Received:         2012.11.11           Accepted:         2013.01.10           Published:         2013.02.22	Medical management of acute type a aortic dissection in association with early open repair of acute limb ischemia may prevent aortic surgery
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	Summary
Background:	Acute type A aortic dissection (AAAD) is a cardiovascular emergency with a high potential for death. Rapid surgical treatment is indicated to prevent fatal complications. Aggressive appropriate medical management starts at first suspicion and is essential to prevent exacerbation or rupture of the dissection. Despite improved surgical techniques, perioperative care and the development of specialized cardiovascular centers, mortality remains high. Organ ischemia is a catastrophic manifestation of aortic dissection, demanding acute surgical intervention in specialized cardiovascular centers.
Case Report:	We present the case of a 62-year-old man with isolated acute limb ischemia due to an acute type A aortic dissection treated in a regional general hospital, without a specialized cardiovascular service, with immediate open malperfusion repair and aggressive medical management. The patient did not undergo further surgical aortic repair, and after a 30-month follow-up he remains symptom free and in good clinical condition, suggesting that although aortic surgery remains the gold standard for treatment of acute Type A dissection, appropriate medical management and early malperfusion repair may offer an initial limb- or life-saving procedure.
Conclusions:	This staged approach gives clinicians more time to properly evaluate and transfer the patient to a specialized cardiovascular center, and in some cases may even offer a definite treatment.
key words:	aortic dissection $ ullet $ limb ischemia $ ullet $ revascularization $ ullet $ minimally invasive management $ ullet $ medical treatment
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## BACKGROUND

Aortic dissection is a lethal condition that is considered to be the most common aortic catastrophe. Risk factors of this phenomenon are hypertension, thoracic aortic aneurysm, atherosclerotic disease, bicuspid aortic valve, aortic coarctation, and connective tissue disorders. Aortic dissection may be classified chronically into acute (less than 2 weeks from the initial dissection), subacute (2 weeks up to 2 months), and chronic (more than 2 months). The Stanford classification subclassifies the aortic dissections into 2 types. Type A involves the ascending aorta (DeBakey types I and II), but type B does not (DeBakey type III) [1]. The estimated total incidence of acute (type A and B) dissection is 30 to 43 per 1 million population per year. Acute type A aortic dissection constitutes more than 50% of all cases, in which DeBakey type I lesions predominate [2].

The mortality rate for untreated acute type A aortic dissection is 1% per hour up to 48 hours, and up to 90% of patients die within 30 days [2]. The most common causes of death are aortic rupture, myocardial ischemia, acute aortic insufficiency, and malperfusion (cerebrovascular, visceral, and spinal). Emergency surgery is usually recommended, although in certain situations the initial management of malperfusion or conservative therapy can be considered prior to proximal aortic repair. The operative mortality, though, is about 10–20%. This percentage is higher in several subsets of patients, including those with severe neurologic deficits and advanced malperfusion [1,3].

Data from the International Registry of Acute Type A Aortic Dissection in 2012 reveals that in a series of 1809 patients with type A acute dissection, only 3.8% presented mesenteric malperfusion; approximately 30% showed clinical symptoms or signs of neurologic complications, 52.2% had acute renal failure, and 30% had limb ischemia. [4] Although the above-mentioned associated complications may not involve malperfusion as the only underlying pathogenetic mechanism, imaging data, showing extremely high rates of arch vessel (52.9%) and any renal artery involvement (70.6%) by the dissection, support the idea that malperfusion plays an important role and that, when it occurs, it is likely to involve more than 1 vascular territory [4].

Acute type A aortic dissection is highly lethal and may be increasing in incidence. Surgery is believed to save and extend life, but despite apparent advances, diagnosis is often delayed, evidence for improving outcomes is modest, and optimal surgical management remains unclear. Recent reviews have directed limited attention to the provision and performance of surgery [2].

Medical management is part of the initial stabilization of any patient with type A dissection, both during clinical and radiographic evaluation and en route to the operating room. There are, however, situations where the patient's treatment stops with medical management: these are patients with completed stroke, comorbid conditions (eg, cancer, advanced multiple organ dysfunction, age), prior aortic valve replacement (AVR), and presentation to the hospital beyond 48–72 hours of the onset of aortic dissection [3].

The aim of this study is to depict our experience with the management of a patient presenting with acute limb-threatening ischemia due to Type A aortic dissection in a regional general hospital in which the primary goal was to ensure lower limb viability and patient stabilization until final evaluation and treatment by a specialized cardiovascular service.

