Fontan conversion to total cavopulmonary connection and arrhythmia ablation: clinical and functional results

G Agnoletti, A Borghi, G Vignati, G C Crupi

Objectives: To assess the early results of conversion from atriopulmonary to total cavopulmonary connection in patients with failing Fontan operation.

Design: Early clinical and instrumental evaluation of patients undergoing conversion from atriopulmonary to total cavopulmonary connection from April 1999 to November 2000.

Setting: Tertiary referral centre for congenital heart disease.

Patients: 11 Fontan patients (mean [SD] age 20.9 (6.7) years) with refractory arrhythmias or ventricular dysfunction.

Interventions: Total cavopulmonary connection, intraoperative ablation, and AAIR pacemaker implantation.

Main outcome measures: Holter monitoring, transoesophageal atrial stimulation, ergometric test, and myocardial scintigraphy at a mean [SD] follow up of 16.8 (5.6) months.

Results: One early postoperative death occurred. During follow up three patients had relapse of atrial tachycardia, controlled by medical treatment, and two were pacemaker dependent. Transoesophageal stimulation did not induce atrial tachycardia in any patient. Ergometric test showed a diminished exercise tolerance in all but one patient. Mean minute ventilation and maximum oxygen consumption were 62% and 40% of their respective predicted values. Myocardial scintigraphy showed reversal of rest or exercise dysfunction in five patients and improved systemic ventricular function in seven. Mean basal ejection fraction increased from 39.4% (95% confidence interval [CI] 32% to 46%) to 46.5% (95% CI 41.7% to 51.2%) and ejection fraction on effort from 42.3% (95% CI 33.9% to 50.7%) to 50.2% (95% CI 44.5% to 55.9%).

Conclusions: Our data show that total cavopulmonary connection associated with intraoperative ablation and pacemaker implantation allows for better control of arrhythmias and improves ventricular function in the majority of patients with failing Fontan.

Fontan operation, first introduced in 1971, has undergone several modifications aimed at diminishing or relieving right atrial enlargement and avoiding the presence of slow, turbulent systemic venous flow, which are the usual late effects of the classic Fontan operation. It is known that patients undergoing the Fontan operation can develop several complications caused by right atrial enlargement, increased systemic venous pressure, and slow, turbulent venous flow, such as atrial arrhythmias, compression of the right pulmonary veins, coronary sinus hypertension, atrial thrombi, and pulmonary embolism. These complications can, in turn, have grave consequences; thus, arrhythmias can lead to myocardial failure and development of atrial thrombi, pulmonary vein compression can cause low cardiac output and pulmonary hypertension, and coronary sinus hypertension can induce myocardial impairment. All these possible events make the late Fontan patient a complex patient, often difficult to treat either medically or surgically.

The present practice of total cavopulmonary connection, introduced in 1990 by Marcelletti, excludes the right atrium from the systemic venous circuit, preserves a laminar systemic venous flow, and is expected to decrease the incidence of atrial arrhythmias and myocardial impairment.

Recently, conversion from atriopulmonary to total extracardiac cavopulmonary connection has been shown to improve myocardial function and functional class and to control refractory arrhythmias in most patients. Whether this operation should be routinely associated with antiarrhythmic ablative procedures such as atrial cryoablation or radiofrequency ablation and atrial pacemaker implantation is still being debated.

From April 1999 to November 2000, 11 patients had conversion of atriopulmonary anastomosis to total cavopulmonary connection associated with antiarrhythmic procedures. We review this experience.

METHODS

Patient population
There were six male and five female patients. The mean (SD) age at reoperation was 20.9 (6.7) years. The mean (SD) time interval between initial Fontan and total cavopulmonary connection was 15.6 (2.5) years. All patients had first time conversion. One patient (patient 7) with double inlet left ventricle and discordant ventriculoarterial junction had atriopulmonary anastomosis and resection of subaortic stenosis.

Table 1 lists the clinical characteristics of the patients, including primary diagnoses, previous Fontan connection, New York Heart Association (NYHA) functional class, and type and onset of arrhythmias. Table 2 lists the haemodynamic data.

Indications for reoperation were as follows.

