

## Coronary Artery Bypass Grafting in Idiopathic Myelofibrosis: A Case Report

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### ABSTRACT

The concomitant presence of myeloproliferative disorders and the need for coronary artery bypass surgery is a surgical dilemma. Thrombosis and hemorrhage can cause difficult problems and might require different approaches during and after surgery. We report a patient who had idiopathic myelofibrosis and underwent a successful coronary artery bypass surgery.

### INTRODUCTION

Idiopathic myelofibrosis (IMF) is the least common form of myeloproliferative disorders (MPDs) and is characterized by mobilization of hematopoietic progenitor cells from the bone marrow to the peripheral blood and other extramedullary sites [Wehmeier 1991]. Thrombotic and bleeding complications are frequent findings in MPDs due to complex alterations in blood cells. The majority of bleeding episodes are minor, but the risk of bleeding after cardiac surgery is unknown. We report a case of coronary artery bypass grafting (CABG) surgery in a patient with a history of IMF and psoriasis vulgaris.

### CASE REPORT

A 65-year-old man was referred to our hospital for CABG due to unstable angina pectoris. Coronary angiography revealed severe triple-vessel coronary artery disease (90% stenosis of the proximal left anterior descending artery, 90% stenosis of the circumflex artery, and 70% stenosis of the mid-right coronary artery). His medical history indicated that he had been treated and followed-up for IMF during the previous year and for psoriasis vulgaris for 40 years. His medical history was also significant for diabetes mellitus, hypertension, and hyperlipidemia. He had been treated by hydroxyurea and erythropoietin and with multiple erythrocyte transfusions for anemia. Examination on arrival revealed psoriatic plaques scattered throughout the body. Hepatomegaly and splenomegaly were other obvious findings. Laboratory data included hemoglobin 8.8 g/dL, hematocrit

25.6%, platelet count 519 K/UL, and white blood cell count 5100/ $\mu$ L. Peripheral blood smear revealed an increased number of platelets in every field without forming aggregates, anisocytosis, poikilocytosis, basophilic stripping, fragmented erythrocytes, and reticulocytosis (5%). Caolin-activated thromboelastography (TEG) revealed hypercoagulation pattern with an increased  $\alpha$  angle of 75°, maximum amplitude of 79.9 mm, and coagulation index of 5.3 (Figure 1). Platelet aggregation was tested by optical aggregometry in platelet-rich plasma, and the patient's platelets revealed a depressed aggregation response to adenosine diphosphate (10 mmol/L) stimulation (Figure 2).

Preoperatively, 2 units of erythrocyte suspensions were transfused to increase the hemoglobin level above 10 g/dL. CABG with left internal mammary artery to left anterior descending artery and a saphenous vein graft to right coronary and circumflex arteries was performed uneventfully. Despite a normal TEG and activated clotting time, the patient had a mediastinal tube drainage of 1300 cc during the first 3 hours in the intensive care unit. To exclude a surgical source, the patient was re-explored, but we could not find any origin and blood oozed from all the surgical sites. After re-exploration, the patient continued to bleed, with additional mediastinal drainage of 1500 cc during the following 4 hours. In spite of normal postoperative TEG measurements and platelet counts, a defective preoperative platelet aggregation on the aggregometry guided us to transfuse platelets eventually. After a transfusion of 6 units of platelet suspension, the bleeding suddenly decreased and ceased within 2 hours. The early postoperative course was otherwise uneventful, and he was transferred to the ward on postoperative day 2. He was discharged from hospital on postoperative day 7, and he is now doing well without any problems.

### DISCUSSION

To the best of our knowledge, this is the first case in the literature describing CABG in a patient with a preoperative diagnosis of IMF. Samuels and associates reported a patient with pyoderma gangrenosum and myelofibrosis occurring concomitantly after CABG [Samuels 1997]. Deeb and associates reported incidentally detecting 2 cases of myeloproliferative syndrome during the operations [Deeb 2002]. They performed CABG without using cardiopulmonary bypass (CPB) due to the limited life expectancy and expected poor wound healing in these patients.

