

basis in hospital inpatients suspected of deep venous thrombosis. The technique is simple to perform, is not operator-dependent, is sensitive to both fresh and aged thrombi and is unaffected by heparin administration. Further work in other patient groups needs to be performed to define the overall clinical utility.

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#### NUCLEAR CARDIOLOGY

## Functional Assessment of Alcapa Syndrome by Dobutamine Stress Thallium-201 SPECT and Echocardiography

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Exercise  $^{201}\text{Tl}$  SPECT has been used as a useful method for the assessment of patients with anomalous left coronary artery communicating to the pulmonary artery (ALCAPA syndrome). In this study, we described an adult patient with this anomaly who was evaluated by dobutamine stress testing in conjunction with simultaneous  $^{201}\text{Tl}$  SPECT and echocardiography before and after surgery. A large perfusion defect in the anterior wall, septum and apex was detected on the preoperative stress scan with partial reversibility on reinjection scan. Worsening of wall motion abnormalities in the septum and anterior wall was detected by stress echocardiography. In the studies performed 3 mo and 1 yr after reimplantation of the left coronary artery in the aorta, a smaller fixed perfusion defect in the anterior wall and apex was detected without reversibility. No stress-induced wall motion abnormalities were detected. Despite the improvement of perfusion, there was no improvement of regional or global left ventricular function at rest. We report that both dobut-

amine  $^{201}\text{Tl}$  SPECT and echocardiography were useful for the detection of reversible ischemia and for the assessment of the surgical outcome of an adult patient with ALCAPA syndrome.

**Key Words:** dobutamine stress echocardiography; thallium-201; SPECT; ALCAPA syndrome

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ALCAPA (anomalous left coronary artery communicating to the pulmonary artery) syndrome is a rare congenital anomaly characterized by an anomalous left coronary artery communicating to the pulmonary artery (1). Most of untreated patients with this anomaly die during childhood from myocardial infarction and heart failure. It is rare when patients survive to adulthood because of the extensive collateralization from the right coronary artery to the left coronary artery (1,2). The detection of myocardial ischemia in patients with ALCAPA is important to identify viable left ventricular (LV) myocardium at jeopardy of irreversible damage. Therefore, exercise thallium

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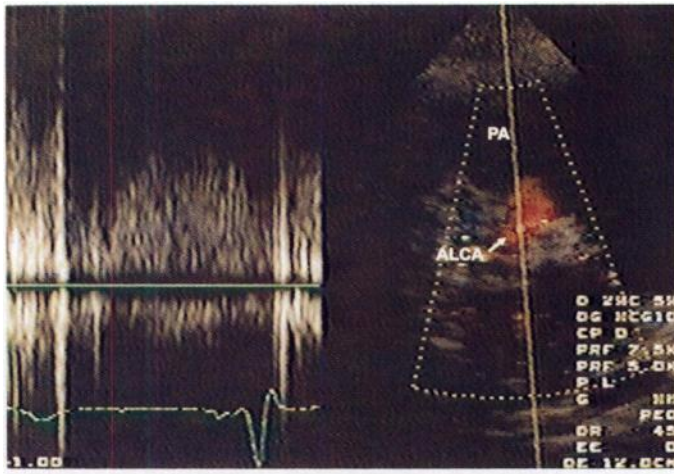


FIGURE 1. Pulsed Doppler sample in the pulmonary artery (PA) shows diastolic flow from the anomalous left coronary artery (ALCA), which is also shown by color flow Doppler.

scintigraphy has been used to assess myocardial perfusion and to evaluate the results of surgery in these patients (3–7). Dobutamine stress test (DST), in conjunction with myocardial perfusion or echocardiographic imaging, is increasingly used for evaluation of coronary artery disease (8–11). In patients with LV dysfunction, low-dose dobutamine echocardiography and  $^{201}\text{Tl}$  scintigraphy are useful for the detection of myocardial viability (12–14). We describe an adult patient with ALCAPA in whom DST, with simultaneous echocardiography and  $^{201}\text{Tl}$  SPECT, was useful for the detection of myocardial ischemia and evaluation of the results of surgery.

