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Coronary Artery Disease in Children

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A simple PubMed search on coronary artery disease AND children returns nearly six thousand citations. Needless to say, majority of them are on Kawasaki disease. It is now well established that Kawasaki disease is an important cause of heart disease in children, to the extend that it is the commonest cause in the developed world [1]. Coronary anomalies as a cause of myocardial infarction in infancy with the classical presentation of anomalous origin of left coronary artery from pulmonary artery (ALCAPA) as a cause of *greying spells* on crying has been known for a long time. Coronary artery calcification in children with chronic kidney disease is another important cause [2]. Coronary artery dilatation has been noted in children with sickle cell disease, though the significance is unknown. In one study, coronary artery dilatation could not predict morbidity in children with sickle cell disease [3]. Other reasons for coronary artery disease in children are progeria, post surgical coronary lesions and cardiac allograft vasculopathy [4]. Coronary artery dilatation has been documented in children with paediatric onset systemic lupus erythematosus [5].

Coronary artery involvement in Kawasaki disease

Kawasaki disease is the most important cause of acquired coronary artery abnormalities in children. It can lead to coronary aneurysms in about one fourth of the untreated cases [1]. Though early initiation of treatment with intravenous immunoglobulin (IVIG) is the sheet anchor of treatment in Kawasaki disease, 10 – 20% may not respond to the initial IVIG treatment and additional treatment may be required. Timely treatment with IVIG can reduce the chance of coronary aneurysm formation from 25% to 4%. Children who develop coronary aneurysms need life long cardiology follow up.

Though very mildly dilated and inflamed coronary arteries may return to normal, this does not occur in large saccular aneurysms. The coronary artery in this region has lost intima, media and elastica and cannot be regenerated. In giant aneurysms, only a thin layer of adventitia is remaining. Psudo resolution of aneurysms may occur with decrease in lumen size by formation of layered mural thrombi. Large aneurysms can rarely rupture leading to pericardial tamponade. Rupture usually occurs in the first two to three weeks and seldom after that. Myocardial infarction can occur due to thrombosis in affected coronary arteries or due to stenosis of the vessels.

Coronary artery anomalies

Though various types of coronary artery anomalies involving different coronary branches have been described, the one which is most familiar to all is the ALCAPA. It may present as cardiac failure in infancy or an anginal equivalent with pallor on crying or feeding (greying spells). ALCAPA is one condition to be kept in mind while evaluating isolated mitral regurgitation in infants. A close look at coronary origins is a must during echocardiographic evaluation. The pressure in pulmonary artery being lower, ALCAPA acts a left to right shunt, though usually small in volume, with flow from collaterals retrogradely into the left anterior descending artery and then into the pulmonary artery.

Other important coronary anomalies are those in tetralogy of Fallot which cross the right ventricular outflow tract and one in which a malignant anomalous origin of right coronary artery from left sinus, passes between the aorta and pulmonary artery. Later can even cause sudden death during exercise, especially in young asymptomatic athletes [6].

Coronary artery fistula and associated ectasia

Coronary artery fistula can be seen occasionally in children. Vinograd CA and associates found that small fistulae mostly arose from left anterior descending coronary artery, drained into pulmonary artery and only 3 out of 92 had associated ectasia. Large fistulae was more often seen in females and most originated form the right coronary artery and drained into right atrium. Of the 12 patients who underwent procedural closure, all had ectatic feeding coronary arteries at a mean follow up period of around 4 years [7].

Transplant coronary artery disease

Cardiac allograft vasculopathy or transplant coronary artery disease is one of the important problems which limit long term survival in pediatric cardiac allograft recepients. Routine surveillance coronary angiography is an established method to detect cardiac allograft vasculopathy. Contrast enhanced cardiac magnetic resonance imaging could detect significant difference in coronary enhancement intensity between those with and without angiographically detectable transplant coronary artery disease in one study [8].

Conclusion

Though coronary artery disease is extremely uncommon in children, one should be on the look out for the rare causes, which can occasionally manifest. Kawasaki disease is the most important cause of acquired coronary artery disease in children and manifest mostly as coronary aneurysms. Coronary involvement in Kawasaki disease can be prevented to a large extend by early IVIG treatment. Other acquired causes are coronary calcification in CKD, cardiac allograft vasculopathy and coronary dilatation in SLE. Coronary anomalies are the major group of congenital coronary lesions, which usually manifest in early infancy.

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