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Coronary Intravascular Ultrasound in 2 Children after Cardiac Transplantation

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Coronary intravascular ultrasound imaging has been found to be the most sensitive method for detecting postcardiac transplantation coronary vasculopathy. Techniques for this type of imaging have been well established for adult patients, but coronary intravascular ultrasound imaging in young children has not been previously reported. We describe the cases of 2 children, ages 8.5 years and 9 months, respectively, who had confusing clinical pictures after cardiac transplantation. Currently available imaging equipment and techniques were used in these children to visualize the coronary arteries. The results of ultrasound imaging were considered helpful in determining future treatment for these 2 pediatric patients. ***(Texas Heart Institute Journal 1994;21:310-3)***

Coronary arterial vasculopathy is the major cause of medium- and late-term failure of cardiac allografts;[1,2] it is an obliterative process that eventually results in multiple small myocardial infarctions.[3,4] Because the intimal changes involved in coronary arterial vasculopathy are generally diffuse and concentric, coronary angiography is unreliable for the sensitive detection and quantitation of this problem.[5,6] Coronary intravascular ultrasound (IVUS) has been shown to be significantly more sensitive for detecting the diffuse intimal thickening that is the hallmark of this vasculopathy in adult patients after cardiac transplantation.[7,8]

Post-transplantation coronary artery disease is also known to affect pediatric patients, but the incidence is not as well known. In small patients, technical limitations of invasive studies, especially via the arterial route, have hindered in-depth evaluations. However, recent advances in IVUS and coronary guiding catheter technology, as well as increased experience with coronary arteriography in small patients, led us to believe that IVUS could be performed in young children if necessary. This report describes our techniques and the results of coronary IVUS evaluation in 2 pediatric patients.

Key words: *Child; coronary angiography; coronary disease; coronary vessels; heart transplantation; ultrasonography, intravascular*

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Case Reports

Patient 1

An 8.5-year-old girl, who had undergone cardiac transplantation approximately 3.5 years earlier, was referred to our institution for coronary IVUS imaging. She had been experiencing intermittent significant reductions in ventricular function even though myocardial biopsy results were negative for cellular rejection. The patient was born with double-inlet left ventricle, L-transposition of the great vessels, and pulmonary stenosis. She had undergone a Fontan procedure approximately 4.5 years before the present evaluation. Severe heart failure followed that procedure, and the patient required cardiac transplantation 10 months later. She did well and had only 1 significant episode of rejection during the 1st 12 months. Coronary arteriograms obtained 1 year after transplantation appeared normal, and the patient was weaned from steroids over the next year. A repeat angiogram 3 years after transplantation showed no focal abnormalities. Six months later, however, the patient developed clinical signs of heart failure, and echocardiography revealed a shortening fraction of 18%. Myocardial biopsy showed no evidence of

cellular infiltration, and coronary arteriograms were again negative for focal lesions. The patient responded to a course of high-dose steroids and she was begun on cyclophosphamide treatment. Her ventricular function improved significantly, but there was evidence of apical septal hypokinesia. This confusing clinical picture caused concern that the patient might have occult diffuse small-vessel disease not detectable by standard coronary arteriography.

We performed coronary IVUS in this 49-kg patient using a technique similar to that used in our adult patients. We engaged the left coronary ostium with a 7-Fr JL 3.5 guiding catheter (Cordis Corporation; Miami, Florida, USA). We were able to maintain this catheter in stable position without obstructing the coronary lumen. An initial arteriogram showed no angiographic evidence of focal coronary stenosis. We carefully advanced a 0.014-inch high-torque floppy guidewire into the distal left anterior descending coronary artery (LAD).

The IVUS catheter that we used was a 30-MHz, 3.5-Fr ultrasound catheter (Boston Scientific; Waterton, Massachusetts, USA) interfaced with a dedicated scanner (Hewlett-Packard; Boston, Massachusetts, USA). The catheter was 135 cm long and was designed to be advanced as a monorail over a 0.014-inch guidewire. The imaging transducer was located 20 mm from the tip of the monorail. The ultrasound images were generated at 30 frames per second and were continuously recorded on one-half-inch super VHS videotape while the catheter was in the coronary arteries.

