Collodion Baby

A. A. Made Berastia Anis Savitri,[1] Ida Ayu Sri Kusuma Dewi[2]

[1]Bali Mandara General Hospital, [2]Pediatric Department, Bali Mandara General Hospital, Denpasar, Bali, Indonesia

ABSTRACT

Collodion baby (CB) is a rare dermatological condition worldwide of 1: 300.000 birth. A Collodion baby is a newborn whose entire body is covered with an adherent, supple, parchment-like membrane. Collodion baby in a manifestation of various hereditary conditions, including Harlequin orchestras' (HI), lamelar orchestras' (LI), non-bullous congenital ichtyosiform erythroderma (NBCIE), trichothiodystrophy metabolic disease, and endocrine disease. We report two collodion babies cases with peeling skin, passive movement, and poor feeding. Both cases were hospitalized in the humidity-controlled incubator. Water and topical treatment were used for daily bathing to prevent infection.

Keywords: Collodion baby

ABSTRAK

Collodion baby merupakan kelainan dermatologi yang sangat jarang; prevalensi di dunia adalah 1:300.000. Istilah ini mengacu pada kondisi bayi baru lahir dengan kulit yang ditutupi membran tipis seperti perkamen. *Collodion baby* merupakan manifestasi berbagai kondisi yang diturunkan seperti *Harlequin ichthyosis* (HI), *lamelar ichthyosis* (LI), *non-bullous congenital ichthyosiform erytroderma* (NBCIE), *trichothiodystrophy metabolic disease,* dan *endocrine disease*. Kami melaporkan dua kasus bayi *collodion* dengan kulit terkelupas, gerakan pasif, dan kesulitan menyusu. Kedua kasus dirawat di ruang Perinatologi dalam inkubator dengan kelembapan terkontrol dan perawatan topikal untuk mencegah infeksi dan menjaga kulit tetap lembap. **A. A. Made Berastia Anis Savitri, Ida Ayu Sri Kusuma Dewi. Collodion Baby – Kasus Serial**

Kata kunci: Bayi *collodion*

Introduction

Collodion Baby is a rare dermatological condition with a worldwide occurrence of 1: 300.000 birth, the incidence of collodion babies in Indonesia is unknown. Collodion baby was first reported in 1884 by Hallopean.[1,2] The term collodion baby refers to a newborn whose entire body is covered with an adherent, supple, parchment-like membrane. The condition is usually associated with ectropion, eclabium, hypotrichosis, hypoplastic nasal and auricular cartilage, and pseudocontractures.[3] Collodion baby is an uncommon dermatological condition, usually as a manifestation of various hereditary conditions, including Harlequin ichthyosis (HI), lamellar ichthyosis (LI), non-bullous congenital ichthyosiform erythroderma (NBCIE), and trichothiodystrophy metabolic disease and endocrine disease.[4] Differentiating this subtype in the neonatal period is difficult because skin histology in the first week of life isn't specific. Final diagnosis by genetic analyses will help provide proper treatments and genetic counseling.[5] So far, six genes were associated with mutations of TGM1 (transglutaminase 1); TGM1 mediate cross-link in the formation of the cell envelope during terminal differentiation of epidermis.[6-8] Although collodion membrane is a transient condition, it can cause complications and can cause death in the first few weeks of neonatal life. These complications include skin abnormalities (e.g., fissuring, bilateral ectropion, autoamputation of digits due to constricting membrane, and limb swelling), ophthalmologic, gastrointestinal (e.g., poor weight gain, constipation, gastroesophageal reflux), respiratory symptoms, and biochemical abnormalities (e.g., hypernatremia, hypocalcemia, and hypoglycemia). Placing the baby in the humidified incubator is essential; most researchers recommend humidity at 40 – 70%.[9,10] Clinical management also relies on daily bathing with water (with or without a mild cleanser).[9,11] Topical treatment such as urea, alpha hydroxy acids, mainly lactic acid, propylene glycol, vegetable oils such as petroleum jelly, and topical retinoid are used.[12] Artificial tears may be needed in severe ectropion conditions.[9] Enteral feeding starts from the first day of life by nasogastric tube, and because of the high transdermal fluid loss, intravenous fluid 250 mL/kg/day is recommended.[13]

Case Series

We present two cases of collodion babies, one female and one male. The patients came to Bali Mandara Hospital for one year (June 2018 – June 2019).

