Percutaneous cardiopulmonary support for circulatory assistance during angioplasty in a patient with Friedreich's ataxia

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Abstract

The case of a 70-year-old-woman affected by Friedreich's ataxia (FRDA), unstable angina and heart failure with left main 'equivalent' lesions is presented. As the patient was in need of revascularisation but would have been at high risk with coronary artery bypass grafting surgery, she underwent coronary angioplasty assisted with percutaneous cardiopulmonary support (PCPS). Despite the onset of temporary complications, the procedure was performed successfully. On the basis of this case and of a review of pertinent literature, the authors discuss the role of PCPS in high-risk patients, with respect to its indications and performance technique.

Key words: percutaneous transluminal coronary angioplasty, Friedreich's ataxia, extracorporeal circulation, percutaneous cardiopulmonary support.

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Introduction

PCPS-assisted percutaneous transluminal coronary angioplasty (PTCA) is usually adopted for high-risk patients who have contraindications to general anaesthesia or severely impaired left ventricular function. Occasionally it may be of help also in cases of inherited neurological disorders.

Case report

A 70-year-old woman affected by type 2 diabetes, hypertension and dyslipidaemia presented with a history of a recent (four months previously) Q-wave myocardial infarction of the anterior wall and heart failure. The patient was affected by Friedreich's ataxia (FRDA), recognised at the age of 25. The diagnosis had been confirmed by genetic testing and by magnetic resonance imaging of the cerebellum and spinal cord.

Clinical examination revealed signs and symptoms compatible with unstable angina: chest pain and dyspnoea at rest, tachypnoea, tachycardia, gallop rhythm, mild hepatomegaly and an increase of plasma creatine kinase MB-isoform and myoglobin. Neurological examination evinced evidence of gait ataxia and dysarthria, lower limb tendon areflexia and loss of vibration sense. Chest X-ray showed cardiomegaly and mild basal effusions. Surface ECG revealed Q waves in leads V1 to V3 and repolarisation changes in the lateral and inferior leads. M-mode and two-dimensional cardiac ultrasonography revealed anterior and apical dyskinesia, minimal mitro-aortic regurgitation and an ejection fraction estimated at 50%. The test was, however, negative for increased septum or left ventricle wall thickness, hypertrophy of papillary muscles or reduction of cavity size, all these characteristic of FRDA-related cardiomyopathy.[1]

Coronary angiography revealed proximal occlusion of the left anterior descending artery and severe stenosis of the proximal portion of the circumflex artery and of the first obtuse marginal branch. The right coronary artery was dominant and presented a subocclusive plaque at the origin of the posterior descending artery (figure 1a). Major congenital abnormalities of the coronary vessels were excluded.

The patient underwent percutaneous coronary revascularisation via catheterisation of the left femoral artery and right femoral vein. Femoro-femoral percutaneous cardiopulmonary support (PCPS) was established for left ventricle assistance during the procedure. The left femoral artery and the contralateral vein were cannulated by 'Bio-Medicus' 19-Fr arterial and 17-Fr venous cannulae, respectively, and connected to 3/8 venous and arterial lines (figures 2a and 2b). The extracorporeal circulation was achieved using a centrifugal Bio-Pump® (Medtronic, Inc.) and an Avecor membrane oxygenator (model: I-3500-2A) (figure 3). Heart rhythm and dynamic pressures were monitored carefully using a central venous catheter, a Swan-Ganz catheter placed in the pulmonary artery and a peripheral arterial catheter placed in the right radial artery. The average flow rate was 2.2 L/min; arterial blood pressure was maintained above 70 mmHg throughout the procedure.

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Figure 1. Coronary angiography before **a**: and after **b**: angioplasty

