Tricuspid atresia 1c accompanying neonatal encephalopathy treated with pulmonary trunk banding and therapeutic hypothermia

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ABSTRACT

The influence of therapeutic hypothermia, known to improve neurodevelopmental outcomes in neonatal encephalopathy, remains unknown in newborns with severe congenital heart diseases. We report a neonate with tricuspid atresia type 1c suffering from moderate neonatal encephalopathy. A burst suppression pattern on amplitude-integrated electroencephalography recovered after inducing moderate therapeutic hypothermia, but exacerbated pulmonary overcirculation still persisted even after the rewarming. Since the medical treatment for pulmonary overcirculation had reached the limit, semi-urgent pulmonary trunk banding was performed on the 4th day of life. Postoperative brain magnetic resonance imaging showed no apparent brain injuries; the patient was discharged uneventfully. We share our perioperative management experience of a patient with tricuspid atresia type Ic who required therapeutic hypothermia for neonatal encephalopathy.

Keywords: Congenital heart disease, neonatal encephalopathy, pulmonary overcirculation, therapeutic hypothermia, tricuspid atresia

INTRODUCTION

Therapeutic hypothermia has become the standard management for newborns with neonatal encephalopathy.[1,2] Since many large clinical trials on therapeutic hypothermia after neonatal encephalopathy have excluded patients with congenital heart diseases (CHD), a little is known about the influence of therapeutic hypothermia in newborns with these anomalies. Although Boos *et al.* have recently focused on the clinical significance of therapeutic hypothermia for neonatal encephalopathy after perinatal asphyxia in infants with ductal-dependent CHD, following a report by Mulkey *et al.*, the management of those undergoing therapeutic hypothermia with pulmonary overcirculation has not been fully investigated.[2,3] We present a case of tricuspid atresia Ic with moderate neonatal encephalopathy treated by pulmonary trunk banding and therapeutic hypothermia.

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CASE REPORT

A 2393 g male neonate prenatally diagnosed with tricuspid atresia 1c was born vaginally at 38 weeks gestation to a 32-year-old gravida 1 para 1 Japanese woman. Fetal heart rate monitoring showed prolonged decelerations during delivery. A triple nuchal cord was realized and reduced afterward, but the neonate was floppy and had a poor respiratory effort at birth. The presence of an acute obstetric event, lethargic, apneic, poor sucking, and hypotonia at 5 h of life were suggestive of moderate neonatal encephalopathy. He was intubated after several apnea attacks, and whole-body therapeutic

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hypothermia at 33.5°C esophageal temperature for 72 h was initiated 5 h after birth [Figure 1].

Following therapeutic hypothermia, inotropic support with dopamine and dobutamine was required due to arterial hypotension combined with bradycardia after 1 h. The use of inodilators, like milrinone, was avoided because its effect on pulmonary and systemic vascular resistance was difficult to anticipate. In addition, oxygen saturation and arterial oxygen tension did not decrease from >90% and 50 mmHg, respectively, even under the arterial hypotension. Hypoxic gas therapy using nitrogen was begun in the first 18 h of life. Since the urine output decreased to 0.4 mL/kg/h and new mitral valve regurgitation emerged, probably due to congestive heart failure caused by pulmonary overcirculation, furosemide was infused. The urine output subsequently rose to ≥2.5 mL/kg/h [Figure 1].

We checked patient's coagulation test every day; however, none of the tests showed abnormal results or indicated any negative effect by therapeutic hypothermia.

Amplitude-integrated electroencephalography showed a burst suppression pattern at 5 h of age, but recovery occurred to a discontinuous normal voltage without any seizure during therapeutic hypothermia. We completed 72 h of therapeutic hypothermia, and rewarming was accomplished in 12 h. As the neonate was rewarmed, arterial oxygen tension decreased to 31 mmHg, despite the unchanging fraction of inspiratory oxygen [Figure 1]. However, the medical treatment for pulmonary overcirculation and associated congestive heart failure was still at a maximum regardless of the rewarming, and the hypotension prevented us from using further vasodilators. Hence, we were forced to schedule a pulmonary trunk banding on the 4th day of life immediately following rewarming. Semi-urgent

