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A RETROSPECTIVE STUDY ON 16 COLLODION BABIES*

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Sixteen collodion babies followed in the Neonatal Care Unit between January 1982 and December 1994 were evaluated retrospectively. The preterm/term ratio was 1.6, and complete shedding of the collodion membrane took an average of 21.9 days (range 18-46 days). Problems noted were marked temperature instability, defective barrier function, increased insensible water loss predisposing to hypernatremic dehydration, cutaneous infections and septicemia. Hypernatremia was observed in 11 (68.7%) and septic infection in seven patients (43.7%). All the infants were treated topically with vaseline containing five percent lactic acid. In the hypernatremic infants, intravenous fluid was administered for rehydration. In the septic infants, antibiotics were used according to the antibiogram. Four of the infants died due to septicemia. The mortality rate was 25 percent, and the major complications included hypernatremia, cutaneous infection and sepsis. *Key words: collodion babies.*

Collodion baby is a rare congenital disorder that resembles harlequin fetus but is milder in degree[1]. The infant is born tightly encased in a shiny membrane resembling parchment or oiled silk, which is perforated by scalp and lanugo hair[1-3]. Collotion babies are often born prematurely[2, 4]. Because of the presence of ectropion, eclabium and flattening of the ears and nose as well as fixation of the lips in an O-shaped configuration, these infants resemble one another in the first few days of life[1-3]. Motion of the limbs is restricted[1]. Soon after birth the collodion membrane peels off, leaving reddish skin which can very easily become infected and may result in pyoderma, causing fatal sepsis and pulmonary infections within the first days or weeks of life. Complete shedding of the collodion membrane may take several weeks[2, 3].

Occasionally, an affected infant is observed to have normal skin subsequent to shedding of the membrane[1, 2]. The collodion baby probably represents a phenotype for several genotypes[1-3]. Complications include marked temperature instability, skin irritation, water loss via the epidermis predisposing to hypernatremic dehydration, pyodermia, septicemia and pneumonia secondary to aspiration of squamous material in the amniotic fluid[1-4, 6]. Cutaneous infections from gram-positive organisms, candida albicans and milk aspiration are common problems[1].

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Supportive care is of primary importance for management of the collodion baby. The encasing membrane should not be manually removed. In the treatment of collodion babies, application of topical antikeratinogenic agents have been used, and measures have been taken to prevent other complications[4-6].

Material and Methods

A total of 16 collodion babies were retrospectively evaluated. All patients were treated in the Neonatal Care Unit at Erciyes University Faculty of Medicine between January 1982 and December 1994. Physical examination, whole blood count and blood chemistry (except in the 1st and 3rd patients) were performed for clinical evaliation. Blood, urine and/or cerebrospinal fluid cultures were taken from patients who had a septic appearance. All patients were diagnosed according to clinical findings. Histopathological examination of the skin was not performed in all patients. Four of the 12 patients who lived were reexaminated when they were two, five, eight and ten years old.

All the infants were treated topically with vaseline containing five percent lactic acid. In the hypernatremic infants, intravenous fluid was administered for rehydration. In the septic infants, antibiotics were used according to the antibiogram, The hyperbilirubinemic infants received phototherapy. An exchange transfusion was given to one infant in addition to phototherapy. In one hypoglycemic infant who had sepsis, concentrated glucose fluid was administered.

Results

The mean age of patients at the time of diagnosis was 17 hours (range 1-62 hours). Ten of the 16 patients were preterm, with a preterm/term ratio of 1.6. The mean age of the preterm infants was 36.1 weeks (range 35-37 weeks). The male/female ratio was 2.2 (Table I). In all but one case the parents were first-degree relatives. Five patients had a family member with the clinical appearance of a collodion baby. Patients were followed for one to 41 days (average 14.7 days). Hyperbilirubinemia complicated the course in eight patients and was managed with phototherapy. One patient needed an exchange transfusion.

Hypernatremia was a frequent complication, observed in eleven patients (68.7%), and improved within the first month of life. Complete shedding of the collodion membrane took an average of 21.9 days (range: 18-46 days, Table I).

Infection occurred in seven patients (43.7%) four of whom were preterm. These infections included sepsis, cutaneous infection and urinary tract infection. Sepsis occurred in five patients, while cutaneous infection due to Staphylococcus albus accompanied sepsis in two cases. In two cases, cutaneous infection and urinary tract infection were the only presenting symptoms. A case who had hypoglycemia,

consumption coagulopathy and cutaneous infection was considered to be septic in spite of the fact that no microorganism was isolated in the blood culture. Microorganisms isolated from blood cultures were proteus and enterococcus in two patients and candida albicans in two other patients with sepsis. Four patients died, yielding a mortality rate of 25 percent.