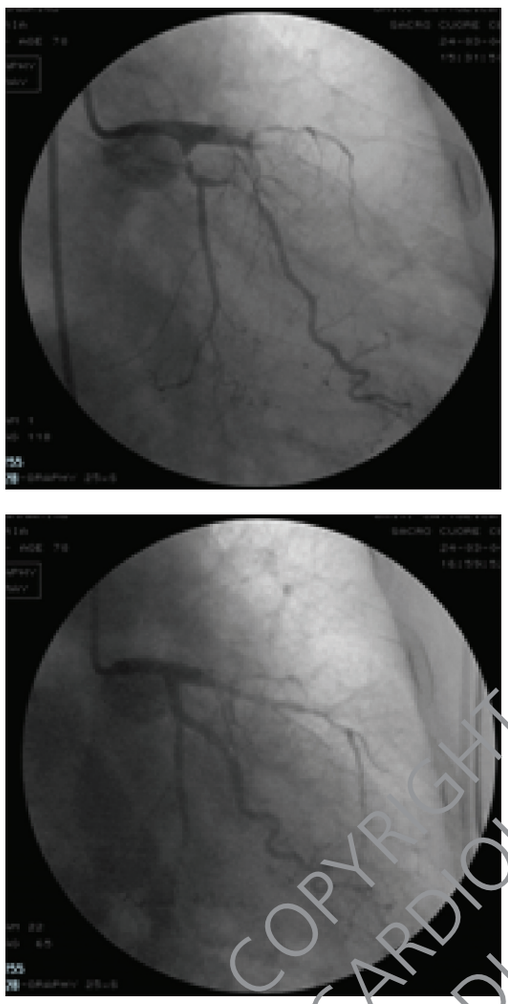
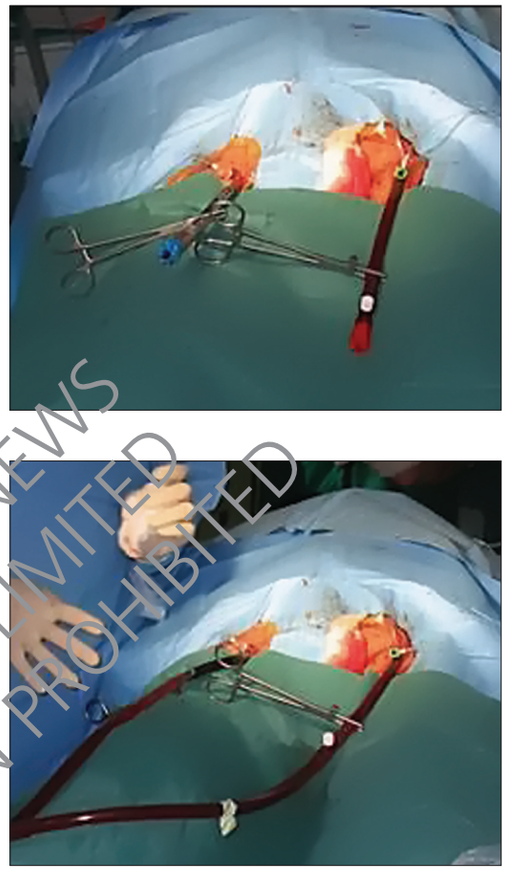


Figure 2. Cannulation of the femoral vessels (**a**) and connection to venous and arterial lines (**b**)



The target lesions were recanalised with balloon dilatations at up to 10 atmospheres and successfully stented (figure 1b). Episodes of intraprocedural ventricular fibrillation and ventricular tachycardia were reversed by an asynchronous 120 J electric shock and by multiple synchronous 120 J shocks, respectively. Pharmacological inotropic support was administered (dopamine 5 µg/kg/min; epinephrine 0.4 µg/kg/min), and the patient was weaned successfully from PCPS (total PCPS time was 48 minutes).

The patient's course in the intensive care unit (ICU) was characterised by acute pulmonary oedema, need for prolonged administration of inotropic drugs and the onset of acute renal failure (peak blood creatinine 2.8 mg/dl and blood urea nitrogen 30 mg/dl). This responded well to frusemide and the patient did not need dialysis. Inotropic support was suspended on the ninth postprocedural day. ECG revealed normalisation of repolarisation in the anterior and inferior leads; echocardiography showed recovery from apical dyskinesia and no pericardial effusion. Since the patient had sustained a significant intraprocedural blood loss (1,200 ml), three units of homologous blood were transfused until optimal haematocrit was achieved. The haemodynamic status progressively normalised, and the patient was disconnected from the mechanical ventilator during the 17th day after PTCA. There was no recurrence of angina and the patient was discharged from the ICU.

At one-year follow-up, the patient is in New York Heart Association (NYHA) class II heart failure and there has been no echocardiographically documented progression of cardiomyopathy.

Discussion

To our knowledge, there are no previous reports in the literature describing elderly patients affected by FRDA and ischaemic heart disease undergoing PCPS-assisted PTCA. FRDA is an autosomal recessively inherited spinocerebellar degenerative disease, frequently associated with cardiomyopathy (up to 90% of cases) and impaired glucose tolerance.[2]

Figure 3. Heart-lung machine employed for percutaneous cardiopulmonary support

Our patient is unusual in that she had reached an advanced age despite FRDA and she presented with severe ischaemic heart disease in the absence of clear signs of cardiomyopathy. Cardiac involvement shows a broad spectrum of phenotypes in FRDA patients,[3] and ECG/neurological features do not correlate with its severity.[4] Myocardial energetic abnormalities are present even in patients without evidence of cardiomyopathy,[5] and in the case presented here this factor was at least likely to worsen the effects of coronary artery disease.