The first case was a 3-day old female neonate who came to the emergency room with her parents because of passive movement and poor feeding. The baby has dry skin with membrane wraps all over the body, especially her skin fold. This membrane has existed since her birth. The neonate was born aterm from a multigravida mother through sectio caesaria. The indication was premature membrane rupture and poor obstetric history. The baby was born with an APGAR score of 8-9, birth weight of 2.940 grams, length, and head circumference were 49 and 34 cm.

Alamat Korespondensi *email: berastia@gmail.com*

Figure 1. First case at emergency room

Figure 2. First case after 5 days

In the emergency room, the neonate heart rate was 130 times per minute, respiratory rate 40 times per minute, and axilla temperature 36,7°C. Physical examination showed yellow skin from head to thigh with parchment-like membrane and thick scale-covered all over the body, predominantly over the chest, abdomen, and lower extremities. She has ectropion and eclabium but no signs of other abnormalities. There is no history of consanguinity or related dermatological disorders in the family, no history of maternal complications during pregnancy, or maternal drug exposure. The first and third pregnancies were aborted; the second child is now four years old.

Complete blood count showed white blood cell 8.74 x 103 /µL, neutrophils 45.4%, lymphocytes 40.8%, haemoglobin 15.9 g/dL, haematocrit 45.0%, and platelets 223.000. Serum glucose, albumin, and electrolytes within normal range. The patient was diagnosed by a Pediatrician (Fellowship of Neonatology) as a collodion baby, lamellar ichthyosis type with risk of infection. The patient was admitted to the perinatology ward on the humidity-controlled incubator.

Figure 3. First case after 17 days

The second case was a first-day male neonate with a patchy-like membrane covering his body, difficulties opening eyes, and lip stiffness (a fish-mouth appearance). The neonate was born aterm from a multigravida mother with sectio caesarea because of locus minoris resistance (LMR) and premature membrane rupture with cloudy amniotic fluid. APGAR scores were 8-9, birth weight 3.100 gram, length and head circumferences were 51 and 33 cm.

Vital signs in emergency room: heart rate 145 times per minute, respiratory rate of 44 minute per minute, and axilla temperature of 36,8°C. Physical examination showed dry skin with a stiff peeled skin-like membrane covering the whole body, especially the face, head, and all extremities. The neonate also has ectropion and eclabium with no signs of other abnormalities. There was no history of consanguinity or related dermatological disorders in the family. This neonate is the third child with no maternal complications during pregnancy or drug exposure.

Complete blood count results: white blood cell 28.44 x 103 /µL, neutrophils 75.4%, lymphocytes 18.8%, haemoglobin 14.5 g/dL, haematocrit 43.5%, and platelets 288.000.

Figure 4. Second case at emergency room

Figure 5. Second case after 7 days

Glucose level, albumin, and electrolytes serum are within normal ranges. The pediatrician diagnosis was collodion baby lamellar ichthyosis type with risk of infection. The patient was admitted to the perinatology ward on the humidity-controlled incubator.

The first case received 70% humidity in the first week, 60% humidity for the next three days, 50% humidity for the next four days, 40% humidity for the next one day, and the next two days in the crib. The second case received 70% humidity in the first week, 65% humidity for the next three days, 50% humidity for the next two days, 40% humidity for the next day. Since the first day in the perinatology ward,

adequate body temperature and fluid were maintained by breastfeeding, orogastric tube, and parenteral nutrition. The first case did not need an orogastric tube since her sucking reflex was still good. Adequate fluid nutrition was given by breastfeeding and the parenteral route. The second case neonate needed an orogastric tube because of the stiffness and dryness of his lips. The orogastric tube was removed after five days because his sucking reflex was already adequate.

Figure 6. Second case after 13 days

The pediatrician prescribed ampicillin 50 mg/kg/dose twice a day and gentamicin 5 mg/kg/dose every 36 hours intravenous for five days and was stopped after the result of bacterial culture was sterile, and the condition was stable.

A combination treated skin lesions of hydrocortisone 2.5% and chloramphenicol cream. Dermatologists suggested histopathology and pathology anatomy examinations. The ophthalmologist told artificial tears twice a day to avoid dry eyes for the first case, while the second case was given levofloxacin eye drop and gentamicin eye ointment three times a day. ENT doctor confirmed no abnormality.

