Nursing and Clinical characteristics of kawasaki disease shock syndrome

[Key words] Kawasaki disease; Kawasaki disease shock syndrome; Nursing;Review

Abstract

Kawasaki shock syndrome is a severe disease that seriously affects cardiac in children. We need for early identification, timely and correct processing. This review of reference to eighteen international important literature, for the onset of KDSS, diagnosis and treatment, improve the level of diagnosis and treatment technology for KDSS.

Introduction

Kawasaki disease (KD) is the leading cause of acquired heart disease in children, and is steadily increasing in prevalence in East Asia. KD is a type of acute febrile vasculitis, which is typically a self – limited condition mostly involving medium-sized blood vessels resulting in coronary artery abnormalities (CAA) in children in developed countries. It usually occurs in children under 5 years of age. Between 15% and 25% of untreated children can develop coronary artery abnormalities (CAAs), including coronary artery dilation and aneurysms. In 2009 Kanegaye et al. [2] defined KD, in which is there is hemodynamic instablity, as Kawasaki disease shock syndrome(KDSS). It's a rare form of KD, and can lead to severe sequelae and adverse outcomes. The incidence rate is 1.9% to 7.0% in KD^[3-6]. The incidence of KDSS in China is lower than in western countries. During the acute phase, about 5% children with KD have low blood pressure and shock. These children are mainly characterized by low tissue perfusion and unstable blood flow mechanics, accompanied by systolic blood pressure with sustained decrease of 20% or more from baseline or systolic hypotension when compared with children of the same age. Children with KDSS have a higher risk of cardiovascular complications than children with non-shock symptoms, such as Mitral regurgitation, myocardial contraction dysfunction and the higher incidence of coronary artery lesions; treatment against immunoglobulin, and often concurrent multiple organ damage.

We use the keyword search, and retireve the relevant files before the analysis.

1. The diagnosis of KDSS

In 2009, Kanegaye et al. proposed the diagnosis of KDSS: children with KD have sustained blood pressure drop of 20% or more from baseline, systolic hypotension when compared to those of the corresponding age group, or peripheral circulatory perfusion disorders. The systolic blood pressure of children from 1 to 12 months <70

mm Hg or the systolic blood pressure of children from 1 to 12 years $<[70 + 2 \times \text{age} \text{ (years)}]$ mm Hg is hypotension^[7].In addition, KD manifestations of normal systolic blood pressure and decreased diastolic blood pressure have also appeared clinically. Xue Chaochao and others^[8] believe that it is between KD and KDSS, which is a transitional period during the disease course. Foreign literature reports that KDSS accounts for 1.9% to 7% of children with KD^[2]. About 1.78% to 1.90% of children with KD in Taiwan have KDSS, and foreign statistics show that KDSS is more common in females, while domestic studies show that there are more males^[6, 8]. The difference may be related to race, environment, number of sample cases, and atypical early manifestations of the disease, resulting in insufficient clinical understanding and missed diagnosis.

2. Early recognition of KDSS

In the incidence of KDSS, there is no obvious gender difference between males and females, although there are reports in the literature that there are slightly more males than females [9]. Compared with non-shock KD, there is no significant difference in the age of onset of KDSS^[10], so it can occur at any age, including adults. In contrast, KDSS is more likely to appear in infants under 6 months of age and children over 10 years of age. The overall median age of onset is higher than that of KD, and it is more likely to appear in patients with incomplete KD^[11].

3. The possible mechanism of KDSS

The mechanism of circulatory and hemodynamic disorders in KD in the acute phase is still unclear. Most of them are considered to be the result of multiple factors. The possible pathogenesis includes capillary leakage caused by small vasculitis, myocardial contractile dysfunction, and abnormal cytokine regulation.

3.1 Capillary leakage

KD is mainly caused by coronary artery damage, but small and medium blood vessels can be affected throughout the body. Inflammatory factor storm, high expression of vascular endothelial growth factor, and endothelial cell dysfunction are the causes of vascular endothelial damage, resulting in increased vascular permeability. When the permeability of small blood vessels and capillaries continues to increase, plasma extravasation and loss of small molecular proteins occurs. Then there was a drop in plasma colloidal osmotic pressure, a significant decrease in plasma albumin, and a decrease in circulating blood volume^[12]. The clinical manifestations include decreased blood pressure, hypoperfusion of tissues, systemic edema and fluid accumulation in the chest and abdominal cavity.

3.2 Myocardial contraction dysfunction

In children with KD, there is not only the coronary artery damage, but also the myocardial cell damage and myocardial ischemia and myocardial infarction caused by coronary artery disease. Endocardial biopsy confirmed that myocardial fibrosis caused by diffuse myocardial inflammatory injury is an important cause of left ventricular systolic dysfunction in patients with KD^[13].Left ventricular ejection fraction decreased in KD, and the degree of ejection fraction decrease is related to the occurrence of KDSS^[3].It is especially important to note that most children with KD are in subclinical cardiac insufficiency, and so this factors that lead to KDSS.