In this patient, we were able to advance the catheter over the guidewire to the distal one third of the LAD. Catheter position was checked with multiple small hand injections of contrast around the IVUS catheter. We did not advance the catheter so far that it would obstruct distal flow. The ultrasound catheter was retracted and the guidewire was manipulated into the circumflex coronary artery. Intravascular imaging was possible throughout the proximal one third of this vessel.

Coronary IVUS images showed normal vessel anatomy (Fig. 1). The intima was quite thin throughout the length of the vessels examined. A brief attempt was made to visualize the right coronary artery. A 7-Fr guiding catheter was used to engage the ostium, and arteriograms showed normal anatomy. The guidewire was advanced to the distal right coronary artery, but the IVUS catheter could not be easily passed over the guidewire without disengaging the guiding catheter. Since the arteriograms and left coronary IVUS images were normal, we elected not to persist. Left ventricular biopsies were obtained to be certain that the absence of cellular infiltrates in previous right ventricular biopsies was not a sampling artifact. No complications resulted from the procedure and the patient was discharged from the hospital the next morning.

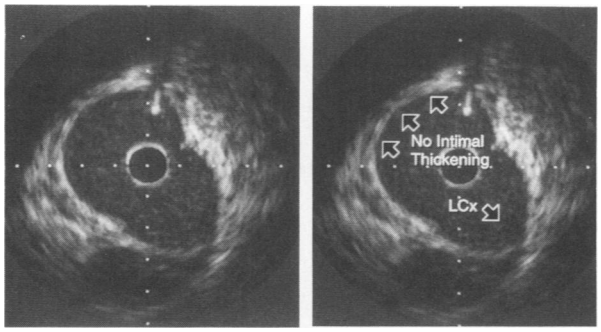

Fig. 1 *Intravascular ultrasound images at the origin of the bifurcation of the left coronary artery in an 8.5-year-old patient. The dark circle in the center of the vessel is the imaging transducer; the bright dot at 1 o'clock is the guidewire artifact. The transducer is in the proximal left anterior descending coronary artery, and the origin of the left circumflex coronary artery (LCx) is seen in an oblique cut (single arrow). There is no evidence of intimal thickening in the left anterior descending coronary artery (arrows).*

Patient 2

A 9-month-old boy who had undergone heart transplantation at 10 days of age was referred to us for coronary IVUS imaging; he was having an episode of decreased left ventricular function with no biopsy evidence of cellular rejection. The patient was born with hypoplastic left-heart syndrome. He initially underwent a Norwood procedure but remained in severe heart failure. He was placed on extracorporeal membrane oxygenation and underwent successful heart transplantation at 10 days of age. Over the next 4 months, he was treated twice for possible rejection; in neither instance was a biopsy performed. At 6 months of age he underwent routine cardiac catheterization. A biopsy at that time showed no rejection, but coronary arteriography showed mild irregularity of the distal LAD, as well as traces of calcium in the proximal LAD. Two months after the catheterization, he again had an episode of reduced ventricular function; biopsy again showed no evidence of rejection. Coronary arteriograms suggested slight additional irregularity and narrowing of the distal LAD.

Because of the patient's small size (8 kg), a 6-Fr sheath and guiding catheter were used. A multipurpose catheter (Cordis) was shaped with the aid of dry heat to approximate the configuration of a JL 1.5 to 2 catheter. This is the smallest catheter we have found with sufficient body to maintain its shape without kinking and still accommodate the 3.5-Fr IVUS catheter. With the addition of anterior angulation to the tip of the catheter, we were able to engage the left coronary ostium in stable position with

no obstruction of coronary flow. A coronary arteriogram showed mild irregularity in the lumen of the mid and distal LAD (Fig. 2). Intravascular ultrasound imaging of the proximal LAD and the circumflex coronary artery was performed as described in the previous patient. It was difficult but possible to perform injections through the guiding catheter with the IVUS catheter in position. Because the monorail tip of the catheter extends 20 mm past the imaging element, particular care was taken to be certain that the catheter tip was not advanced so far distally that it would occlude flow (Fig. 2).

Intravascular ultrasound images revealed mild but definite intimal thickening in the proximal coronary arteries of this very young patient (Fig. 3). The thickening was clearly evident as far as the transducer was advanced in both the main coronary artery and the LAD, but was most apparent at the bifurcation. The patient was discharged the morning after the catheterization. Because of the evidence of intimal thickening in the proximal vessels and the mild angiographic changes in the distal LAD, the patient was started on heparin injections for presumed post-transplant coronary artery disease at the referring institution.