# **CASE REPORT**

A 62-year-old white man with history of poorly controlled hypertension presented in the emergency department of a regional general hospital during his vacation, with sudden paraesthesia and paralysis of his right lower limb, burning sensation and numbness of his neck, trunk, and upper extremities. The symptoms started 45 minutes prior to his presentation to the hospital. His blood pressure was 200/120 mmHg. On physical examination, the right lower limb was pale, cold, and infrainguinally pulseless, with complete neurological deficit. On Doppler assessment of right anterior and posterior tibial arteries, the signal flow was absent. On the contrary left lower limb, pulses were palpable at all examination levels, and the left Ankle-Brachial Index (ABI) was calculated to be 1.0. Furthermore, pulses were detectable in right and left upper extremities, with no significant difference in blood pressure measurements and no signs of ischemia. Physical examination did not reveal any other remarkable findings. ECG showed no signs of atrial fibrillation and no signs of acute myocardial infarction. Complete blood count tests, creatine phosphokinase, and lactate dehydrogenase, were within normal values. Blood creatinine was 1.3 mg/dl. The patient was diagnosed with complete neurologic deficit due to acute limb ischemia (ALI) and malignant hypertension with a strong suspicion of local (iliac) or acute aortic dissection.

In the absence of a functional computed tomography (CT) scanner in the hospital during the last 3 months (due to hospital budget cuts), transthoracic echocardiography was performed, which showed normal aortic root diameter, as well as normal values of left atrial and ventricular dimension, but no signs of dissection. Since complete neurological deficit due to acute limb ischemia was already diagnosed the consultant vascular surgeon decided to proceed to immediate limb reperfusion.

Surgical exploration and assessment of the right common femoral artery was initially performed. No signs of thrombosis or atherosclerosis were detected proximal or distal to the right femoral bifurcation. Because insufficient inflow was observed, a left to right femoro-femoral crossover bypass was performed using a 7 mm PTFE ringed graft. Although the hospital was equipped with an angio-suite, diagnostic digital subtraction angiography and endovascular interventions were unavailable due to hospital budget that are part of the national austerity program. An immediate improvement of blood perfusion of the right lower limb was noted, with restoration of palpable peripheral pulses and complete recovery of the neurologic deficit. High blood pressure was treated and controlled in the ward during the postoperative period with appropriate medication, which included intravenous beta-blockers and per os calcium channel blockers. A small deterioration of renal function (Cr=1.6 mg/dl) was observed during the first postoperative day, which returned to preoperative values on the third postoperative day; aside from this, the patient's postoperative

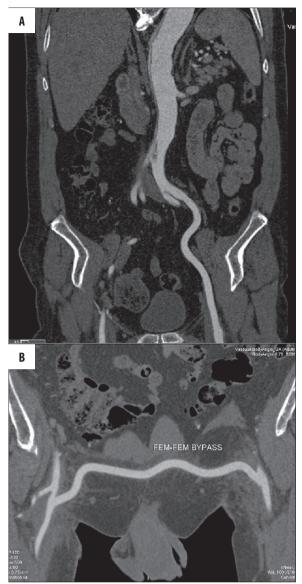


Figure 1. (A) The dissection flap terminates into the left internal and external iliac arteries. (B) Crossover fem-fem bypass from left to right restorates blood flow to right limb.

clinical course proceeded uneventfully. The patient was not admitted for aggressive blood pressure control at the ICU post-operatively because diagnosis of acute aortic dissection was not established.

Due to severe fiscal difficulties, which compromised the ordinary hospital function and unavailability of imaging modalities (CT angiogram, absence of transesophageal ultrasound, functioning angio-suite), and in order to establish diagnosis, the patient was referred to a private clinic for an emergency CTA on the day of admission, but the patient refused the examination and was discharged on the fourth postoperative day on his own volition in good clinical condition in order to continue his scheduled vacation, despite the treating physicians strong recommendation for further clinical and imaging investigation.

The medical treatment prescribed consisted of calcium channel blockers, beta blockers, aspirin, and statins. A CT

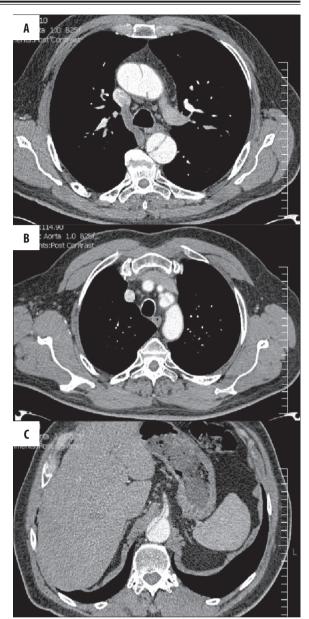


Figure 2. Computer tomography angiography performed 15 days after emergency reperfusion of the lower limb revealing:
(A) A dissection flap in the ascending aorta which extends down to the descending thoracic aorta. (B) The dissection flap continues for a short distance into the left common carotid and left subclavian arteries without causing any symptoms. The brachiocephalic artery is not involved.
(C) Within the abdominal aorta, the dissection flap extends into the celiac axis.