- Functional class higher than II—for four patients were in class III and one (patient 9) developed transient congestive heart failure during atrial arrhythmia.
- Debilitating atrial arrhythmias—eight patients had intraatrial re-entry tachycardia and two suffered from episodes of atrial fibrillation. Medical treatment was partially or completely ineffective in six and three patients, respectively. Endocardial radiofrequency ablation had been attempted, without success, in two patients (3 and 6), and two (5 and 9) required multiple cardioversions. Two patients with atrial
fibrillation were symptomatic for syncope (10) and congestive heart failure (9). At time of repeat Fontan nine patients were in sinus rhythm, one had junctional rhythm (10), one had chronic atrial tachycardia (8).

- Anatomical obstruction—one patient had obstruction at the atriopulmonary junction caused by calcification of the homograft, with a peak gradient of 13 mm Hg.
- Severe ventricular dysfunction—the majority of patients had ventricular dysfunction at rest, shown by echocardiography or scintigraphy; five had ventricular dysfunction on effort.

Thus, the primary indications for repeat Fontan were debilitating arrhythmias in seven patients, anatomical obstruction in one, ventricular dysfunction in five, and coexistence of more than one indication in three. Preoperative analysis of arrhythmias consisted of Holter monitoring and, when possible, an ergometric test. Electrophysiological study was not performed either preoperatively or intraoperatively, as it was not considered to be useful.

Statistical evaluation was performed with paired Student’s t test.

Operative procedure

All patients had right atriopulmonary anastomosis. The operative technique has been previously described. After preparation of the femoral vessels and median sternotomy, the right atrium, the superior and inferior venae cavae, and the atriopulmonary anastomosis were widely dissected and the right atrium, the superior and inferior venae cavae, and the atriopulmonary branches mobilized. Aortobicaval cardiopulmonary bypass and moderate hypothermia (32°C) were used. During cooling a longitudinal right atriotomy was performed so as to achieve adequate exposure of the atriopulmonary anastomosis. The superior and inferior venae cavae and the atriopulmonary anastomosis were divided and a generous right atrial linear lesions were performed with radiofrequency ablation in the atrioventricular valve anulus, the coronary sinus and the superior vena cava, and along the atrial septum, following the procedure illustrated by Mavroudis and colleagues. Similar linear lesions were performed with radiofrequency multipolar catheter (60° for 90 seconds each).

The right pulmonary artery was incised and a superior cavopulmonary connection was fashioned, placing the anastomosis towards the bifurcation. The inferior aspect of the right pulmonary artery was incised and a superior cavopulmonary anastomosis was fashioned, placing the anastomosis towards the bifurcation. The inferior vena cava, the inferior vena cava and the atriopulmonary anastomosis and the superior vena cava, and along the atrial septum, following the procedure illustrated by Mavroudis and colleagues. Similar linear lesions were performed with radiofrequency multipolar catheter (60° for 90 seconds each).

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Intraoperative treatment of arrhythmias included cryoablation (Frigitronics, Shelton, Connecticut, USA) in six patients and radiofrequency ablation (Thermaline Boston Scientific, San Jose, California, USA) in five. The same surgical approach was used regardless of the type of arrhythmias. When cryoablation was used, multiple lesions (–60° for 90 seconds each) were placed in linear fashion between the coronary sinus and the atriocentral valve anulus, the coronary sinus and the inferior vena cava, the inferior vena cava and the atriocentral valve anulus, the atriopulmonary anastomosis and the superior vena cava, and along the atrial septum, following the procedure illustrated by Mavroudis and colleagues. Similar linear lesions were performed with radiofrequency multipolar catheter (60° for 90 seconds each). The right pulmonary artery was incised and a superior cavopulmonary connection was fashioned, placing the anastomosis towards the bifurcation. The inferior aspect of the right pulmonary artery was incised and an inferior cavopulmonary connection was created by interposition of a PTFE (polytetrafluoroethylene) conduit. The diameter of the conduit was 22 mm in 10 patients and 24 mm in the last patient. In the first two patients an atrial pacemaker lead was placed in...
the left atrium according to the technique of Hoyer and colleagues; in the remaining patients an eluting steroid epicardial lead was used. In nine patients the lead was connected to a pacemaker implanted in a subfascial abdominal pouch.