#### CASE REPORT

A 35-yr-old woman presented with palpitations and atypical chest pain. Physical examination revealed a grade III/VI diastolic murmur in the second left intercostal space. Baseline ECG showed Q-waves in I-aVL-V6 and poor R-wave progression in precordial leads. Echocardiography revealed a dilated LV, a kinetic apex and hypokinetic anterior septum and anterolateral wall. The left coro-

nary artery was seen coming out from the posterior wall of the pulmonary artery with a diastolic flow into the pulmonary artery detected by continuous and color flow Doppler (Fig. 1). Coronary angiography revealed a tortuous widely-dilated right coronary artery communicating through extensive collaterals with the left coronary artery which was filling the pulmonary artery (Fig. 2A). Gated blood-pool scintigraphy showed hypokinesis of the anterior wall and anterior septum and LV ejection fraction (EF) of 51%. DST with simultaneous  $^{201}\text{Tl}$  SPECT and echocardiography was performed for the detection of myocardial ischemia and viability. Typical angina occurred during the test. A large perfusion defect in the anterior wall, apex and anterior septum was detected on stress imaging with partial reversibility on reinjection. The apex was considered viable on the basis of partial  $^{201}\text{Tl}$  reversibility. Thallium-201 counts in the apex were 9116 at stress and 11,051 units at reinjection, comprising 70% and 85% of the maximal normal counts, respectively. Baseline echocardiogram showed akinesis of the apex and hypokinesis of the anterior wall and anterior septum. During low-dose dobutamine, no contractile response was observed in the apex. At peak stress, worsening of wall motion abnormalities was detected in the anterior wall and septum.

The patient underwent reimplantation of the left coronary artery in the aorta. DST was repeated 3 mo and 1 yr after surgery which revealed a fixed perfusion defect in the anterior wall and the apex without reversibility (Fig. 3). The defect size, however, was smaller compared to the preoperative scan by quantitative analysis. No stress-induced wall motion abnormalities were detected. There was no improvement of regional LV wall motion on the echocardiogram or of global function by gated blood-pool scintigraphy (EF = 43% after 1 yr). The values of EF, quantitative  $^{201}\text{Tl}$  defects and maximal dobutamine stress heart rate before and after surgery are summarized in Table 1. Follow-up coronary angiography performed 14 mo after surgery showed antegrade filling from the aorta of a dilated left coronary artery with slow flow and abrupt reduction of the calliper distally (Fig. 2B). Collateral circulation resolved completely (Fig. 2C). Symptoms of palpitations and atypical chest pain did not improve. The course of the patient was uneventful during a follow-up period of 2 yr, with persistent