angiogram was performed after finally persuading the patient, 20 days after his discharge; it revealed a type A aortic dissection with a dissection flap (entry point) arising from the mid ascending aorta and extending into the left internal and external iliac arteries (Figures 1 and 2). It is most likely he suffered the dissection during initial presentation with acute limb ischemia. The patient denied readmission to the hospital or transfer to a specialized cardiothoracic center in Greece and returned to his country (the United Kingdom) where he then visited the local specialized center.

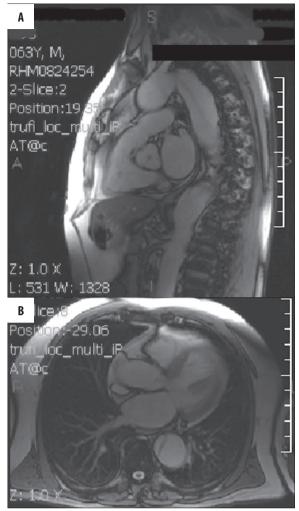


Figure 3. MR Angiogram 3 months later showing no significant change in the appearance of the initial dissection.

The patient is under the care of the specialists of his regional Cardiovascular Center and is continuing his initial medical treatment with statins, aspirin, and strict control of blood pressure (systolic BP <120 mmHg) using calcium channel blocker, angiotensin receptor blocker, and beta blocker. No further open aortic repair has been required. Follow-up imaging studies with CT and/or MR angiogram performed regularly shows a stable dissection and the patient remains symptom free (Figure 3). Thirty months after initial treatment the patient is stable and has no further need for surgical treatment.

## DISCUSSION

Acute aortic dissection involves blood flow through an intimal tear into the aortic media of an often weakened aortic wall, resulting from degeneration (eg, atherosclerosis, aging, hypertension) and/or genetic predisposition (e.g., Marfan syndrome). Treatment principally consists to limiting propagation of the false lumen with all negative consequences on end-organ perfusion, by reducing and stabilizing the hemodynamic stress on the aortic wall.

Surgical repair is preferred for type A dissection. Medical therapy, focused on the use of antihypertensive agents, is

generally used to achieve this goal in uncomplicated/stable type B dissection. Anti-hypertensive therapy remains the cornerstone in maintenance of hemodynamic stability during follow-up to promote aortic stability and prevent aortic expansion with possible rupture and recurrent dissection. The effects of medications on the outcomes in patients with aortic dissection have been examined using the IRAD database [5], which reports that 96% of patients with aortic dissections are discharged with antihypertensive medication. In all of them, as well as in those with type A overall and in those with type A treated surgically, the IRAD study has shown that administration of  $\beta$ -blockers, the most commonly used agent in patients with a ortic dissection (88.6%), are associated with improved survival [5]. In contrast, for those with type B overall and for those treated medically, the IRAD study showed that administration of calcium channel blockers were selectively associated with improved survival, but renin-angiotensin system inhibitors were not significantly associated with survival [5].

It has been believed that without surgical therapy, acute type A aortic dissection was nearly invariably lethal, with a stated mortality of 1% per hour and an expected 90-day mortality of 70% to 90% [6,7]. With modern ICU care, anti-impulse therapy, and medical management, it is becoming apparent that survival rates are higher than previously thought [3].

Feldman et al achieved a hospital survival rate of 88% in patients who underwent interval or permanent non-operative management, and 80% in patients who underwent entirely non-operative management. The authors indicate that the survival over the first 2 years was essentially equivalent in the immediate surgery, delayed surgery, and exclusively medical groups. They pointed out that this data did not deter any enthusiasm for urgent surgery as a general principle for acute aortic dissection. Rather, they wished to indicate that, when necessary, interval or exclusive medical management can produce better results than previously expected [8]. Interestingly, with regard to patients who present later than 48 hours after the initial onset of pain, a 10-year follow-up study by the same group has shown that medical and surgical treatment modalities resulted in similar survival, but there is a trend towards better survival in the surgical group [3,9,10].

