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Recovery from Fontan circulation failure by application of continuous negative extrathoracic pressure

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Abstract
A 2-year-old girl developed lethal circulatory failure, general edema, and hepatic dysfunction in an acute phase after total cavopulmonary connection, a Fontan-type operation. Application of continuous negative extrathoracic pressure (CNEP) with a cuirass ventilator at −4 cmH_2O under spontaneous respiration dramatically improved hemodynamics, with systolic arterial pressure increasing from 82 mmHg to 90 mmHg, and central venous pressure decreasing from 15 mmHg to 13 mmHg; also, urine output increased, from 1.6 $ml \cdot kg^{-1} \cdot h^{-1}$ to 6.4 $ml \cdot kg^{-1} \cdot h^{-1}$. Improvements in hepatic function and fluid retention (reduction of pleural fluid and ascites) were also observed. The patient was successfully weaned from CNEP after 5 days. CNEP is an easily applicable, noninvasive tool to reduce pulmonary impedance, and is specifically useful to improve hemodynamics in patients after a Fontan-type operation. Our result suggests that CNEP may represent a first-line option to save patients from critical circulatory failure after a Fontan-type operation.

Key words Fontan circulation · Continuous negative extrathoracic pressure · Hepatic dysfunction

Introduction

A Fontan-type operation, including total cavopulmonary shunt, is a procedure to achieve palliative cure of a single-ventricle heart. Failure of Fontan circulation frequently occurs when impedance in the pulmonary circulation becomes high in comparison with passive, nonpumped venous flow. A low-output state and venous congestion-related organ injury represent significant adverse outcomes. We encountered a child who became critically ill due to severe hepatic damage associated with low cardiac output in the early postoperative period after total cavopulmonary shunt. The patient was saved from the failed Fontan circulation with the application of continuous negative extrathoracic pressure (CNEP).

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Case report

A 2-year-old girl underwent total cavopulmonary shunt using an extracardiac cavopulmonary conduit. Preoperative catheterization data were as follows: pulmonary vascular resistance, 1.98 Wood units·m^2; pulmonary artery index, 576 $mm^2 \cdot m^{-2}$; ejection fraction of single right ventricle, 52%; regurgitation of common atrium-ventricle valve, grade I; and normal pulmonary artery. These data fulfilled the criteria for a Fontan-type operation. The operation was completed without any complications and the patient was successfully weaned from positive-pressure ventilation and extubated at 9 h postoperatively. For the next several days following extubation, inotropic support, using dopamine at 3–6 $mg \cdot kg^{-1} \cdot min^{-1}$ and massive fluid loading, was still needed to maintain stable hemodynamics. Furosemide was needed to keep urine output at more than 1 $ml \cdot kg^{-1} \cdot min^{-1}$. Severe ascites and pleural fluid accumulation were observed and were associated with progressive hepatic dysfunction, with serum transaminase levels at more than 100 $IU \cdot l^{-1}$ (Fig. 1).

These findings were consistent with the status of "failed" Fontan circulation. Marked blood flow stagnation in the inferior vena cava was detected on an ultrasonogram and on angiography (Fig. 2). Cardiac ventricular wall motion was well maintained under the support of low-dose dopamine. We therefore considered additional therapeutic strategies to reduce pulmonary vascular resistance. A noninvasive, easy-applicable CNEP method was first applied for the aforementioned purpose while the patient kept breathing spontaneously.

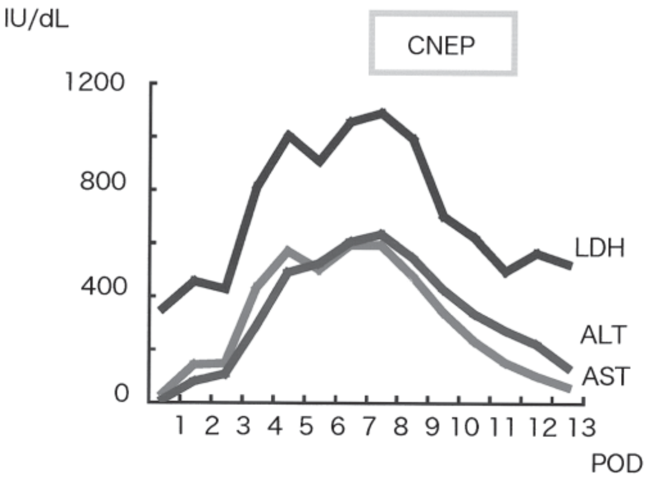

Fig. 1. The patient developed progressive hepatic dysfunction in the postoperative period. The increased levels of hepatic transaminases were significantly decreased after the application of continuous negative extrathoracic pressure (*CNEP*). *LDH*, lactate dehydrogenase; *ALT*, alanine aminotransferase; *AST*, aspartate aminotransferase; *POD*, postoperative day

