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# A Rare Case of Congenital Varicella in 14 Days-Old Girl Baby

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***Abstract:*** ***Varicella is a rare disease in infant. Maternal varicella between day 5 antepartum to day 2 postpartum may develop congenital varicella with fatal illness. We reported a 14-days-old girl baby presented with chief complaint widespread pleomorphic rash and fever. Patients complained rash on whole body since 3 days before admitted to hospital. The rash initially appeared on chest then spread to face, both of her hands and feet. Redness spots filled with fluid, ruptured, then dry and form crust. There was history of fever since 4 days before admitted to hospital, with the highest temperature 39$^0$C. The patient had laziness to drink. Her mother had fever 2 hours after delivery and varicella rashes 6 hours after delivery. During physical examination, the patient's body temperature was 38.5°C. Laboratories examination revealed reactive anti-varicella zoster IgM. The mother's antibody revealed reactive anti-varicella zoster IgG. The patient was administered intravenous acyclovir for 7 days. We don't have varicella-zoster immune globulin (VariZIG) in our hospital. The lesions recovered on day 9 of treatment and she was discharged. Varicella in the neonates is associated with high morbidity and mortality, but treatment with acyclovir combined with supportive therapy has resulted good outcome.***

**Keywords:** varicella, congenital, acyclovir

## 1.Introduction

Varicella-zoster virus (VZV) is a highly contagious infectious agent that causes primary varicella infection, known as varicella. Its main target is T lymphocytes, epithelial cells and ganglia [1]-[3]. Varicella can affect all age groups with the highest proportion were children less than 10 years of age. The incidence peaks at 5-9 years of age. The incidence of congenital varicella in Indonesia is not certain, but the overall incidence of maternal and congenital varicella has decreased over the past 10-15 years, presumably due to varicella vaccination [1].

There are three forms of varicella-zoster infections involving in fetus and neonate: fetal, congenital (early neonatal) and postnatal [1]-[2]. Congenital (early neonatal) varicella infection occurs when pregnant woman suffers varicella during the last 3 weeks of pregnancy or within the first few days postpartum. Disease begins just before delivery or within the first 10-12 days of life [2], [4].

The clinical manifestation include widespread, intensely pruritic vesicular rashes. Affected individuals typically have 250-500 lesions with varying stages of evolution, often associated with fever or other systemic symptoms. Although the most common complication of varicella is bacterial superinfection from skin lesions, varicella pneumonia is the most common cause of mortality. Other complications of varicella infection include acute cerebellar ataxia, encephalitis, thrombocytopenia, and Reye Syndrome [2].

Varicella-zoster immune globulin (VariZig) should be adminitered as soon as possible, ideally within 72-96 hours after exposure, although administration may be useful up to 10 days postexposure. Intravenous immune globulin (IVIG) may also be considered if VariZig is unavailable. Antiviral therapy with acyclovir may also be indicated [1], [3], [5].

## 2.Case Report

A 14 days-old girl baby with chief complaint widespread pleomorphic rash on the whole body since 3 days before admitted to the hospital. The rash initially appears on chest then spread to face, back and both of hands and feet. Redness spots filled with fluid and rupture. There was history of fever since 4 days before admitted to the hospital, with the highest temperature 39$^0$C. The baby developed laziness to drink since 1 day before admitted to the hospital.

Patient was born spontaneously from mother with varicella. Mother had fever 2 hours after delivery and also had redness spots on stomach since 6 hours after delivery and the next day spread to the whole body. Mother received acyclovir therapy so that the mother prohibited from giving breast milk during taking medication.

On the first day of hospitalization**,** patient still suffered fever and lazy to drink, with the body temperature was measured 38.5°C. Other vital signs were within normal limit. From physical examination, there was multiple vesicles with erythema skin, rounded, scattered throughout body, diameter 0.1-0.3 cm, partially ruptured to form erosion covered brownish black crust. Complete blood count revealed white blood cell 12.9x10$^3$/uL (neutrophils 5.5x10$^3$/uL (42.63%)); haemoglobin 15.27 g/dL; haematocrit 46.09%; platelets 150.2x10$^3$/uL. Patient was assessed with congenital varicella and treated in isolation room. Patient was given formula milk and parenteral nutrition. Patient was planned to get VariVIG/IVIG but no VariVIG/IVIG in our hospital. Patient got acyclovir 15 mg/kg/dose every 8 hours (intravenous), planned for 7 days.