Most procedures for type A dissections involve replacement of the ascending aorta under profound hypothermic circulatory arrest. The extent of distal repair is determined by the pathology of the dissection. It is generally accepted that the goal of type A aortic dissection repair is to resect the area of the primary intimal tear, in order to achieve occlusion of the false lumen, preventing further development of descending aortic aneurysms [11]. Aortic valve preservation is usually attempted by commissural resuspension or local repair unless aortic valve disease and coronary malperfusion exist. Recent work using stent grafts has been reported [12]. Most patients who have type A dissection, and malperfusion and who undergo early repair, demonstrate resolution of the malperfusion by proximal repair alone. However, patients with malperfusion and ischemic end-organ dysfunction or acute limb ischemia are an extremely high-risk group; the occurrence of malperfusion predicts poorer long-term survival and higher early mortality [3,13,14].

The aim of this case report is to raise concern about the treatment of acute type A aortic dissection after early and proper repair of limb ischemia in an otherwise stable patient. In current practice, aortic repair is almost always recommended (either emergency or delayed). In our case, the patient is completely asymptomatic 2 years after the initial dissection and immediate blood restoration of the limb. According to repeated CT and MR angiograms, which revealed no overall changes in the appearance of the dissection in association with complete blood flow restoration of the limb and patient's good clinical condition (absence of neurologic deficit, chronic renal failure, chronic limb ischemia, or signs of intestinal malperfusion), the cardiologists and cardiothoracic surgeons who took over his care after returning home suggested that there is no need for further surgical intervention. The efficient control of blood pressure and heart rate, as well as the absence of further risk factors, is of paramount importance. Thus, taking into account that the perioperative risk for elective aortic repair is quite high, and since the patient remains asymptomatic, it is important that we reconsider the role of conservative (medical) treatment after early limb ischemia repair in acute type A aortic dissection as acceptable management in patients who have survived the initial perilous period.

Finally, 2 years later, the Thoracic Surgery Department and the angiography suite in this regional general hospital are out of service and the medical team has been dispersed elsewhere. The global financial crisis, in association with the national fiscal deficit, is having a serious impact on the Greek national health care system. The introduction of "linear" cuts (justified by the risk of a debt default) in the allocation of resources creates a devolution process in health fiscal policy, which clearly increases inequalities in access to health rate in the southern Europe [15]. Deterioration of health indicators and the population's welfare is sharpening the already existing differences in the quality of care, not only between different regions of the country, but also between Greece and the majority of Western nations.

The global financial crisis, which started in 2008 and is still ongoing, may have considerable impact on government budgets and the funding available for health services. Past economic recessions (especially the 1997 Asian financial crisis) have shown that the impact on public health can be severe [16]. Buysse et al, in their report for the World Health Organization, found that the economic recession has had a mixed effect on pharmaceutical consumption, expenditures, and prices. The largest changes have occurred in highincome countries and in Europe. They conclude that this recession provides an opportunity to identify which policy approaches most effectively prevented or contributed to declines in pharmaceutical consumption [16]. This may work for government and health organizations aiding in reducing unnecessary costs. On the other hand, linear cuts across the board on all public health disciplines may be dangerous, and will eventually have a negative impact on public health. Borowy reported that the collapse of the Soviet bloc, which caused a devastating economic crises in the Russian Federation, triggered negative public health responses, with a substantial increase in mortality rates [17]. Falagas et al reported that economic crises in less affluent countries are accompanied with an increase in all-cause mortality, as well as mortality from most of the major specific causes [18].

The above-mentioned association could be attributed to increased psychosocial stress during such periods, among other factors. Economic depression has been found to increase suicides [19]. Stuckler et al reported that every 1% increase in unemployment was associated with a 0.79% rise in suicides, and a greater than 3% increase in unemployment had an even greater effect among people younger than 65 years of age [19].

Physician awareness and proper management is important for good clinical outcomes in pathologies with very high mortality [14]. Despite the remarkable daily efforts of the medical staff, the Greek national healthcare system experiences new major reductions, not only in investments for preventive medicine, evidence-based medicine infrastructures, health information systems, and physical capital renewal, but also in the funds necessary for its ordinary daily functioning. Waiting lists, continuity of health care and patient's centeredness, and integration between social and health care are all negatively affected by resource cuts.

The public health authorities should be aware of this issue and consider appropriate preventive and control measures. In the case of Greece, where there is an increased demand for hospital budget cuts by the austerity program and adequately staffing all hospitals is not an option, we propose concentrating specialized personnel in appropriate specialized centers and developing a very efficient ambulance/transport service to connect outlying areas with the specialized centers.

# CONCLUSIONS

Surgery remains the gold standard for treatment of acute type A dissections, but in cases presenting with isolated lower limb ischemia, surgical reperfusion may ensure limb viability until the patient can be transferred to a specialized center. In selective cases, limb reperfusion may even offer a long-term solution, in association with intensive and appropriate medical management at first suspicion.

The recent financial crisis negatively affects the incidence of diseases due to increased inequalities in access to health care.

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#### **Conflict of interest**

None to declare.

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