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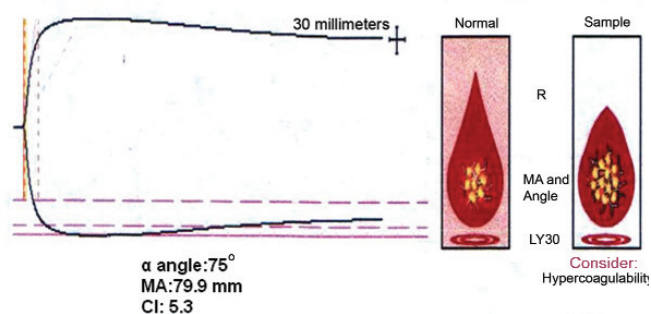


Figure 1. Hypercoagulable thromboelastography pattern taken preoperatively. MA indicates maximum amplitude; CI, coagulation index.

MPDs are characterized by a primary defect at the level of multipotent hematopoietic stem cell, which leads to increased production of 1 or more blood cells [Wehmeier 1991]. Thrombotic and bleeding complications are common during the natural history of MPDs. Thrombosis affects the large arteries of the cardiovascular and cerebrovascular system and may cause fatal myocardial infarction and stroke. In addition, patients with IMF have a higher prevalence of coronary artery disease, mainly due to older age at diagnosis of IMF [Ganti 2003].

Severe symptoms and multivessel disease in our patient made revascularization unavoidable. The two major concerns were bleeding after surgery and early graft thrombosis. The type of revascularization to be used in this particular patient was also debated. Because of the unfavorable long-term prognosis of MPDs and recent advances in stent technology, percutaneous intervention might be a reasonable alternative to CABG in these patients. On the other hand, the chance of early stent thrombosis in this type of patient is unknown. To achieve complete revascularization and avoid possible early hypercoagulation seen after off-pump surgery, we performed on-pump CABG in our case.

The unfavorable effect of CPB on hemostatic alterations such as platelet dysfunction and increased fibrinolysis are common after cardiac surgery [Despotis 2001]. The characterization of the defect is important as a guide to the treatment of the patient. TEG is a sensitive tool in detecting CPB-induced alterations in the hemostatic system, with a negative predictive value of 97% [Ereth 1997]. In the presence of increased postoperative bleeding, normal post-CPB TEG usually indicates a surgical source. Therefore, we re-explored this patient. However, there was no surgical bleeding but the patient continued to bleed after re-exploration until platelet transfusion.

Concomitant presence of psoriasis vulgaris in this case was thought to be incidental, and tests revealed negative autoimmune serology. Psoriasis vulgaris has been reported to disturb the wound healing process during the early postoperative period and cause recurrent cellulites [File 1984]. However, we did not observe any cardiac or skin complications related to psoriasis vulgaris postoperatively.

In conclusion, MPDs in patients undergoing CABG present a challenging clinical situation. Preoperative evalua-

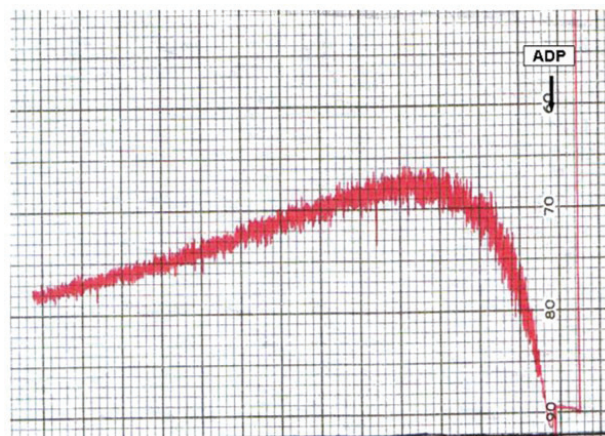


Figure 2. Depressed preoperative platelet aggregation pattern in response to adenosine diphosphate by optical aggregometry.

tion of such patients warrants a demanding work-up of the hemostatic system to prevent life-threatening postoperative bleeding. Special laboratory tests to measure platelet functions such as flow cytometry, aggregometry, adhesion tests, secretion assays, and measurements of platelet activation markers such as  $\beta$ -thromboglobulin or thromboxane  $A_2$  may provide useful information for both the preoperative hemostatic status and postoperative transfusion therapy. In addition, less invasive procedures such as percutaneous interventions or off-pump CABG might be reasonable alternatives in patients with an underlying hematological disease and limited life expectancy. However, as in our case, standard CABG with CPB can be safely performed in these patients.

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