Clinical Analysis of Kawasaki Disease Shock Syndrome

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Kawasaki disease shock syndrome (KDSS) refers to Kawasaki disease (KD) patients who present more than 20% decrease in systolic blood pressure compared to healthy individuals of the same age, or to those patients who show peripheral blood circulation perfusion disorder. KDSS may have varied clinical courses and could be easily ignored and misdiagnosed. Yet, this syndrome could be deleterious, especially when combined with coronary artery injury or multiple organ dysfunctions. Therefore, early diagnosis of KDSS through specific clinical manifestations is critical. This manuscript summarizes the clinical features and treatment methods of 11 children with KDSS who were able to fully recover in our hospital. The results shed lights on understanding the early identification of KDSS and could assist in reducing the complications of KD and improving the quality of life in KDSS patients.

All patients met the criteria for KD according to the standard of American Academy of Pediatrics and Cardiology Society,[1] and they also met the requirements for concurrent shock of KD proposed by Kanegaye *et al*. in 2009.[2] We retrospectively reviewed the medical records of 11 children diagnosed with KDSS in our hospital from February 2012 to February 2017. Among these 11 patients, 7 were male and 4 were female. Ages ranged from 1 month to 9 years. Clinical manifestations of shock occurred in the first 3–10 days during the disease course with a mean time of 6.1 days. Five of the 11 patients experienced shock within 5 days of KD onset. These data suggested that patients already exhibited shock even before KD diagnosis could be made. All the 11 cases showed persistent high fever, lymph node enlargement, and cardiac insufficiency. More than 50% of patients had multiple organ dysfunctions as listed in Table 1.

Table 1: Clinical presentations of children with Kawasaki disease shock syndrome (n = 11)

Clinical presentations	Cases (n)
Physical examination	
Lymphadenectasis	11
Rash	10
Conjunctival hyperemia	10
Joint swelling of the palms or feet	9
Chapped lips	6
Clinical manifestations	
Coronary artery dilatation	9
Shock within 5 days	5
Pneumonitis	7
Auxiliary examinations	
Hypoalbuminemia	10
Hepatic insufficiency	8
Anemia	8
Hyponatremia	6
Electrocardiogram abnormalities	8
Urine routine abnormalities	5
Hypokalemia	3

Intravenous immunoglobulin (IVIG) and oral administration of aspirin were given in all KDSS patients, and re-treatment with IVIG was done in five patients who presented with IVIG resistance. In addition, all patients received fluid volume resuscitation, vasoactive drugs, and nutrition support treatments. Six patients received additional treatments including respiratory support and methylprednisolone.

KDSS is considered a rare disease around the world, yet in the recent years, more interests have focused on the early diagnosis of KDSS. Clinical manifestations of KDSS are

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atypical.[3,4] It can rapidly develop into shock, and often with strong inflammatory responses which could lead to coronary artery disease and multiple organ dysfunctions. Therefore, early identification is particularly important. Through literature review and examination of our 11 patients, we have summarized the following characteristics for KDSS: (1) It is more common in males; (2) symptoms include lymphadenectasis, hypoalbuminemia, hyponatremia, hepatic insufficiency, anemia, and electrocardiogram abnormalities; the incidence of coronary artery dilatation is high, and some children may have severe gastrointestinal symptoms; (3) inflammatory indicators are significantly increased; (4) patients often present with IVIG resistance; (5) 50% of patients need hormone therapy; and (6) shock appears at an early stage. For the treatment of KDSS, IVIG combined with aspirin and vasoactive drugs is the current standard of care. Recent studies[5] have shown that glucocorticoids combined with a large dose of gamma globulin treatment can reduce the incidence of coronary artery disease in patients with severe KD. In this study, 11 patients were given anti-shock therapy, such as fluid resuscitation, vasoactive drugs, and respiratory support therapy. Further, six children underwent hormone therapy due to inflammatory reactions. In our study, a high dosage of methylprednisolone was given to one patient in the initial treatment. Disease progression was controlled, yet heart rate declined afterward. Arrhythmia occurred with the junctional escape rhythm and the onset of cyanosis followed. Vasoactive drugs could not maintain a normal range of blood pressure; therefore, a temporary pacemaker was implanted. For the other patients in our cohort, vital signs and blood and inflammation indicators returned to normal after shock through interventions.

In summary, the cases in our study shared similar clinical manifestations with previously reported cases. However, in addition to standard care, respiratory support and hormone therapy were used in our patients, and temporary pacemakers were implanted when vasoactive drugs could not maintain a normal range of blood pressure. Our study indicated that early diagnosis and comprehensive therapeutic methods based on the disease progression are important in treating KDSS.

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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