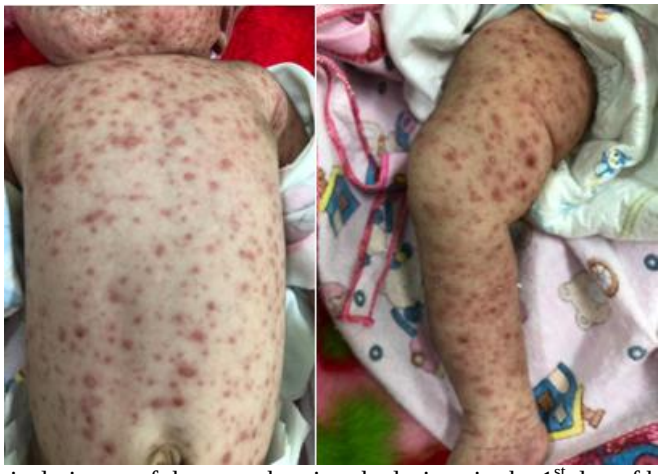

**Figure 1:** Clinical picture of the case showing the lesions in the 1st day of hospitalization

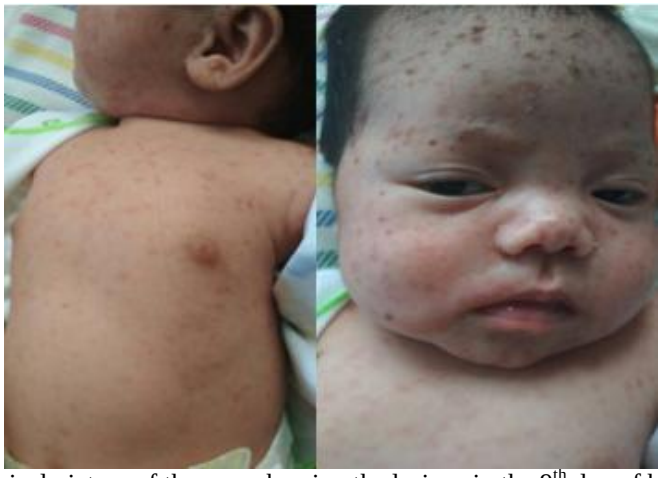

**Figure 2:** Clinical picture of the case showing the lesions in the 9th day of hospitalization

On the 2nd day of hospitalization, patient was consulted to division of dermatology and venereology and recommended salicyl powder and sodium fusidic acid cream at lesions every 12 hours. Laboratory examinations anti-varicella zoster antibodies revealed reactive anti-varicella zoster IgM. We check the antibody of mother too which resulted reactive anti-varicella antibody IgG.

On the 9th day of hospitalization**,** there was no complaint, with vital signs within normal limit. Complete blood count evaluation revealed white blood cell $11.2x10^3$/uL (neutrophils $2.65x10^3$/uL (23.54%)); haemoglobin 12.96 g/dL; haematocrit 38.39%; platelets $415.3x10^3$/uL. Patient was discharged from hospital.

## 3.Discussion

Congenital varicella is the form of disease that occurs when pregnant woman suffers varicella during the last 3 weeks of pregnancy or within the first few days postpartum. Disease begins in neonate just before delivery or within the first 10-12 days of life [1]. Maternal varicella can affect the baby through transplacental viremia, ascending infection during delivery. Maternal varicella in late pregnancy may result in congenital varicella before passive immunity from mother to baby can be conferred and the cell-mediated immune response of the neonate is insufficient to prevent the haematogenous spread of VZV [1], [2]. If maternal varicella occurs <5 days before delivery to 2 days after delivery, up to 50% of neonates will be infected, with mortality 20% [5]. In case of intrauterine acquired disease, the characteristic point in time and the maternal history of varicella during the last weeks of pregnancy have to be considered. The differential diagnosis includes HSV and enterovirus infections [5], [6].

The diagnosis of congenital varicella usually is made clinically based on the characteristics appearance of skin lesions. Clinical presentation of congenital varicella is variable. There may be seen on the skin, lungs and other organs [1], [5]. In the skin, a centripetal rash (beginning on the trunk and spreading to face and scalp, sparing to the extremities) begins as red macules and progresses to vesicles and encrustation. Lesions are more common in diaper area and skin folds. Lung involvement is seen in all fatal cases. It usually appears 2-4 days after the onset of rash but may be seen up to 10 days. Sign include shortness of breath, fever, cyanosis, rales and hemoptysis. Chest radiograph shows diffuse nodular-miliary pattern, especially in perihilar region. In the other hands, gocal necrosis may be seen in the liver, adrenals, intestines, kidneys and thymus. Glomerulonephritis, myocarditis, encephalitis and cerebellar ataxia are sometimes seen [1], [5].