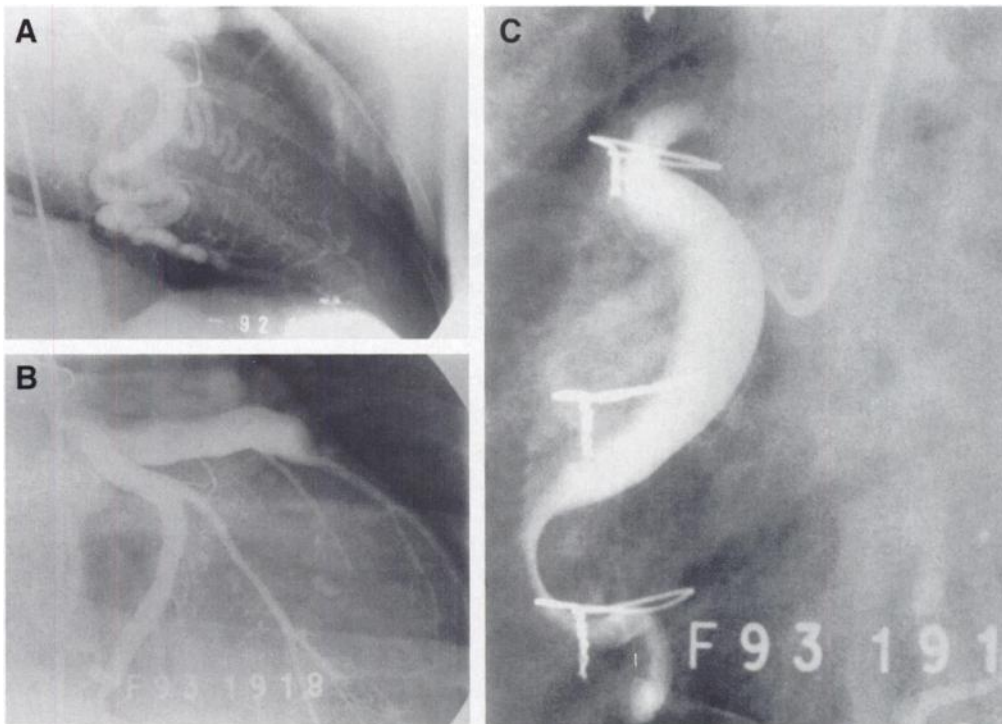
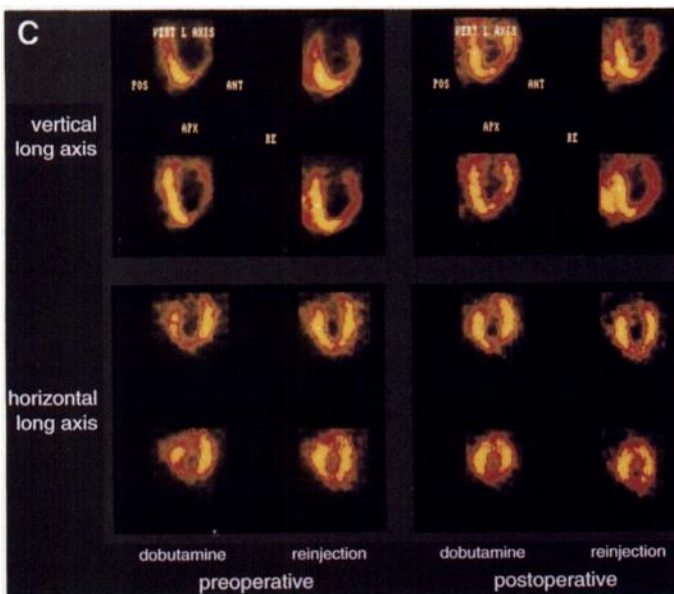