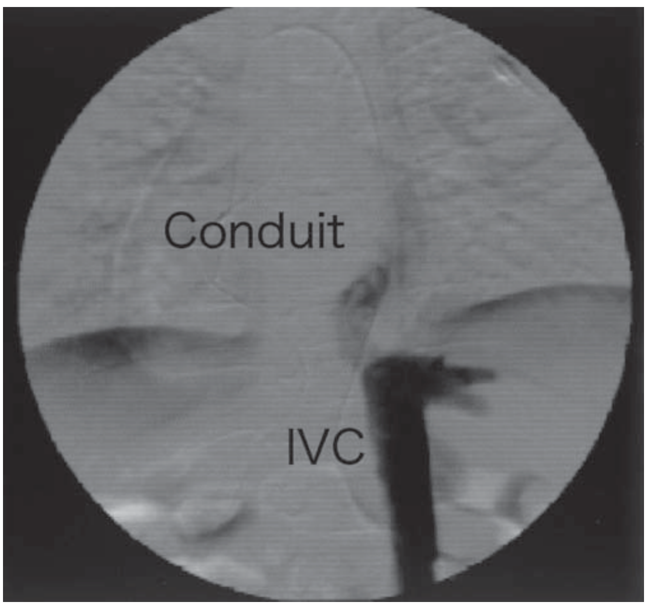

Fig. 2. Postoperative angiography shows severe stagnation of blood flow in the inferior vena cava (*IVC*)

We thought that reducing impedance in the pulmonary circulation with the application of CNEP might relieve the systemic venous stagnation. Written informed consent for this special treatment was obtained from both parents. A biphasic cuirass ventilator (RTX; Medivent, London, UK) was used to attain CNEP. A size 2 cuirass was placed over the chest and upper abdomen. Negative extrathoracic pressure of –3 to –4 cmH_2O was continuously applied. The patient was breathing spontaneously with nasal oxygen supplementation ($2 l\cdot min^{-1}$) throughout the application of CNEP. Prompt changes in hemodynamic parameters were observed after the application of CNEP. Initiation of CNEP decreased central venous pressure from 15 mmHg to 13 mmHg, and increased systolic arterial pressure from 82 mmHg to 90 mmHg. Moreover, a dramatic increase in urine output, from $1.6 ml\cdot kg^{-1}\cdot h^{-1}$ to $6.4 ml\cdot kg^{-1}\cdot h^{-1}$, was observed. The superior vena cava (SVC) flow-velocity waveform, as measured by Doppler echocardiography, increased by 20% after applying CNEP. Hemodynamic improvements were followed by dramatic decreases in serum hepatic transaminase levels, with aspartate aminotransferase (AST) falling from $630 IU\cdot l^{-1}$ to $79 IU\cdot l^{-1}$ and alanine aminotransferase (ALT) falling from $589 IU\cdot l^{-1}$ to $44 IU\cdot l^{-1}$ (Fig. 1). Simultaneously, the amounts of pleural fluid and ascites decreased. The patient was successfully weaned from CNEP after 5 days. No signs of recurrent circulatory failure have since been observed. CNEP appeared to increase pulmonary and systemic blood flow by decreasing intrathoracic pressure and impedance. The present case suggests that CNEP may represent a useful, first-line option to save patients with critical circulatory failure after a Fontan-type operation.

Discussion

A Fontan-type operation is performed for patients with a single-ventricle heart. In these patients, anastomosis between the vena cava and pulmonary artery directs systemic venous return to the pulmonary vascular beds without the intervention of a pumping ventricle. Maintaining low pulmonary vascular impedance is thus crucial to avoid circulatory failure, represented by low cardiac output and organ congestion [1,2].

As pulmonary blood flow is easily stagnated by high pulmonary impedance, strategies to avoid positive intrathoracic pressure are specifically beneficial for Fontan patients [3]. Decreased cardiac output becomes significant at a positive end-expiratory pressure of more than 6 cmH_2O [4]. Some authors have therefore suggested the importance of early extubation [5]. Recent studies have indicated significant improvements in short-term hemodynamics in Fontan patients with the application of negative-pressure ventilation [6].

Shekerdemian et al. [7,8] reported that pulmonary blood flow was increased by 42% after the application of negative-pressure mandatory ventilation in intubated Fontan patients. Our patient received continuous negative pressure under spontaneous breathing, and showed significant short-term improvements in hemodynamics comparable to those in the above reports [7,8]. Significant increases in urine output following the application of CNEP could contribute to the rapid removal of excess body fluids from the thorax and abdomen. Effective fluid removal, in conjunction with enhanced negative intrathoracic pressure, was specifically beneficial in saving the hemodynamics from the malignant cycle of exacerbated Fontan circulation. Immediate recovery of the circulation saved the patient from lethal venous congestion-related organ injury.

One might consider several alternatives to improve Fontan circulation; reintubation, higher inhaled oxygen, inhaled nitric oxide, or the additional use of inotropics with or without vasodilators. These therapeutic interventions, however, may be invasive and could confer possible side effects, i.e., positive-pressure ventilation or tachycardia might, conversely, exacerbate Fontan circulation. The possible benefit of CNEP consists of its easy applicability and noninvasiveness. We therefore conclude that the application of CNEP under spontaneous breathing can be used as a primary rescue option to avoid takedown surgery in Fontan patients. Prospective clinical studies to evaluate the therapeutic effects of CNEP in children following a Fontan-type operation should be explored in future.

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