The most sensitive and specific method for the detection of VZV in clinical specimens is the amplification of conserved sequences of viral DNA by polymerase chain reaction (PCR). This technique is clearly the best method for investigation of skin swabs or biopsies, cerebrospinal fluid,

tissue samples and amniotic fluid for the prenatal diagnosis of fetal infections. Early in the course of illness with varicella, PCR analysis of saliva and buccal swabs can also detect the virus. PCR also can be used to distinguish between wild-type and vaccine strain VZV. Viral culture and direct fluorescent antibody assay are less sensitive than PCR and usually not recommended. Positive serology for VZV was not necessary to define as case, but detection of VZV-specific IgM class antibodies by immunoassay procedure was performed in atypical cases. IgM antibody may be detected as soon as 3 days after the appearance of VZV symptoms, but the test may not be reliable [1], [3], [5], [7].

Infection due to the VZV normally affects children. It is associated with several complications, the most serious complication of which is pneumonia. Primary maternal VZV infection during the last trimester can cause pneumonia with significant morbidity and mortality. Pulmonary manifestations, although initially mild, may progress to respiratory failure and cause acute respiratory distress syndrome [6], [8]. The mortality rate due to pneumonia is 10–30% [9]. Varicella-zoster virus pneumonia was defined as the development of respiratory signs and symptoms together with unilateral or bilateral interstitial or alveolar diffuse infiltrates visible on chest X-ray, with or without hypoxaemia, within 10 days after the onset of generalized cutaneous infection due to varicella [8], [10].

Management of congenital varicella consist of VariZig, Acyclovir, and antibiotics. Infant of mothers who develop VZV infection (rash) within 5 days before or 2 days after delivery should receive 125 unit of VariZig intramuscularly as soon as possible and not later than 10 days. Intravenous immunoglobulin (IVIG) 400 mg/kg should be used if VariZig not available. VariZig is not expected to reduce the clinical attack rate in treated newborn, however these infants tend to develop mild infections than the untreated neonates, but it has no benefit once signs of varicella become evident. Prophylactic administration of oral acyclovir beginning 7 days after exposure also may prevent or attenuate varicella disease in exposed infants. Infant of mothers who develop VariZig infection (rash) >7 days before delivery does not need VariZig. It is believed that infants will receive antibodies via the placenta [1], [6].

If signs of neonatal infection develop despite VariZig, the neonate should be treated with Acyclovir 15 mg/kg/dose every 8 hours for 7 days as post exposure prophylaxis as well as treatment in symptomatic neonates. However oral acyclovir has low bioavailability and must be given in frequent doses to achieve therapeutic levels [12], [13]. Antibiotics can be used if secondary bacterial skin infections occur [2], [11].

Prognosis is good if the onset of maternal varicella occurs >5 days before delivery, because the mother has enough time to develop antibodies and pass it to the infant. The infant has mild case of varicella with excellent prognosis. If the mother has onset of disease within 5 days before delivery or 2 days after delivery, the infant is exposed with no antibodies. The disease usually severe with dissemination [1], [12]. Overwhelming sepsis and multiple organ failure lead to mortality rate as high as 30% [9]. The usual causes of death are pneumonia, fulminant hepatitis and disseminated intravascular coagulation. The usage of VariZIG reduce mortality rate to 7%. However there is an increase risk of developing zoster in the first 2 years of life [1], [13].

Prevention of congenital varicella is by avoiding varicella infection and early detection of varicella in pregnant women. Nowadays varicella can be prevented through effective vaccination. The varicella vaccine is a live virus, single-valent vaccine. The vaccine also can be administered in quadrivalent form, in combination with measles, mumps and rubella vaccine. Women in reproductive age who are susceptible to varicella should be offered varicella vaccine at the time of their annual examination or preconception counseling appointment. Similarly, susceptible pregnant women should be offered the vaccine immediately after delivery. Adverse reactions of the vaccine such as mild fever, inflammation and pain at the injection site, and rash are uncommon [14].

## 4.Conclusion

We reported a case of Varicella Congenital to emphasize the importance of evaluation and therapy. Congenital varicella is associated with serious feto-maternal morbidity and mortality. Vaccination against Varicella zoster virus can prevent the disease and outbreak control limits the exposure of pregnant women to the infectious agent. The early recognition of Varicella Congenital would lead to safe and effective treatment by medication which can decrease morbidity and long term complication. Appropriate treatment for Varicella Congenital must be started early by the time of diagnosis in specialized centers.

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