The neonates were bathed with special soap containing lactic acid, lactoserum, and pH for normal skin. After the bath, mixed retinoid cream prescribed by the dermatologist was given to the whole body, and after 2 hours, petroleum jelly was given for moisture.

The parents did not consent to genetic analysis, histopathology, and pathology anatomy examination.

The first case was discharged after 17 days with good general health condition. The peeling skin only appears in the forehead, chest and abdomen. The second case was discharged after 13 days with better health condition.

Discussion

Collodion Baby is a rare dermatological condition; the entire body is covered with an adherent, supple, parchment-like membrane. The collodion membrane exerts mechanical compression, distorting the face and extremities, resulting in a striking appearance that may initially frighten the family members and physicians who may have never confronted such a case. These newborns can have malformed ears, averted eyelids (ectropion), and lips, resulting in a fish-mouth appearance.[1,3,14]

A 3-day old female neonate and a 0-day old male neonate came to our hospital with dry skin, and membrane wraps over the whole body with predominance over the chest, abdomen, and both of the lower extremity for the first case and was predominantly in his face, head, and all extremities for the second case. On physical examination, we found the fish-mouth appearance (eclabium) and ectropion which are pathognomonic for collodion babies. Laboratory examination results were within normal limits, but there was a slight increase of white blood cells for the second case. The bacterial culture of these cases was also sterile.

Due to the impairment of skin barrier function, these neonates risk many complications, including hypernatremic dehydration, thermal instability like hypothermia, percutaneous toxicity, infection, fissures, conjunctivitis, sepsis, dehydration, and constriction bands of the extremities resulting in vascular compromise and edema.[15-17] There is a high risk of dehydration and electrolyte imbalances and high insensible water loss from the skin defect, requiring placement in a high-humidity incubator, close monitoring of body temperature, and good nutritional support in enteral and parenteral forms.[14-16] Placing collodion baby in the humidified incubator is essential, with the incubator humidity set at 40-70%. For daily bathing, use water (with or without a mild cleanser) and topical treatment (e.g., urea, alpha hydroxy acid, lactid acid, propylene glycol, vegetable oil, and retinoid topical). And artificial tears may be needed if the baby has ectropion.[3,9,11]

The cases were treated in the incubator with 40-70% humidity, adequate body temperature, and fluid maintained by breastfeeding, orogastric tube, and parenteral nutrition. Ampicillin and gentamicin were prescribed in both cases because of the high risk of sepsis neonatorum.[17] They were bathed with special soap containing lactic acid, lactoserum, and pH for normal skin. Artificial tears were prescribed twice a day for the first case, and levofloxacin eye drops and gentamicin eye ointment three times a day for the second case. After the bath, mixed retinoid cream prescribed by the dermatologist was given to the whole body, and after the next 2 hours, petroleum jelly was given for moisture.

The neonate's skin moisture should be maintained because of impaired skin barrier function risk. Although the collodion membrane is only a temporary condition of the newborn, neonatal complications can occur in 45% of collodion babies, leading to a mortality rate of ~11% in the first few weeks of life. Morbidity and mortality during the neonatal period are increased due to an impaired skin barrier function.[16,18] These cases were discharged with a good general health condition.

Table 1. Humidity of incubator for first case

Day	Humidity of Incubator
1 – 7	70%
8 – 10	60%
11-14	50%
15	40%

Table 2. Humidity of incubator for second case

Day	Humidity of Incubator
1 – 7	70%
8 – 10	65%
11-12	50%
13	40%

Loss of skin integrity may cause hypothermia, increased insensible water loss and electrolyte disorders, skin infection, and sepsis. Management of this devastating condition requires a multidisciplinary approach with the combined effort of dermatologists, neonatologists, and ophthalmologists.[10,17] The management strategy should focus on protecting skin barrier, infections control, maintenance of fluid and electrolyte balance, and early initiation of retinoid therapy,[17,19] accompanied with enhanced moisturization, electrolyte monitoring in the early neonatal period, sepsis prevention and physiotherapy, long term survival of 81% has been reported.19 Both patients were continued to be under multidisciplinary surveillance and management.

Conclusion

Collodion baby is rare but may suffer from complications and death in the early weeks of life. Management is to prevent infection, to keep skin moisture through the humidified incubator, topical treatment, and nutritional support.

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