The only patient receiving associated procedures (patient 7) had patch exclusion of a right ventricular aneurysm.

Follow up
All patients were assessed with echocardiography, Holter monitoring, transoesophageal stimulation, ergometric test with oxygen consumption, and myocardial scintigraphy. Echocardiography (Acouos Sequoia C 256 with a 2.5–3.5 probe) was used to rule out the presence of turbulent flow or thrombi, to evaluate the presence of atrioventricular valve(s) incompetence, and to assess the ejection fraction at rest in the four chamber view.

Transoesophageal atrial stimulation was performed, under mild sedation, according to the protocol described previously. The following parameters were evaluated: firstly, corrected atrial recovery time, used as an expression of sinus node function; secondly, the heart rate at which Luciani Wenckebach 2° atrioventricular block appeared during asynchronous atrial stimulation; and lastly, inducibility of re-entry atrial tachycardia.

Ergometric test was performed on a stationary cycle, following the protocol of James and colleagues. The following parameters were evaluated: maximal work load, maximal heart rate and blood pressure, minute ventilation, maximum oxygen consumption, and ventilatory anaerobic threshold. Patients were classified in accordance with the Weber functional class.

Multiple gated equilibrium radionuclide angiography was performed with the subject in the semioorthostatic position on the stress table. Every three minutes the work load was increased by 25 W until maximum exercise was reached or symptoms appeared. If patients became exhausted before completing the test, the work load was reduced by 25 W to allow acquisition for at least two minutes. Heart rate, blood pressure, and ECG were monitored throughout the exercise. Ejection fraction was calculated after background subtraction by interactive semiautomated software. The normal value for ejection fraction at rest was > 50%. An increase of > 5 of resting ejection fraction was considered to be a normal response to exercise, and a decrease of < 5% was considered to indicate exercise dysfunction. A single operator processed all the studies.

RESULTS
Operative results
Table 3 shows the perioperative data. Mean (SD) duration of extracorporeal circulation and aortic cross clamp was 6 (1.7) hours and 35.4 (23.5) minutes, respectively. Mean (SD) duration of mechanical ventilation was 3.5 (3.5) days. Mean (SD) intensive care unit and hospital stay was 5.9 (4.8) and 22 (6.7) days, respectively.

Early complications were one transient low output syndrome and one limb ischaemia, requiring left tight fasciotomy. One hospital death occurred as a result of pneumonia 11 days after intervention.

At discharge all patients were in NYHA class I or II. ECGs showed sinus rhythm in four patients and paced rhythm in six. One late complication was observed in patient 8, in whom, because of poor control of anticoagulation, a thrombus at the junction of the inferior vena cava and the hepatic veins, successfully treated with heparin infusion, was observed two months after reoperation.

Table 3 Operative data

<table>
<thead>
<tr>
<th>Patient</th>
<th>Conduit size (mm)</th>
<th>Type of ablation</th>
<th>ECC (hours)</th>
<th>Aortic cross clamp time (min)</th>
<th>Mechanical ventilation (days)</th>
<th>ICU stay (days)</th>
<th>Hospital stay (days)</th>
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</tr>
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<td>22</td>
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<td>5</td>
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<tr>
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<td>11</td>
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<td>Pneumonia</td>
<td>NA</td>
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<tr>
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<td>34</td>
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<td>2</td>
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</tr>
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</table>

ECC, extracorporeal circulation; ICU, intensive care unit; PM, pacemaker; RFA, radiofrequency ablation; SR, sinus rhythm.

Table 4 Follow up data

<table>
<thead>
<tr>
<th>Patient</th>
<th>NHYA class</th>
<th>Basal rhythm</th>
<th>Relapse of arrhythmia</th>
<th>Transoesophageal stimulation</th>
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<td></td>
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<td>I</td>
<td>SR</td>
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<tr>
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<td>II</td>
<td>SR/PM</td>
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<tr>
<td>10</td>
<td>I</td>
<td>SR/PM</td>
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<td>150</td>
</tr>
<tr>
<td>11</td>
<td>I</td>
<td>PM</td>
<td>No</td>
<td>150</td>
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</table>

AT, atrial tachycardia; cSNRT, corrected sinus node recovery time; RAP, refractory atrial period; SVT, supraventricular tachycardia; W point, Wenckebach point. *Exceeds normal limit.
Follow up

