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Steroid replacement therapy for severe heart failure after Norwood procedure

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Abstract
A 15-day-old neonate demonstrated severe heart failure and capillary leak syndrome after undergoing a Norwood procedure for hypoplastic left heart syndrome. Because she developed severe subcutaneous edema and baseline blood cortisol was low, we suspected relative adrenal insufficiency. After 18 days of dexamethasone administration, her hemodynamics and respiratory function improved, and she was successfully extubated and discharged from hospital. When hemodynamics are unstable in neonates after major cardiac surgery, relative adrenal insufficiency and steroid replacement should be considered.

Key words Adrenal insufficiency · Corticosteroids · Heart failure · Norwood procedure

Introduction

Steroid replacement therapy has been reported to improve hemodynamics and survival in patients with septic shock [1,2] and in premature infants [3,4], possibly because of their insufficient adrenal function. However, reports are limited regarding steroid replacement therapy in post-cardiac surgery neonates [5]. We hereby report one patient in whom long-term steroid replacement therapy improved severe heart failure and capillary leak syndrome, that possibly resulted from relative adrenal insufficiency.

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Case report

A 15-day-old neonate with body weight of 1780 g was electively scheduled for a modified Norwood procedure, because of hypoplastic left heart syndrome (mitral stenosis and aortic stenosis). The diagnosis had been established immediately after normal vaginal delivery at 38 weeks of pregnancy at a birth weight of 2189 g. The Norwood procedure consisted of reconstruction of the aortic arch, conduit implantation from the right ventricle to the pulmonary artery, and atrial septectomy. The anesthesia time was 625 min, operation 460 min, cardiopulmonary bypass 176 min, and aortic crossclamp 20 min. Intraoperative bleeding was 272 mL.

She was sent to the intensive care unit (ICU) with her chest opened, because of hemodynamic instability, oliguria, high blood lactate (7.9 $mmol \cdot l^{-1}$), and low mixed venous oxygen saturation (39%). On postoperative day (POD) 13, we performed a delayed sternal closure with dopamine (4.9 $\mu g \cdot kg^{-1} \cdot min^{-1}$), epinephrine (0.045 $\mu g \cdot kg^{-1} \cdot min^{-1}$), milrinone (0.56 $\mu g \cdot kg^{-1} \cdot min^{-1}$), and phentolamine (2.3 $\mu g \cdot kg^{-1} \cdot min^{-1}$) when her body weight was 2493 g. However, her circulatory condition deteriorated, resulting in oliguria, high lactate (11.5 $mmol \cdot l^{-1}$), and low mixed venous oxygen saturation (28%). The respiratory system compliance decreased from 1.5 $ml \cdot cmH_2O^{-1}$ to 1.0 $ml \cdot cmH_2O^{-1}$ because of severe subcutaneous edema (Fig. 1) and her body weight increased to 3286 g. On POD 38, in the morning, her blood cortisol level was low (11.5 $\mu g \cdot dl^{-1}$). Taking excessive capillary leakage and low cortisol level into consideration, we suspected relative adrenal insufficiency and started steroid therapy (20 $mg \cdot day^{-1}$ methylprednisolone) for 3 days. After the steroid administration, blood pressure and urine output increased (Fig. 2). On the third day of steroid therapy, her body weight had decreased to 2900 g and the dosage of epinephrine was reduced from 0.06 to 0.02 $\mu g \cdot kg^{-1} \cdot min^{-1}$.

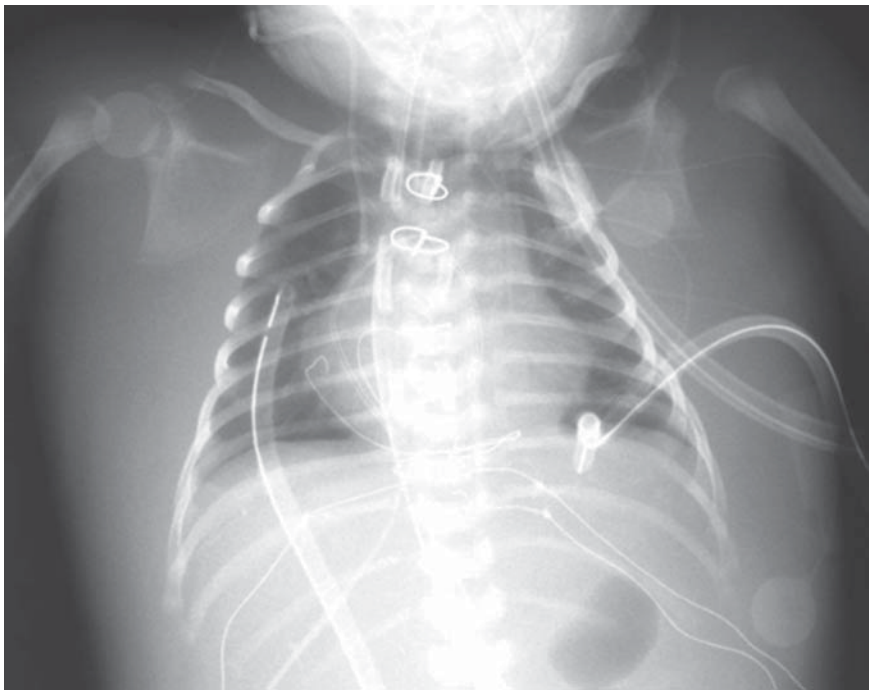

Fig. 1. A chest X-ray film on postoperative day 35. Severe subcutaneous edema was seen