3.3 Cytokine dysregulation

The waterfall response of cytokines occurring during shock has an important role and significance. Many scholars believe that KDSS and septic shock have similar clinical features and immunological mechanisms. Under the mediation of superantigens^[14],cytokine dysregulation and activation of intracellular signaling pathways occur, and the continuous expansion of microvessels leads to a decrease in peripheral vascular resistance, which leads to circulatory congestion.

4. Nursing

4.1 Nursing Care of KD Fever

Fever is the most common symptom of KD. Children with KDSS are prone to have recurrent fever, and their body temperature fluctuates between 38.5 and 40°C. It is difficult to be normal. It is necessary to closely observe the mental state and vital signs of the children to prevent and treat febrile convulsions. When the general body temperature is lower than 38.5°C, take physical cooling measures as much as possible. For example, use antipyretic ointment, warm water to wipe the bath, drink appropriate amount of warm water etc. When the body temperature exceeds 38.5°C, you can use antipyretic drugs to cool down according to the doctor's including physically measures. Monitor it every half hour to one hour after . If the temperature drops, it often manifests as sweating, and it should be wiped dry and changed clothes or bedding in time to avoid aggravating the illness.

4.2 KDSS emergency care

When the child has KDSS, the nursing staff should move the child to the rescue bed immediately, raise the head 15 degrees, and the lower limbs 20 degrees, to ensure the blood volume is returned. Connect to ECG monitoring, observe and record heart rate, pulse, blood pressure, percutaneous oxygen saturation, capillary filling time,etc. The temperature and color of the extremities can reflect the circulation and perfusion of the extremities to a certain extent. You can touch the extremities to get the shock situation during nursing. Maintain the effective oxygenation index and give oxygen to the mask (5L/min). If the ventilation cannot be improved, mechanical ventilation should be given. Rapid venous expansion can effectively increase the circulating blood volume and prevent the shock from getting worse which is the key to save. For critically ill children, caregivers should establish two venous accesses suitable for expansion within 5 minutes, choose thick and straight proximal veins, and firstly use expensive veins or external jugular veins. Deep venous puncture are necessary to prepare the use of vasoactive drugs.

4.3 How to monitor blood pressure

It is very important to measure and record the blood pressure correctly when the child first admitted to the hospital has shock with lower blood. Choose cuffs of different widths according to the age of the child. The width should be 2/3 of the length of the upper arm. The normal average blood pressure of different ages can be calculated by this formula: systolic blood pressure (mmHg)=80+(age×2).Diastolic blood pressure is 2/3 of systolic blood pressure. Measure blood pressure every hour during the entire nursing process of the child. When using dopamine or dobutamine, measure blood pressure every half an hour if there is a fluctuation in blood pressure,

which will be helpful for the doctor to judge.

4.4 Volume expansion drugs use care

Volume expansion is very important for the children with shock. Normal saline 20ml/kg is the first choice, and completed within 10 minutes. After the nurses for rapid infusion of normal saline, monitor blood pressure and heart rate immediately, and use colloidal fluid if necessary.

4.5 Vasoactive drug use care

At present, the combination of norepinephrine and dobutamine can play a synergistic effect and significantly improve in systemic blood flow. However, both of them are high-risk drugs. Care should be taken when use. Check whether the infusion pump is working properly before using the drug, and paste red marks on the syringe and the infusion tube. Calculate the normal blood pressure of children of the age, set the initial drug speed according to the normal blood pressure and cooperate with the doctor. Follow the principle from low concentration to high concentration, monitor blood pressure, heart rate, blood oxygen saturation and other indicators every 30 minutes. Change to monitoring once every hour after stabilization. When using norepinephrine in peripheral veins, closely observe the changes in local skin color to prevent venous leakage. If venous leakage occurs, stop the infusion immediately, and change the infusion channel in time to keep the blood pressure continuously stable. and give phentola immediately. When stopping the drug, the concentration should be gradually reduced, and it is strictly forbidden to stop the drug suddenly. If the child has lowered blood pressure, lethargy or even coma during the process,inform the doctor immediately.

4.6 Nursing care of gamma globulin shock therapy

Gamma globulin is a blood product. Strict aseptic operation should be performed before infusion. Assessed body temperature. If it reaches 38.5°C or higher, inform the doctor that the infusion should be postponed to prevent infusion reactions until below 38.5°C or lower. Flush the tube with normal saline before and after the infusion to avoid mixing with other drugs. it is about 10ml/min at first. Accelerated not more than 20ml/min if there are no adverse reactions after 15 minutes^[15]. During the medication, you should pay close attention to any adverse reactions. Once fever or rash occurs, suspend the infusion immediately and call the doctor to get the treatment.