Mean (SD) duration of follow up was 16.8 (5.6) months. Relapse of atrial tachycardia occurred in three patients 11, 3, and 6 months after reintervention and was controlled with medical treatment. In patient 7, because of coexistent atrial tachycardia and bradycardia, a pacemaker generator was implanted 11 months after total cavopulmonary connection and medical treatment was started, with complete control of the arrhythmia. In all the remaining patients Holter registration did not show any arrhythmia. All patients were treated with antiaggregating or anticoagulating agents. Three patients (7, 8, and 11) with suboptimal ventricular function were pre-treated with antiarrhythmic drugs or with radiofrequency ablation. More recently relapse of atrial tachycardia occurred in three patients 11, 3, and 6 months after reintervention and was controlled with antiarrhythmic drugs or with radiofrequency ablation. More recently an atrial tachycardia pacemaker be implanted.

Table 5 shows the results of ergometric tests. At rest seven patients were in sinus rhythm and two were paced. On effort four were paced. The mean (SD) maximal work load, heart rate of 188.3 (29.9) beats/min. Supraventricular tachycardia was not induced in any patients. Echocardiography showed a mean (SD) ejection fraction of 48.9 (11.2%), being < 50% in two patients and > 40% in one. A mild incompetence of the atrioventricular valve was present in four patients (2, 4, 6, and 9). All patients had laminar flow in the systemic venous conduit, without a smoke-like effect. Ventricular dysfunction at rest was reversed in three and on effort was found in five and two patients, respectively. Postoperatively ventricular dysfunction at rest and on effort was found in five and two patients, respectively. Thus, ventricular dysfunction at rest was reversed in three patients and dysfunction on effort in four.

**DISCUSSION**

Total cavopulmonary anastomosis was introduced with the aim of reducing the late complications usually observed in patients with atrio pulmonary Fontan, such as giant right atrium, refractory or debilitating arrhythmias, pulmonary venous obstruction, ventricular dysfunction, and protein losing enteropathy. Studies of relatively recent relapse of atrial tachycardia and patient 9 declined to collaborate). Atrial recovery time was detectable in only six patients who were not paced and was prolonged in two. Atrioventricular conduction was normal in all patients; Luciani Wenckebach 2nd atrioventricular block appeared at a mean (SD) heart rate of 188.3 (29.9) beats/min. Supraventricular tachycardia was not induced in any patients.

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**DISCUSSION**

Total cavopulmonary anastomosis was introduced with the aim of reducing the late complications usually observed in patients with atrio pulmonary Fontan, such as giant right atrium, refractory or debilitating arrhythmias, pulmonary venous obstruction, ventricular dysfunction, and protein losing enteropathy. In some patients these complications can be controlled with anticoagulating, anticongestive, and antiarrhythmic drugs or with radiofrequency ablation. More often myocardial dysfunction caused by coronary sinus hypertension, pulmonary venous compression caused by giant right atrium, atrial arrhythmias caused by previous suture lines, and right atrial enlargement are difficult to manage medically. In the past, these patients usually underwent cardiac transplantation.

