The Role of Transoesophageal Echocardiography during Repair of Multiple Congenital Coronary Artery Fistulas

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Coronary artery fistula (CAF) is a very rare lesion occurring in approximately 1 in every 50,000 patients with congential heart disease, but at the same time CAF is the most common congenital anomaly of the coronary artery. CAF is characterized by a normal aortic origin of the coronary artery involved, but with a fistulous communication with the atria, ventricles or with the pulmonary artery, coronary sinus or vena cava. Two pediatric patients (3 years and 4 years) presented with multiple congenital coronary arterial fistulas. In both cases the fistulas entered the right ventricle. The fistulas in both patients were successfully closed surgically.

Intraoperative transesophageal echocardiography with color flow Doppler was used for a precise location of the fistulous communication, selective demonstration of vessels feeding the fistula and documentation of abolition of fistulous flow.

Key words: congenital heart disease, coronary artery fistula, transesophageal echocardiography, surgical repair

INTRODUCTION

Coronary artery fistula (CAF) is a very rare lesion occurring in approximately 1 in every 50,000 patients with congenital heart disease (1), but at the same time CAF is the most common congenital anomaly of the coronary artery (2). Wearn et al. (3) has described three types of pathological communication between the coronary arteries and the cardiac chambers: the arterioluminal type, in which the CAF enters the cardiac chamber directly; the arteriosinusoidal type, with the communication through the sinusoidal network, and the arterio-capillary type, which allows fistulization into the capillaries and through the thebesian system into the heart chamber.

The affected coronary artery becomes progressively dilated and tortuous, and at times truly aneurysmal. The fistula may involve the right or the left coronary artery or one of their branches. The site of abnormal termination may be a cardiac chamber, the pulmonary artery or vein, the coronary sinus or vena cava.

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CAF can originate from the right coronary artery in about 60%, from the left coronary artery in 32%, from both arteies in 2% and from single artery in 7% (1, 6). Patients usually are asymptomatic. A continuous murmur, similar to the murmur of patent ductus arteriosus, is audible over the precardium.

The ECG is normal but may show right ventricle hypertrophy, or left ventricle hypertrophy if the CAF is large.

Chest X-ray usually shows a normal heart size. Pulmonary artery hypertension is rare and pulmonary vascular resistance elevation has not been reported. Echo studies usually suggest the site and type of the CAF. Closure of the CAF is indicated as soon as the diagnosis is determined, because congestive heart failure (19%), subacute bacterial endocarditis (4%), fistula rupture (0.5%), myocardial infarction (9%) and sudden death (14%) are significant risk factors (2, 4, 5, 11).

Successful percutaneous catheter coil embolisation or a double-umbrella device was reported (7–9).

In the surgical closure of CAF, also by using cardiopulmonary bypass (CPB), the fistulous point is closed nearer to the entry to the cardiac chamber without compressing the coronary circulation. If the termination site of the CAF is at the distal aspect of the coronary and no significant myocardium is in

jeopardy, the coronary may be ligated proximal to the termination site. If CAF terminates in the midportion of the left descending coronary artery or right coronary artery, the fistulous communication may be closed with multiple pledgetted sutures (1, 6). The operative mortality for CAF repair is very low – 0–4% (1, 4, 8, 9, 10).

The late results of repair are excellent and very few patients have recurrence (12).

In this article, we describe our experience with multiple CAF in two patients to whom CAF closure under transoesophageal echo (TEE) supervision during repair was performed by using CPB.

MATERIALS AND METHODS

1. Case report

A three-year-old boy was referred to the Latvian State Cardiology Center for Children because of cardiac asymtomatic murmur. The patient had a normal weight and height for his age. His blood pressure was 90/50 mmHg in both arms. The pulses were symmetrical in all extremities and pulse pressure was wide. Palpation of the anterior chest identified a normal impulse and there was no thrill. Auscultation revealed a 3/6 systolic–diastolic murmur in the left second and third intercostal space, and the second heart sound was normal. Chest X-ray showed a slight cardiomegaly with an increased pulmonary blood flow. The electrocardiogram was normal. All other clinical findings were normal. A complete echocardiogram examination was performed, including two-dimensional and M-mode imaging, pulse wave and continous wave Doppler studies, and color Doppler flow mapping. (Fig. 1). A slight increase in the left ventricle diastolic diameter was revealed. There were no hypokinetic segments in the left ventricle. The origin of the coronary artery was normal. The proximal left coronary artery was enlarged. In the apical four-chamber view, color flow mapping showed a short systolic and a large diastolic jet in the apex of the right ventricle. A large aneurysm in the site of drainage was observed.