Discussion

At the present time, there are no IVUS catheters designed specifically for pediatric patients. However, these 2 cases illustrate that at least limited studies are technically feasible with currently available equipment. In patients weighing more than 40 kg, it should be possible to apply the same standard techniques and equipment used in adult patients. In very small pediatric patients, we have found that it is necessary to custom-shape a guiding catheter. With current IVUS catheters, the smallest guiding catheter that can be used is 6 Fr. Although the tip of the IVUS catheter that we used extends 20 mm past the imaging element, we found it possible to visualize the proximal portions of the LAD in our 8-kg patient without risking distal coronary artery obstruction.

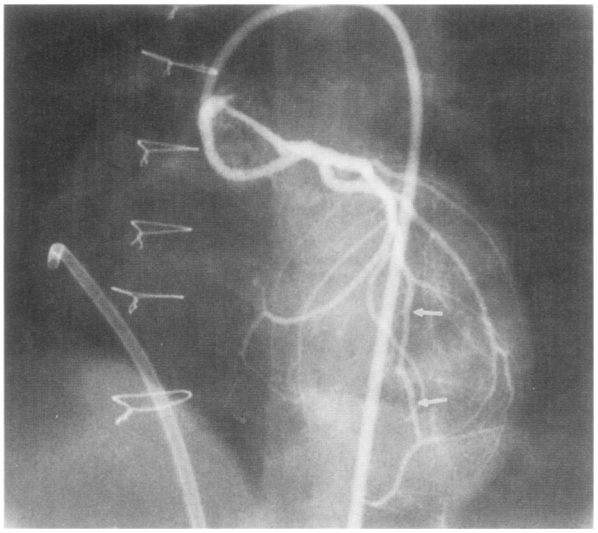

Fig. 2 *Left coronary arteriogram obtained through a 6-Fr coronary guiding catheter in a 9-month-old patient. Arrows indicate mild irregularity of the lumen of the mid and distal left anterior descending coronary artery.*

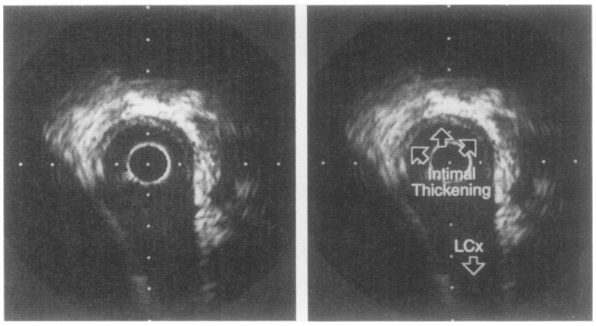

Fig. 3 *Intravascular ultrasound images at the bifurcation of the left coronary artery in a 9-month-old patient. Three arrows in the right panel indicate the thickened intima, which was most prominent in the area opposite the origin of the left circumflex coronary artery (LCx, single arrow).*

Modifications of the catheter design (such as shortening of the monorail tip) will be needed in order to evaluate the more-distal coronary tree in small children, but studies confined to the proximal vessels may be of some benefit. In our 9-month-old patient, the finding of definite diffuse intimal thickening proximally (in angiographically normal portions of the vessels) strengthened the suspicion that mild distal angiographic irregularities of the LAD were not merely artifacts but were indicative of true coronary artery disease. In reviewing the records of more than 120 adult transplant patients from our institution, we have noted proximal intimal thickening by IVUS without significant distal disease, but this is not generally the case when the arteriograms of the distal segments appear abnormal. Similarly, if there is no evidence of intimal thickening in the proximal coronary arteries, it is unlikely that diffuse intimal thickening will be found in more-distal vessels. In their adult transplant patients, St. Goar and colleagues[7] have found that IVUS provides information that may prove to be a marker of prognosis. They also point out that IVUS offers significant potential for monitoring interventional therapy and for improving our understanding of the pathophysiology of post-transplant coronary disease. Further studies are needed to determine whether IVUS in children provides sufficient information about prognosis or treatment to justify more routine use of this

modality in pediatric patients after cardiac transplantation.

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