The extracardiac conduit approach excludes the right atrium from the systemic venous circuit, avoids additional atrial suture lines, and allows extensive atrioplasty, with removal of arrhythmogenic atrial tissue. At the time of total cavopulmonary connection, some authors have suggested that arrhythmia circuit cryoablation be performed and an anti-tachycardia pacemaker be implanted. Studies of relatively
small numbers of patients showed that conversion of atrio-pulmonary to total cavopulmonary connection provides better control of arrhythmias, allowing antiarrhythmic treatment to be stopped in about 50% of patients. Relapse of arrhythmias, easily controlled by reintroduction of medical treatment, was observed in 30% of patients. No further relapse of arrhythmia was observed after the reintroduction of medical treatment. In the setting of Fontan reconstruction relapse of atrial arrhythmia has multiple explanations. Firstly, not all right atrial tissue can be removed. Secondly, new suture lines are necessarily created. Thirdly, gaps in a linear lesion are created by cryoablation and, less frequently, by radiofrequency ablation. In terms of relapse of arrhythmias, radiofrequency ablation is theoretically superior to cryoablation, as with this technique it is more difficult to leave gaps in a linear lesion. In addition, radiofrequency ablation is less time consuming than cryoablation. However, in our series, we applied cryoablation in six patients and radiofrequency ablation in five, and observed relapse of arrhythmias in both groups. The most important complication of intraoperative ablation was sinus node dysfunction. Sinus node dysfunction was probably caused in part by the extensive atrial wall resection and in part by ablation lines placed near the sinus node. Sinus node dysfunction may be present immediately after discontinuation of bypass but may appear later, as in two of our patients who developed sick sinus syndrome after one and 24 months, respectively. According to our results, an atrial wire should be inserted in every patient undergoing Fontan conversion even if sinus rhythm is present at the end of the operation. Transoesophageal atrial stimulation confirmed the good early results of intraoperative ablation, as no patient had inducible atrial arrhythmia. On the contrary, sinus node dysfunction was present in most patients, as two had a permanent AAIIR pacemaker rhythm and three had a low heart rate increment during the stress test. In our patients atrial wall reduction and ablative lesions close to the sinus node probably worsened the sinus node dysfunction, often present in Fontan patients. These observations reinforce the need for implanting an atrial epicardial wire during reoperation, with the aim of preserving atrial contraction and optimising ventricular function.

Early follow up of repeat Fontan surgery showed that conversion of atrio-pulmonary to total cavopulmonary connection, in accordance with other authors’ reports, has a relatively low mortality and morbidity. Although, before reintervention, none of our patients was in a functional class higher than III, at a mean follow up of 17 months all but one were in class I. Previous data showed that patients in class IV can also improve, after reconnection, to class I–II. We confirmed that intra-atrial tachycardia frequently occurs in Fontan patients. Its anatomical substrates are scars, patches, fibrotic areas related to atrial dilatation, and natural barriers such as the atrioventricular valves, and the orifices of the venae cavae and of the pulmonary veins. In this setting, we performed a compartmentalisation of the right atrium, as suggested by Mavroudis and colleagues. As this arrhythmia may be caused by a macro re-entry or to a micro re-entry circuit, an extensive right atrial wall resection seemed ideal to reduce the occurrence of micro re-entry circuits. Ablative procedures have been performed without a preoperative electrophysiological evaluation. The increasing experience with atrial arrhythmias, obtained with percutaneous ablative procedures, improved the understanding of the correlations between the electrocardiographic patterns and the anatomical substrate of arrhythmias, reducing the number of diagnostic electrophysiological studies. Concerns may be raised for intra-atrial re-entry tachycardia, in which this correlation is usually lacking, because of the coexistence, in the same patient, of multiple re-entrant circuits, which may develop. However, when a surgical ablative procedure is planned, every effort should be made to ablate all possible re-entrant circuits and, for this purpose, a preoperative electrophysiological evaluation was not considered to be essential.

Our data confirm that reconnection of atrio-pulmonary to total cavopulmonary connection provides better control of arrhythmias, allowing antiarrhythmic treatment to be stopped in about 50% of patients. Relapse of arrhythmias, easily controlled by reintroduction of medical treatment, was observed in 30% of patients. No further relapse of arrhythmia was observed after the reintroduction of medical treatment. In the setting of Fontan reconnection relapse of atrial arrhythmia has multiple explanations. Firstly, not all right atrial tissue can be removed. Secondly, new suture lines are necessarily created. Thirdly, gaps in a linear lesion are created by cryoablation and, less frequently, by radiofrequency ablation. In terms of relapse of arrhythmias, radiofrequency ablation is theoretically superior to cryoablation, as with this technique it is more difficult to leave gaps in a linear lesion. In addition, radiofrequency ablation is less time consuming than cryoablation. However, in our series, we applied cryoablation in six patients and radiofrequency ablation in five, and observed relapse of arrhythmias in both groups. The most important complication of intraoperative ablation was sinus node dysfunction. Sinus node dysfunction was probably caused in part by the extensive atrial wall resection and in part by ablation lines placed near the sinus node. Sinus node dysfunction may be present immediately after discontinuation of bypass but may appear later, as in two of our patients who developed sick sinus syndrome after one and 24 months, respectively. According to our results, an atrial wire should be inserted in every patient undergoing Fontan conversion even if sinus rhythm is present at the end of the operation. Transoesophageal atrial stimulation confirmed the good early results of intraoperative ablation, as no patient had inducible atrial arrhythmia. On the contrary, sinus node dysfunction was present in most patients, as two had a permanent AAIIR pacemaker rhythm and three had a low heart rate increment during the stress test. In our patients atrial wall reduction and ablative lesions close to the sinus node probably worsened the sinus node dysfunction, often present in Fontan patients. These observations reinforce the need for implanting an atrial epicardial wire during reoperation, with the aim of preserving atrial contraction and optimising ventricular function.