Even if there are no definitive reports that vasospasm and congenital coronary abnormalities associated with FRDA may cause angina,[6] onset of ischaemic cardiac manifestations should be carefully monitored in these patients, as they may progress rapidly and catastrophically. Our patient was actually at high risk and not eligible for CABG despite her severe and multivessel disease; at the same time she could not be given PTCA alone due to her angiographic lesions (involvement of both left anterior descending and circumflex arteries). Muscle relaxants are, moreover, a potential source of complications in FRDA. Though there is a higher risk of minor complications (local haematoma and blood loss), PCPS under regional anaesthesia is reported to enhance the safety of PTCA in high-risk patients.[7-11]

In our centre, prophylactic PCPS is usually used for assistance to PTCA according to the following indications: a) left ventricular ejection fraction lower than 30% plus severe coronary disease in regions other than the primary infarcted/ischaemic area; b) target vessel(s) supplying more than 50% of the still viable myocardium; c) if the patient refuses surgical revascularisation or has prohibitive surgical risk. One advantage is that PCPS ensures both myocardial and systemic perfusion in the event of intraprocedural arrest and ventricular failure. Further, in the event of failed percutaneous revascularisation the procedure can be converted immediately to surgical coronary intervention in the catheterisation laboratory itself. PCPS can be prolonged if ventricular failure is protracted after CABG.

We used PCPS instead of aortic counterpulsation in this patient, given the high risk of intraprocedural arrest; in the event of ventricular fibrillation the heart rhythm-triggered balloon pump would probably have arrested too. There are reports in the literature that aortic counterpulsation may be not sufficient for patient recovery during cardiogenic shock; in such a setting emergency PCPS can be extremely beneficial.[12] PCPS can provide full cardiopulmonary support, including blood oxygenation and venous drainage, until the heart rhythm recovers or the procedure is converted to an open-chest procedure.

Some authors claim higher rates of vascular complications related to PCPS, arguing that the intraaortic counterpulsator can be easily inserted and left *in situ* for longer periods. The concept that PTCA success rates are improved by PCPS in comparison to balloon counterpulsation alone is supported by evidence from the literature.[8] Further, the advantage in terms of procedural safety seems established.[9] There is disagreement on whether the adoption of PCPS is effective in reducing postprocedural major cardiac events.[8] In our opinion, operators will feel more confident during the balloon inflation phase if prophylactic PCPS is instituted, rather than counterpulsation alone, in those individuals at highest risk.

The availability of standby PCPS during all coronary procedures has been recommended,[11] but whether to use standby or prophylactic PCPS for high-risk individuals is another area of debate. Standby PCPS has to be activated promptly if the patient develops complications during the procedure, but time is needed for cannulation of vessels and circuit priming. This procedure carries a 'latency time' before cardiopulmonary support is fully working, potentially placing the patient at risk. Therefore, although there is consent that prophylactic PCPS should be insti-

Key messages

- We describe the case of a patient with Friedreich's ataxia who underwent coronary angioplasty assisted with percutaneous cardiopulmonary support (PCPS)
- We employ prophylactic PCPS for indications that include target vessel(s) supplying more than 50% of the still viable myocardium and prohibitive surgical risk
- One area for debate is the use of standby or prophylactic PCPS in coronary procedures

tuted when the procedural risk is highest, there are no broadly accepted indicators of highest risk. Some authors propose the threshold of left ventricular ejection fraction < 20%.[7]

The patient presented here should be included in a separate category in any case, as neurological co-morbidity placed her at highest risk. The impact of Friedreich's ataxia on the choice of prophylactic PCPS is still to be quantified.

At the present moment, it remains unknown whether relief of ischaemia in our patient has prevented or slowed the progression to cardiomyopathy, or even delayed the need for transplantation. This case suggests that PCPS-assisted angioplasty is safe in the elderly with degenerative spinocerebellar disease and compromised cardiovascular function; it relieved symptoms and almost certainly lengthened the patient's life expectancy. PCPS played a pivotal role in the success of the stenting procedure, by allowing longer times of balloon dilatation, reducing pre- and afterload so decreasing myocardial oxygen requirement and finally avoiding intra-procedural circulatory arrest. In fact, PCPS represents the best method of temporary circulatory support in intraprocedural cardiac arrest or major arrhythmias.

In conclusion, the indications for prophylactic PCPS need to be further refined. Studies on large cohorts having as the primary end point the analysis of the effects of standby/prophylactic PCPS on the risk factor profile would probably suffice. These effects should be assessed in both the general population undergoing PTCA and in individuals characterised by neurodegenerative disorders.

Conflict of interest

None declared.

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