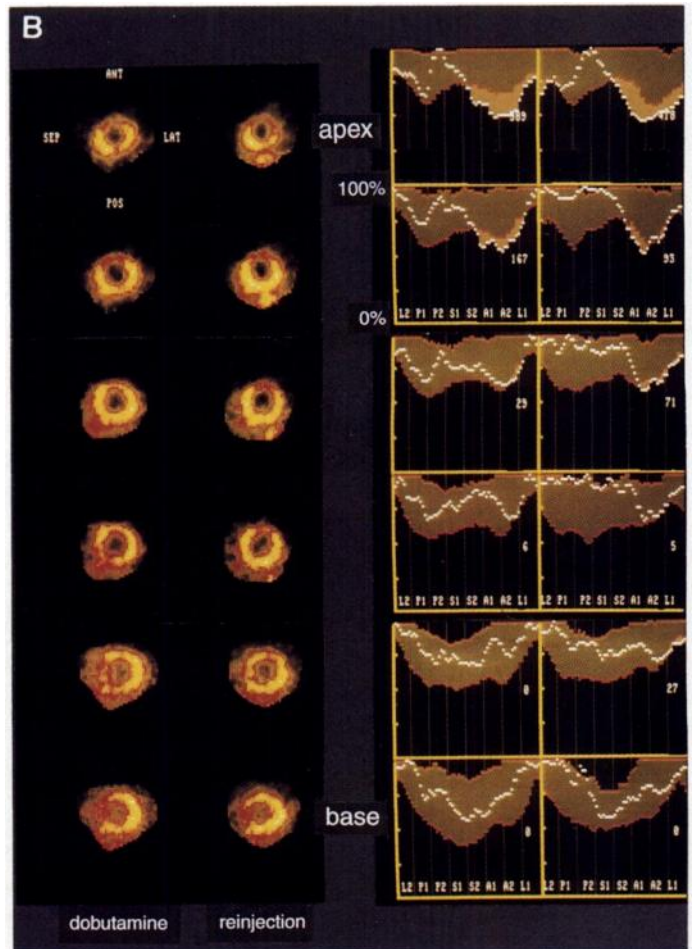
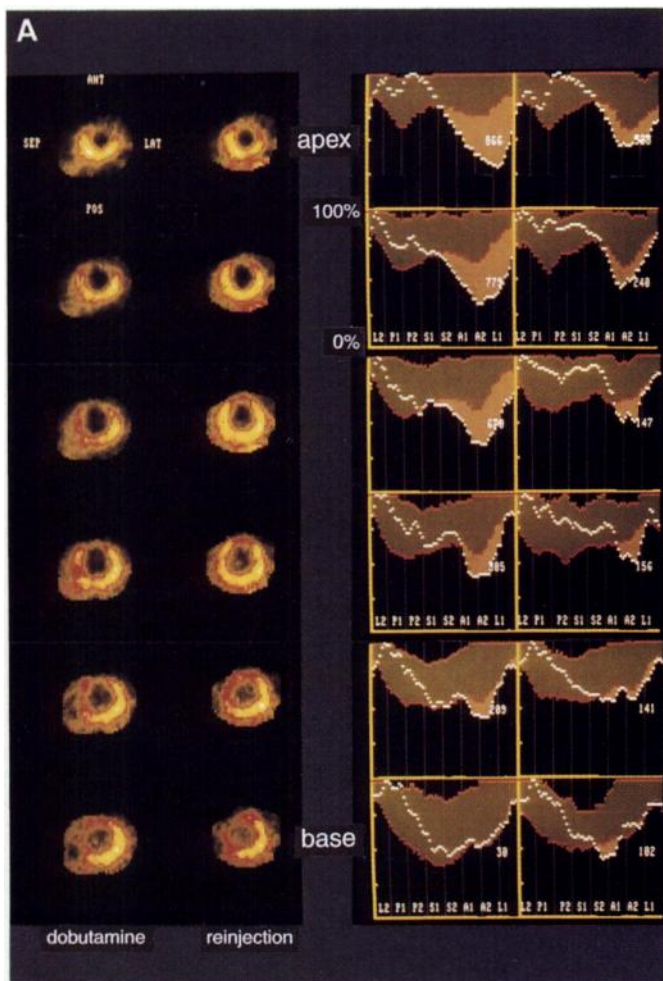