4.7 Nursing care for correction of hypoproteinemia and hyponatremia

Children with KDSS have different degrees of hypoproteinemia and hyponatremia; both are significantly related to vascular inflammation and damage^[4]. It should be corrected slowly during the anti-shock treatment. Albumin infusion can be given in hypoalbuminemia and complete within 4 hours, because it is a blood product. Hyponatremia can cause nausea, vomiting, coma, convulsions, edema etc. Sodium should be supplemented in time, usually 24 to 48 hours, and monitor the blood sodium and other electrolytes.

4.8 Aspirin use care

Aspirin is an indispensable drug for the treatment of KD, and it is also the first choice. It has anti-inflammatory and anticoagulant effects^[16]. The early combined application with immunoglobulin can effectively control the acute inflammatory

process and reduce coronary artery disease. Take the medicine every day. In order to reduce the irritation of the medicine and the formation of drug-induced ulcers, the children should take the medicine after meals and eat moderately at night. Avoid taking the medicine on an empty stomach. If you have fever when you get aspirin, you must take at least 6 hours between the withdrawal of ibuprofen or acetaminophen. During the period of taking aspirin, pay attention to observe the child where has bleeding in the nasal cavity, skin, mucous membrane or gums. Not to pick the nose with hands. Use a soft-bristled toothbrush when brushing or gargle. If necessary, use a mouth guard to help the child clean the mouth, give the child a light and easy-to-digest diet, and keep the stool smooth.

4.9 Coronary aneurysm care

KDSS with coronary artery aneurysms can easily rupture and cause death. Nursing staff instruct the child stay in bed absolutely to reduce cardiac work. Observe closely whether the heart rate and heart rhythm is murmur during auscultation. Pat back gently for children in shock stage, and when defectaion is difficult, Kaisailu can be used to avoid coronary aneurysm rupture. Take targeted nursing measures for children in different ages, personalities, and psychological characteristics. Such as communicate more with older children and give encouragement to avoid aggravating their burden; for young children, they often cry because of pain, fear, etc. Toys and cartoons can be given to divert their attention and cooperate with treatment.

4.10 Nutritional care

When KDSS, children should stop eating, receive total intravenous nutrition, and supplement 60-80ml/kg of fluid daily. Children need to supplement water-soluble vitamins daily to facilitate skin repair because of fever, rash, skin erosion and other reasons. After the blood pressure is stable and consciousness is restored, the child should be given a liquid or semi-liquid diet with high calorie, high protein, and high vitamin content. Avoid eating raw, cold, hot, hard, sour, and spicy foods, so as not to affect the recovery process of the disease.

4.11 Skin and mucous membrane care

Children with KD have conjunctival hyperemia in both eyes. Don't rub. When there are more ocular secretions, rinse the eyes with normal saline 1-2 times a day and apply erythromycin ointment twice a day. Avoid strong light exposure. When oral mucosa is chapped or eroded, rinse mouth with sodium bicarbonate solution or normal saline every day, and evenly coat lips with vitamin E every day. Cut peeling off with sterile scissors, and instruct the child not to tear it to avoid infection. Change sheets and clothing once a day to avoid skin infections. Norepinephrine can cause strong contraction of small arteries and venules throughout the body. Superficial vasoconstriction is more pronounced, causing the skin temperature to be dry and cold. It can be used with blankets, warm and dry towels and other measures^[17].

4.12 Psychological care

Children are relatively young. Give more comfort and encouragement during the irritable and irritable stage, especially hugging and shaking hands^[18]. Safe physical contact can increase the children's sense of security. You can also prepare toys and books that the children like, and organize the children and their parents to take some

feasible and meaningful activities to enrich the children's life during the hospitalization and reduce the pressure. Parents are worried so we should explain the condition of the child, and give the knowledge of KD, provide psychological support and spiritual comfort, and build confidence in treatment.

4.13 Health education

Explain the condition of the illness to the family in time and provide psychological support. Do well in discharge guidance, including taking medication according to the doctor's instructions, and observing whether there are digestive symptoms. Supple the nutrition with easy-digestible, high-vitamin,high-protein food. Regular review; enough rest and no exercise. Review after discharge from 1 month, 3 months, half a year to avoid coronary artery disease. Individual children need to be followed to adulthood.