Ergometric test data, available in only two patients before conversion, did not allow the evaluation of a possible improvement of exercise capacity after reconnection. All our patients were able to perform a significant test, reaching the anaerobic threshold. All but two had a poor exercise tolerance. This is in accordance with previous data showing that Fontan patients have a decreased exercise tolerance, a reduced peak and mean maximal oxygen uptake, and prolonged recovery time for heart rate and maximum oxygen uptake. This response to exercise has been attributed to a reduction in stroke volume at both low and high intensity exercise, to an abnormal linkage of ventilation to tissue carbon dioxide production, and to an increased dependence on anaerobic metabolism in skeletal muscles. In most patients the increment of heart rate during effort was very low, suggesting a latent sinus dysfunction.

Previous studies of ventricular function after Fontan operation found an abnormal systolic function and a depressed response to exercise in most patients. Ventricular filling abnormalities have been found to be common after atrio-pulmonary anastomosis, regardless of the type of dominant ventricular morphology, and are thought to be related to either intrinsic myocardial or acquired factors, such as an increased mass to volume ratio and raised systemic vascular resistances. Our data confirm a depressed ventricular function either at rest or on effort in Fontan patients, whereas they show an improvement in myocardial performance after reconnection in most of them. Previous data on scintigraphic evaluation of ventricular function before and after repeat Fontan operation are not available. The observed improvement in myocardial function after reconnection may be attributed to reversion of the hypertension in the coronary sinus and subsequent improvement of coronary perfusion, to reversion of pulmonary vein compressions, to removal of the atrial reservoir, and to restoration of laminar systemic venous flow. In addition, the preservation of the spontaneous or paced sinus rhythm, maintaining the synchronism between atrial and ventricular contraction, may optimise the function of the single ventricle.

In conclusion, conversion from atrio-pulmonary to total cavopulmonary connection can constitute an alternative to cardiac transplantation in patients with atrio-pulmonary connection with uncontrolled arrhythmias, ventricular dysfunction, or obstruction of the conduit. A longer follow up is required to test the incidence of arrhythmia relapse and the durability of the improvement of ventricular function and functional class.

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REFERENCES
A 50 year old woman was admitted with central crushing chest pain. She had a strong family history of ischaemic heart disease (IHD). Her cholesterol was 4.7 mmol/l. There were no other risk factors for IH. At the time of admission she was haemodynamically stable. Her ECG showed 2–6 mm ST segment elevation in the inferior leads. She was thrombolysed with tenecteplase. The ECG showed complete resolution of the ST segment. There was no elevation of cardiac troponin. She developed further chest pain with ST segment elevation in the inferior leads, and coronary angiography showed complete occlusion of the proximal right coronary artery (upper panel, middle). A prophylactic temporary pacemaker was inserted because the patient developed severe sinus bradycardia. This lesion did not respond to intracoronary nitrates. Hence the lesion was predilated and stented (Guidant Penta 4/38) with a good result angiographically (lower panel, middle). Twenty four hours later the patient developed further chest pain and ST segment elevation in the inferior leads. Repeat coronary angiography showed the stent was widely patent but there was a severe diffuse coronary artery spasm distal to the stent (upper panel, right) which responded very well with intracoronary nitrates (lower panel, right).

We believe that the complete occlusion of the right coronary artery initially was probably caused by severe coronary artery spasm which was resistant to intracoronary nitrates. The patient was discharged on maximal dose of calcium channel blockers and nitrates. She still has recurrent chest pain and a recent angiogram showed mild spasm of the proximal coronary artery just proximal to the origin of the stent, which responded to intracoronary nitrates.

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