FIGURE 2. (A) Preoperative right coronary injection shows a dilated, tortuous right coronary artery filling the left coronary artery through extensive collaterals with retrograde filling of the pulmonary artery. (B) Postoperative selective left coronary artery injection showing proximal dilatation of the left coronary artery with abrupt calliper reduction distally. (C) Postoperative selective right coronary injection showing disappearance of collaterals and no filling of left coronary artery.





**FIGURE 3.** Dobutamine and reinjection  $^{201}\text{Tl}$  SPECT images. (A) Preoperative images of the six short-axis slices are provided with the corresponding circumferential profile. The images show a partially reversible defect in the apical anterior wall and a completely reversible defect in the septum. (B) Postoperative images show the absence of reversible perfusion defects and improvement of perfusion compared to preoperative images. (C) Images of the long-axis slices show a partially reversible apical defect in the preoperative scan which was absent in the postoperative scan.

regional LV dysfunction on serial echocardiograms. Holter monitoring revealed no high-grade arrhythmias.

#### METHODS

Dobutamine was infused intravenously starting at a dose of 5  $\mu\text{g}/\text{kg}/\text{min}$  increasing every 3 min to 10, 20, 30 and 40  $\mu\text{g}/\text{kg}/\text{min}$ . Echocardiography was performed at rest and throughout the test. One minute before termination of infusion, 80 MBq of  $^{201}\text{Tl}$  was injected intravenously. Images were acquired within 5 min after the

end of the test and 4 hr following the test after reinjection of 40 MBq  $^{201}\text{Tl}$ . Image acquisition and interpretation was performed according to a previously described protocol (8). The diagnosis of ischemia relied upon the occurrence of reversible perfusion defects and new or worsened wall motion abnormalities. The diagnosis of viability in dyssynergic segments relied upon the occurrence of a contractile response during low-dose dobutamine (5–10  $\mu\text{g}/\text{kg}/\text{min}$ ) and the presence of reversibility or a fixed defect containing  $\geq 50\%$  of the maximal thallium uptake.

**TABLE 1**  
LVEF, Quantitative Perfusion Defect Size\* and Maximal Stress Heart Rate before and after Surgery

	Ejection fraction (%)	Stress defect	Reinjection defect	Maximal stress heart rate
Preoperative	51	2879	1314	130
Postoperative (3 mo)	35	661	793	134
Postoperative (1 yr)	43	539	627	143

\*Defect size is a unitless measure of the area between the lower limit of normal values ( $\pm 2$  s.d.) and the actual circumferential profile in the six short-axis slices.

## DISCUSSION

The detection of myocardial ischemia in patients with ALCAPA is important to identify the myocardium at risk of irreversible dysfunction. The combined effect of an increase of myocardial oxygen demand and flow malperfusion induced by dobutamine (15,16) may serve as an appropriate mechanism of eliciting ischemia in patients with ALCAPA, in whom the mechanism of ischemia is a combination of coronary steal into the pulmonary artery and inadequate collateral flow in face of an increased demand (6,17). Since these patients may develop an acquired native coronary artery or graft disease, the ability to establish a two coronary artery system provides an advantage over ligation of the left coronary artery (18).

In our patient, myocardial ischemia could be elicited before surgery during DST as manifested by angina, reversible  $^{201}\text{Tl}$  defect and stress-induced wall motion abnormalities. Echocardiography and  $^{201}\text{Tl}$  SPECT concordantly localized ischemia. The absence of these ischemic markers postoperatively identified a successful surgical correction, which was confirmed by coronary angiography.

A reversible  $^{201}\text{Tl}$  defect without a contractile response to low-dose dobutamine was detected in the apex. This viability pattern on  $^{201}\text{Tl}$  SPECT was predictive of a significant improvement of perfusion postoperatively. No improvement of LV function occurred in a follow-up period of 2 yr. The discrepancy of viability patterns between echocardiography and  $^{201}\text{Tl}$  SPECT may be explained by the high sensitivity of  $^{201}\text{Tl}$  for the detection of small islands of ischemic myocardium which are not capable of restoration of contractility after revascularization of a segment with extensive scarring as we have previously reported (13). The characteristics of chronic ischemia in ALCAPA patients are apparently different from those with atherosclerotic coronary heart disease in terms of the mechanism and duration of ischemia which starts since birth due to a drop of pulmonary artery pressure below coronary perfusion pressure (6). A recent study described a delayed improvement of function up to 3 yr after surgery in children with ALCAPA (19). The lack of improvement of LV function after improvement of perfusion has been attributed to a delayed subcellular adaptive response, impairment of energy production and transfer and altered sensitivity of myofilaments to calcium (19–21). The unique anatomy of the left coronary artery with dilatation

and slow flow proximally and a small calliper distally, shown in the postoperative angiogram, may have an impact on functional recovery. The change of the pressure and direction of flow postoperatively may result in a change of vessel morphology on the long term, and consequently improvement of LV function.

## CONCLUSION

Both dobutamine  $^{201}\text{Tl}$  SPECT and echocardiography were useful for the detection of reversible ischemia and for the assessment of the surgical outcome of an adult patient with ALCAPA syndrome.

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