5. Conclusion

All in all, KDSS is an extremely serious complication of KD. Some early clinical symptoms of KDSS are not typical and are easily missed or misdiagnosed. Therefore, early recognition, timely and appropriate treatment can quickly correct shock ,reduce coronary artery damage and multiple organ injuries and other serious complications. It is also important to pay attention to the nursing knowledge for children, and it will also improve the prognosis of children with KDSS.

References

- [1] Fuyong Jiao, Zhilong Mu, Xian Wang. Update on Study of Kawasaki Disease. Journal of Pediatric and Womens Healthcae. [J]. (2018)1:1013-1018. (Crosscheck)
- [2] Kanegaye JT, Wilder MS, Molkara D, et al. Recognition of a Kawasaki disease shock syndrome. Pediatrics. 2009. 123(5): e783-9.
- [3] Qiu H, Li C, He Y, et al. Association between left ventricular ejection fraction and Kawasaki disease shock syndrome. Cardiol Young. 2019. 29(2): 178-184.
- [4] Chen PS, Chi H, Huang FY, Peng CC, Chen MR, Chiu NC. Clinical manifestations of Kawasaki disease shock syndrome: a case-control study. J Microbiol Immunol Infect. 2015. 48(1): 43-50.
- [5] Taddio A, Rossi ED, Monasta L, et al. Describing Kawasaki shock syndrome: results from a retrospective study and literature review. Clin Rheumatol. 2017. 36(1): 223-228.
- [6] Lin MT, Fu CM, Huang SK, Huang SC, Wu MH. Population-based study of Kawasaki disease shock syndrome in Taiwan. Pediatr Infect Dis J. 2013. 32(12): 1384-6.
- [7] Newburger JW, Takahashi M, Burns JC. Kawasaki Disease. J Am Coll Cardiol. 2016. 67(14): 1738-49.
- [8] Honda T, Honda Y, Yoshizawa H, Gribble GW. AN EFFICIENT SYNTHESIS OF TRICYCLIC COMPOUNDS, (+/-)-(4abeta8abeta10aalpha)-1,2,3,4,4a,6,7,8,8a,9,10,10a-DODECAHYDRO-1,1,4a-TRIMETHYL-2-OXOPHENANTHRENE-8a-CARBOXYLIC ACID, ITS METHYL ESTER, AND (+/-)-(4abeta,8abeta10aalpha)-3,4,4a,6,7,8,8a,9,10,10a-DECAHYDRO-8a-HYDROXYMETHYL-1,1,4a-TRIMETHYLPHENANTHREN-2(1H)-ONE. Org Prep Proced Int. 2005. 37(6): 546-550.
- [9] Gamez-Gonzalez LB, Moribe-Quintero I, Cisneros-Castolo M, et al. Kawasaki disease shock syndrome:

- Unique and severe subtype of Kawasaki disease. Pediatr Int. 2018. 60(9): 781-790.
- [10] Qi Y, Wu YH, Yang YL, Yang W, Yang J, Ma W. Clinical analysis of 16 cases of Kawasaki disease shock syndrome [J]. Chinese Journal of Pediatric Emergency Medicine, 2017, 24(12): 925-928. Chinese Pediatric Emergency Medicine
- [11] Singh S, Sharma A, Jiao F. Kawasaki Disease: Issues in Diagnosis and Treatment--A Developing Country Perspective. Indian J Pediatr. 2016. 83(2): 140-5.
- [12] Chen Y, ZHANG Q. Progress in pathogenesis of capillary leak syndrome. Chinese Pediatric Emergency Medicine, 2019, 26(12): 461-465.
- [13] Harada M, Yokouchi Y, Oharaseki T, et al. Histopathological characteristics of myocarditis in acute-phase Kawasaki disease. Histopathology. 2012. 61(6): 1156-67.
- [14] Nagata S, Yamashiro Y, Ohtsuka Y, et al. Heat shock proteins and superantigenic properties of bacteria from the gastrointestinal tract of patients with Kawasaki disease. Immunology. 2009. 128(4): 511-20.
- [15] Shao Ying, XIE Chunhong, Wei Linlin, et al. Early observation and nursing of kawasaki disease complicated with intestinal injury [J]. Nursing and rehabilitation, 2014,13 (12): 1150. Crosscheck
- [16] Mercier JC, Ouldali N, Melki I, et al. Severe acute respiratory syndrome coronavirus 2-related multisystem inflammatory syndrome in children mimicking Kawasaki disease. Arch Cardiovasc Dis. 2021. 114(5): 426-433.
- [17] Niu Q, Zhao X Y, Study on optimal nursing of 46 children with Kawasaki disease [J]. Shaanxi Med J, 2015, 44(4): 512. Crosscheck
- [18] Pan Hong. Psychological characteristics and nursing strategies of preschool patients [J]. Health Vision: Medical Edition, 2014, 22(9): 703 704. Crosscheck