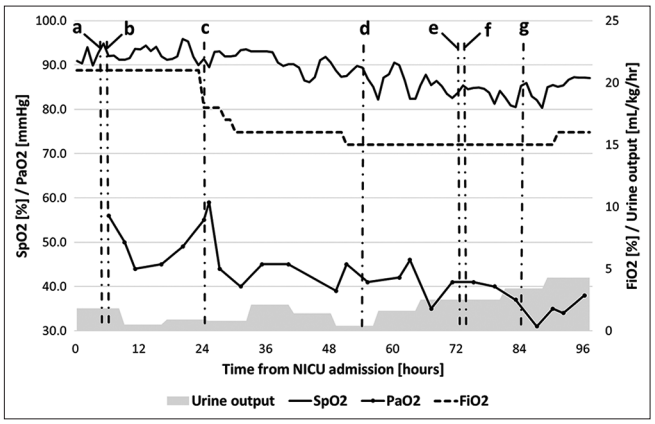

Figure 1: Preoperative clinical course. The vertical lines indicate the following events: (a) intubation; (b) initiation of therapeutic hypothermia and inotropic support; (c) initiation of hypoxic gas therapy using nitrogen; (d) initiation of continuous infusion of furosemide; (e) initiation of rewarming; (f) dose increase of intravenous furosemide; and (g) end of rewarming

pulmonary trunk banding was performed on the 4th day of life, and the patient was extubated on the postoperative day 3. He underwent brain magnetic resonance imaging (MRI) on the 10th day of life, which did not show any apparent brain injury. He was discharged on day 30 postoperation without any complication.

DISCUSSION

This is the first report describing a neonate with tricuspid atresia 1c and moderate neonatal encephalopathy who underwent therapeutic hypothermia. We focused on the clinical course before and after inducing therapeutic hypothermia and the intensive treatment for the congestive heart failure caused by the progressing pulmonary overcirculation. The patient eventually underwent pulmonary trunk banding following the completion of therapeutic hypothermia and was discharged without any apparent brain injury on MRI.

Although a few studies on the combination of tricuspid atresia and neonatal encephalopathy have been published, the clinical significance of therapeutic hypothermia for hypoxic-ischemic encephalopathy after perinatal asphyxia in infants with ductal-dependent CHD has been discussed.[2] One report stated that 4/5 surviving patients had normal/subnormal brain MRI after therapeutic hypothermia, and only one patient presented with nonspecific signal alterations.[2] MRI was normal in our patient, and no acute neurological complications occurred, which indicates a good neurological outcome after therapeutic hypothermia.

The most common adverse effects during therapeutic hypothermia include arterial hypotension and a need for inotropic support or mechanical ventilation.[2] Our patient experienced all of these events, likely due to the atypically progressing pulmonary overcirculation in addition to the therapeutic hypothermia. We could successfully resolve complicated and rapidly changing conditions with extensive treatments.

Pulmonary trunk banding for tricuspid atresia with excessive pulmonary blood flow is recommended after distal pulmonary resistance sufficiently decreases, usually performed in the 2nd, 3rd, or 4th week of life.[4] This may be attributable to the fact that physiological neonatal pulmonary hypertension initially causes the shunt at the level of the ventricular septal defect to be less prominent. It is unclear why the patient in this case needed early banding, atypically, in the 1st week of life; it may possibly be the result of the concomitant therapeutic hypothermia. Surface-induced hypothermia exacerbates pulmonary overcirculation in the patient with a left-to-right shunt, by increasing the systemic vascular resistance almost without changing the pulmonary vascular resistance,[5] which potentially gives a clue of

the atypically progressing pulmonary overcirculation in this case. Although there remained a possibility of postponing the pulmonary trunk banding by vasodilator usage after the rewarming, the situation was difficult due to the additional complications regarding the hypotension and congestive heart failure with almost a maximum treatment.

We completed 72 h of therapeutic hypothermia, but the timing of rewarming was controversial in our case due to the unstable circulatory status. Considering that therapeutic hypothermia for <72 h has also been shown to improve neurologic outcomes,[2] we recommend considering the discontinuation of therapeutic hypothermia when circulatory instability occurs.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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