Surgical repair. After median sternotomy the pericardium was opened. The coronary anatomy was carefully inspected and the distribution of coronary arteries was noted. The left anterior descending coronary artery was enlarged from the very beginning (cross-diameter 7 mm): 20 mm before the apex of the right ventricle and terminal end of the coronary artery an elevated cardiac surface zone 30 × 30 mm was noted. On this area an intensive thrill was localized. The TEE was started, and CAFs were visualized 25 mm from the right ventricle apex and followed with multiple (6–7) small CAFs from the angiomatous malformed region. All CAFs terminated in the right ventricle cavity (Fig. 2).

The ascending aorta and both venae cavae were canulated and CPB was initiated. The heart was observed for signs of ischemia, and the ECG was monitored. A ligature was placed around the coronary artery immediately proximally to the CAF and after the control occlusion (no signs of ischemia and myocardium perfusion remain adequate) the ligature was tied. A second suture ligature was placed quite close to the end of the CAF to ensure obliteration.

Supplementary horizontal pledgeted mattress sutures were placed quite near the angiomatous malformed region where the thrill was localized. TEE control suggested a 100% closure of all CAFs. The postoperative course was uneventful and the patient was discharged home on day 8. Six months after surgery, the patient had no cardiac symptoms or residual cardiac murmur. His chest X-ray electrocardiogram and echocardiogram were normal.

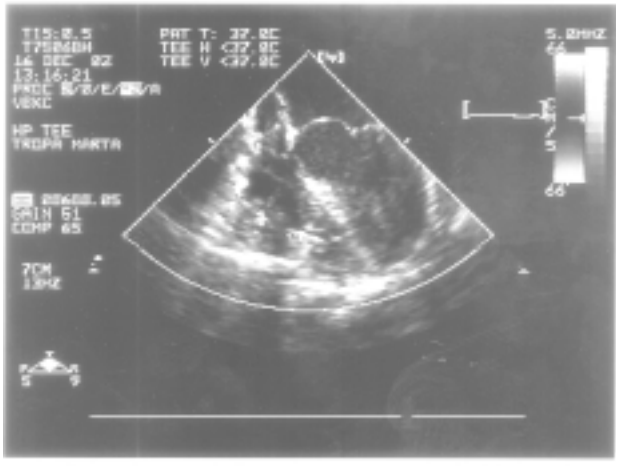

Fig. 1. Perioperative multiplane transoesophageal echocardiography showing multiple coronary artery fistulas from left anterior descending coronary artery into right ventricle

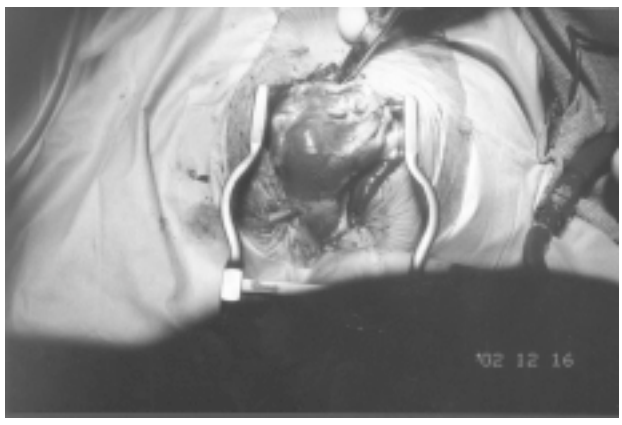

Fig. 2. Perioperative view of the dilated left anterior descending coronary artery

2. Case report

A four-year-old girl was hospitalized in our clinic for assessment following a 4-month history of dyspnea and fatigue on exertion. The patient had a normal weight and height for her age. Her blood pressure was 95/40 mmHg in both arms. The pulses were symmetrical in all extremities and the pulse pressure was wide. Palpation of the anterior chest indentified a normal impulse and there was no thrill.

Examination demonstrated the presence of a continuous murmur of 3/6 grade intensity at the third intercostal space, and the second heart sound was normal. Chest X-ray showed a mild cardiomegaly. The electrocardiogram showed the sinus rhythm with the right axis, right bundle branch block. A complete echocardiogram examination was performed. There was a dilated right ventricle, dilated left coronary arteries with a diameter of 7–8 mm and continuous turbulent multiple flows through a fistulous drainage site into the apex of the right ventricle. There were no hypokinetic segments in the left ventricle. The origin of the coronary arteries was normal.