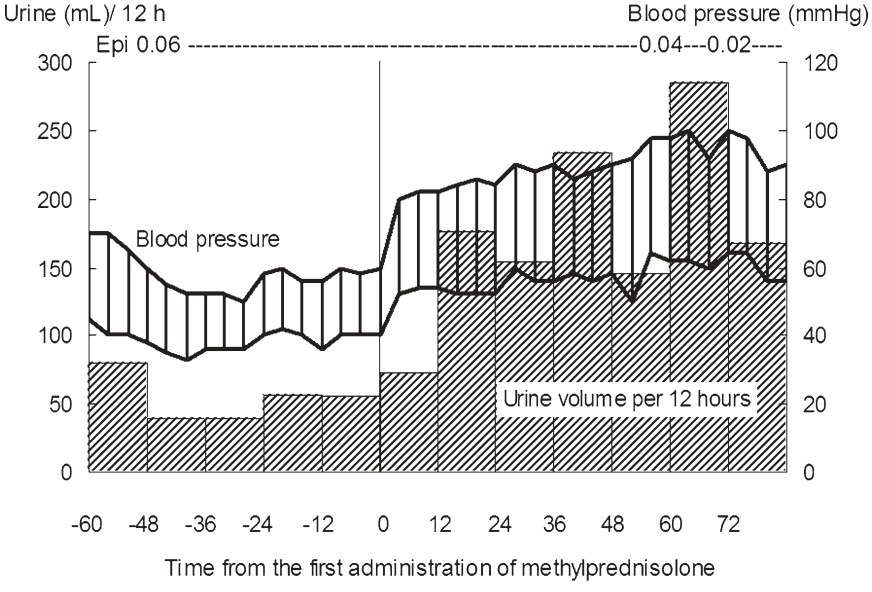

Fig. 2. Urine amount (*left Y axis*) and blood pressure (*right Y axis*) before and after the first administration of methylprednisolone (6 mg) treatment on postoperative day 38. Methylprednisolone (10 mg) was administered every 12 h. After the start of steroid treatment (*time 0*), blood pressure and urine output increased, and epinephrine (*Epi*) was decreased

However, weaning from mechanical ventilation was difficult after the first steroid therapy. Therefore, we started long-term steroid replacement therapy, on POD 45. We decreased the dosage of dexamethasone from 0.5 $mg \cdot kg^{-1} \cdot day^{-1}$ to 0.3, 0.2, 0.1, 0.05, 0.025, 0.0125, 0.006, and then 0.003. Each dose was repeated for 2 days. At the end of the protocol, hemodynamics had become stable with low-dose dopamine (3 $\mu g \cdot kg^{-1} \cdot min^{-1}$) and lactate had decreased to 1.1 $mmol \cdot l^{-1}$. Furthermore, her body weight had decreased to less than 2000 g, subcutaneous edema had disappeared, and respiratory system compliance had increased to 2.5 $ml \cdot cmH_2O^{-1}$. Finally, she was extubated on POD 55. Through this protocol, there were no adverse effects relating to steroid, such as gastrointestinal bleeding, infection, or glucose intolerance. The baseline cortisol level was 16.2 $\mu g \cdot dl^{-1}$ on POD 64, and it increased to 80.3 $\mu g \cdot dl^{-1}$ on POD 70. She was discharged from the ICU on POD 77, and discharged from the hospital on POD 133.

Discussion

We experienced a high-risk neonate who was successfully treated with long-term administration of dexamethasone after a Norwood procedure. When she developed severe heart failure with severe subcutaneous edema, her cortisol level was suppressed. To our knowledge, this is the first report of successful long-term steroid replacement therapy in a post-cardiac surgery neonate whose cortisol level was serially monitored.

It is well known that patients with septic shock often present with relative adrenal insufficiency [2], and steroid replacement at low dose improves the prognosis of these patients [1]. This strategy may be extended to other scenarios. Both Seri et al. [3] and Ng et al. [4] reported, in preterm hypotensive infants with adrenal insufficiency, that corticosteroid increased mean arterial blood pressure. In our patient, the excessive capillary leak, hemodynamic instability, and rapid improvement of the symptoms after steroid administration were consistent with adrenal insufficiency. Although normal values have not been determined for infants, our patient's baseline cortisol ($11.5\,\mu g \cdot dl^{-1}$) was lower than the reported minimum requirement ($15\,\mu g \cdot dl^{-1}$) in the stressed condition for adults [2], suggesting relative adrenal insufficiency. We speculate that transfusion, cardiopulmonary bypass, and mechanical ventilation may induce systemic inflammation and inhibit adrenal cortisol synthesis [2,6]. In addition, the intraoperative use of corticosteroid (methylprednisolone 50 mg) may have inhibited adrenal function [7].

In post-cardiac surgery infants, corticosteroid often improves the hemodynamics and respiratory course [8,9]. However, caution must be exercised against the routine use of corticosteroid, because corticosteroid may cause intraventricular hemorrhage, neuromotor dysfunction, and growth retardation [10,11]. In addition, reports are limited regarding the relationship between adrenal function and the effects of steroid replacement therapy in post-cardiac surgery neonates [5]. We believe this case adds some information about this relationship.

There are several unsolved questions for the use of corticosteroids in post-cardiac surgery neonates. First, we do not know what kind of corticosteroid is the most beneficial in these patients. Although we applied a high dose of methylprednisolone followed by a low dose of dexamethasone in our patient, the short biological half-life of hydrocortisone could be as beneficial as it is in adult patients with septic shock [1]. Second, standard methods and criteria for the diagnosis of adrenal insufficiency in pediatric patients are not established. We used the baseline cortisol level as a guide for adrenal insufficiency, but we should have done corticotropin stimulation tests.

When hemodynamics are unstable in neonates after major cardiac surgery, relative adrenal insufficiency and steroid replacement therapy should be considered.

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