentation included LV dysfunction in 11 (61%), significant mitral regurgitation in 8 (44%). Mitral regurgitation was considered as the primary pathology in 8 patients (44%) hence its association with ALCAPA was overlooked for as long as 5 years. On the contrary of general view, with ultrasound examination only 4 (22%) patients exhibited dilated right coronary artery, and in 5 (27%) the origin of left coronary artery could not be clearly visualised. 8 (44%) showed abnormal retrograde flow in the pulmonary artery. The commonest ECG findings were nonspecific ST segment changes in 11 (61%), pathological Q wave in 14 (77%) in lead aVL, and T wave inversion in 10 (55%) in leads aVL and V6. 8 (44%) patients underwent cardiac catheterization to confirm the diagnosis. All 18 patients had surgery immediately after diagnosis. Post-surgical follow up echocardiogram showed resolution of LV dysfunction in 10 (55%) patients. 3 (16%) patients died following surgery.

Conclusion: Isolated mitral regurgitation is an important and frequent finding in children with ALCAPA syndrome. Any child with echocardiographic diagnosis of isolated significant mitral regurgitation and left ventricular dysfunction or dilatation should prompt a search for ALCAPA syndrome.