Surgical repair. After median sternotomy the pericardium was opened. The coronary anatomy was carefully inspected and the distribution of the coronary arteries was estimated. The left anterior descending artery was enlarged and tortuous following its termination in the apex of the right ventricle. Supplementary three enlarged and tortous coronary arteries branched from the right coronary artery and crossed the anterior wall of the right ventricle. Multiple CAF connections from both left (3) and right (3) coronary artery with the right ventricle cavity were visualized by TEE. After cannulation of the ascending aorta and both venae cavae, CPB was initiated. Three CAFs that arose beginning from the middle of the left anterior coronary artery were closed by using horizontal pledgeted mattress sutures, and similar three CAFs on the branches of the right coronary artery were closed (Fig 3). No ischemic signs on ECG were noted. TEE showed the necessity of supplementary sutures since CAF was completely closed. The postoperative course was uneventful and the patient was discharged home on day 5.

Four months after surgery, the patient had no cardiac symptoms or residual cardiac murmur. Her chest X-ray, electrocardiogram and echocardiogram were normal.

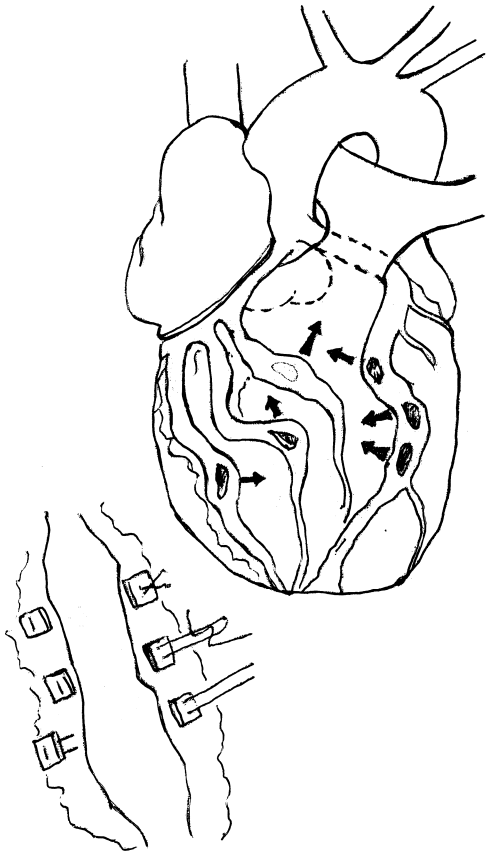

Fig. 3. Perioperative view: schematic information on anatomic findings and suture technique used for CAF closure

DISCUSSION

Most of the CAF directed to ventricular cavities are single, but there is a separate group of patients in whom there is a diffuse sponge work of tiny connections from a number of, if not most, branches of the left (our 1st case) and sometimes the right coronary arteries also (13–15). These presumably represent persistence of embryonic trabecular spaces. Some believe that the prognosis of a surgically untreated CAF is excellent and that operation is indicated only if symptoms are present. However, in view of CAF increase in size and eventual development of symptoms and failure, of the tendency for the development of bacterial endocarditis in these patients, of a very low probability of spontaneous closure (16–19) plus the safety and efficacy of the operation, it is recommended to regard the diagnosis of CAF as an indication for operation unless the shunt is small ($Q_p/Q_s < 1.3$) (6). More active CPB is indicated when the coronary is dilated and tortuous, in order to prevent catastrophic hemorrhage during closure of the CAF. It is also indicated when CAF is in the course of the coronary artery rather than at its termination, so that the CAF itself can be closed without ligation of the coronary artery. In such instances a pledgeted suture ligature is placed around the "feeding" coronary artery very close to the fistulous connection. When the CAF is less clearly localized and consists of a leash of vessels, a secure closure requires a running

suture that encloses all the involved vessels and the underlying wall (6).

Intraoperative (perioperative) and postoperative myocardial ischemia should be minimzed by a prompt intraeoperative detection of any ECG signs. Significant information for the diagnosis and control of 100% closed CAF can be obtained by TEE. The in-hospital mortality for repair of CAF in the absence of giant aneurysm formation approaches zero (6, 8–10, 20–22). A review of the literature by Libertthson and colleagues (11) indicated a postoperative mortality of 4% in 173 patients. Complications of the operation are rare. Myocardial ischemia, either temporary or with infarction, has been reported in 3% of cases and fistula recurrence in 4% (11). However, late results of repair are excellent. Admission of surgery in young patients and in those with small or asymptomatic CAF has been questioned.

In our cases, surgical repair of CAF under the preoperative TEE control was a safe and the effective treatment resulting in a 100% surveyed and 100% closed CAF shunts.

Transoesophageal echocardiography with color flow Doppler facilitated the diagnosis and operative management of this condition, and it has been suggested that it may be of even sufficient accuracy to preclude coronary angiography (11). The current work has shown that intraoperative transoesophageal echocardiography can provide precise information during the intraoperative management of coronary artery fistulae. The clear visualization of the site and extent of the fistulous communication as well as the effect of shunt occlusion on region wall motion permitted the successful ligation of abnormal coronary fistula.

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