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

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Background

Origin of left coronary artery from the pulmonary trunk is one of the more common coronary arterial abnormalities encountered in children. It is a rare congenital anomaly and occurs from 1 in 250 to 1 in 400 of all congenitally malformed hearts with an overall incidence of approximately 1 in 300000 children. This anomaly is seen in greater frequency in boys than in the girls with the ratio being 2.3 to 1. It usually presents in the neonatal period with congestive cardiac failure due to myocardial ischemia. Children can present with progressive mitral regurgitation which may or may not be accompanied by electrocardiographic signs of ischemia. Although there are isolated case reports of diagnosis of ALCAPA in adults, failure of early and accurate diagnosis leads to overall poor outlook.

Aims

To evaluate the frequency and association of isolated mitral regurgitation with ALCAPA syndrome in children presenting with left ventricular dysfunction or dilated left ventricle to a tertiary paediatric cardiac center.

Patients and methods

We undertook a twenty year review of ALCAPA syndrome from two paediatric cardiac centers in United Kingdom namely UHW, Cardiff and LGI, Leeds. Patients with abnormal coronary artery origins apart from ALCAPA syndrome were excluded. The data was identified from medical and surgical records and follow-up data was obtained from departmental database and review of patients notes. The analyzed data is presented in the form of total numbers and percentages unless stated otherwise.

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.

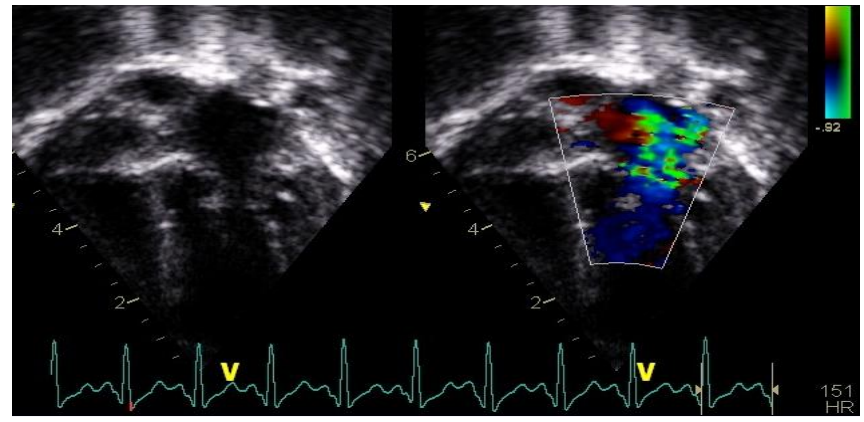

Figure 1 . Severe MR with dilated LV

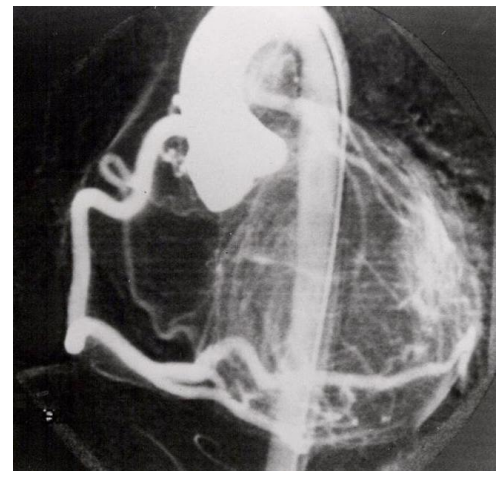

Figure 2. Cardiac catheterisation showing enlarged RCA which fills the LCA via collaterals

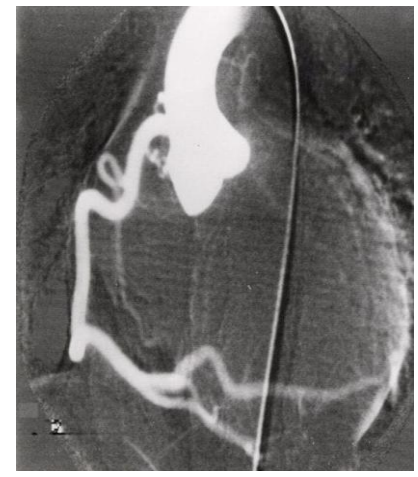

Figure 3. Coronary angiography showing failure of LCA to arise from aorta

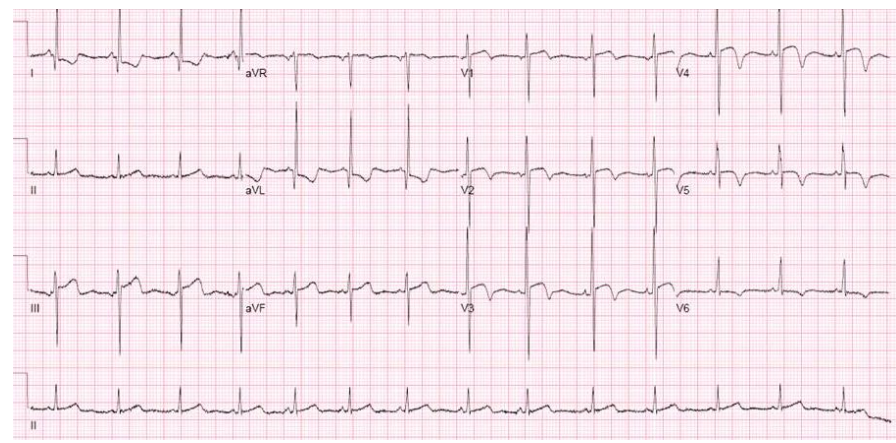

Figure 4. An anterolateral infarct pattern with abnormal deep (>3 mm) and wide (>30 msec) q waves is observed in leads I, aVL, absent q waves in leads II, III, and aVF, and poor R wave progression across the precordial leads.

Conclusions

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.