Table I: Cases of Collodion Baby

Case no/Age (hours)/Sex	Gestational age (Weeks)	Initial serum Na^+ values (mEg/L)	Complete shedding of collodion membrane (days)	Outcome
1/05/B	40	–	–	Exitus
2/19/B	36	132	18	Unknown
3/24/B	40	–	–	Exitus
4/36/B	36	143	25	Unknown
5/05/B	35	168	23	Unknown
6/48/B	37	161	26	CNBIE
7/48/G	38	158	21	CNBIE
8/12/G	36	138	19	Unknown
9/04/G	36	157	21	Unknown
10/24/B	40	159	25	Unknown
11/07/B	36	144	–	Exitus
12/48/G	36	141	37	CNBIE
13/03/B	38	144	–	Exitus
14/03/G	36	160	28	CNBIE
15/62/B	37	148	46	Unknown
16/17/B	40	138	31	Unknown

B: Boy, G: Girl, CNBIE: Congenital non-bullous ichthyosiform erythroderma.

Four of the 12 patients who survived were reexaminated later in their lives. Normal physical and neurological development, but mild ichthyotic scaling was observed in all patients. There was no information about the outcome in the other eight patients.

Discussion

Collodion babies are seen rarely. This condition often eventuates in lamellar ichthyosis or congenital ichthyosiform erythroderma but may evolve into other disorders, such as Netherton's and Conradi's syndrome, ichthyosis vulgaris, recessive X-linked ichthyosis and Sjögren-Larsson syndrome[2,4,7]. Van Scott[8] suggested a classification for ichthyosis with four groups based on anatomical, cellular, kinetic and genetic observations, which seem to provide the best framework for approaching these disorders.

The presence of positive family histories, the absence of congenital abnormalites, and normal motor and mental development led us to conclude that the collodion

appearances in the four patients who were reexaminated later in their lives were manifestations of the autosomal-recessive form of lamellar ichthyosis (congenital non-bullous ichthyosiform erythroderma).

Light microscopically the neonatal collodion skin is characterized by a thick compact stratum corneum which was PAS positive in its upper two thirds, a thin stratum granulosum, and a non-acanthotic stratum spinosum with normal mitotic activity[9].

Two siblings with Netherton's syndrome complicated by neonatal hypernatremia have recently been reported[6]. It is known that hypernatremia may occur when there is a considerable increase in the area of thin erythematous skin or increased skin loss (burn, phototherapy, preterm or generalized dermatoses)[6].

In the presented cases, histopathological examination of the skin was not performed. In eleven of our patients, serum sodium (Na^+) levels were greater than 140 mEq/L in the absence of symptoms such as vomiting, tachypnea, polyuria, fever and ambient temperature, suggesting transepidermal water loss. Moreover, the hypernatremia was improved within the first month of the life in all patients. In one study, transepidermal water loss values four days after birth were shown to be abnormally high compared with those of normal infants of the same age. The transepidermal water loss measurements returned to normal within the first month, parallel to the improvement of both the skin signs and the electrolyte and fluid balance[10].

Loricrin and involucrin are major precursor proteins to the cornified cell envelope expressed late in epidermal differentiation[11]. Involucrin represents 25 percent of the living epidermis in normal humans and 38 percent in collordion babies.

Kanitakis et al.[12] demonstrated altered involucrin expression in a variety of keratinization disorders including collodion babies, and suggested that in such conditions, cellular functions other than keratin metabolism are also affected.

Larreque et al.[13] showed in 267 collodion babies that the mortality rate was 33 percent in 1976 and 11 percent in 1984. Skin infection with systemic spread was the major complication. Larreque et al[14] in another study showed that the collodion membrane in 198 collodion babies spontaneously desquamated between the 15th day and the third month of life, and during the neonatal period one-third of the infants died due to pulmonary complications or infection.

Topical preparations containing alphahydroxy acids and closely related compounds have been found to exert profound antikeratinogenic effects on ichthyosis or other forms of epidermal keratinization[8, 15]. A topical preparation containing five percent lactic acid, an alphahydroxy acid, has been firstly exerted in the treatment of collodion babies in Turkey and by Erdem[5, 16]. He reported that shedding of the collodion membrane took an average of 16 days (range

11-22 days) on five collodion babies. In addition, he observed decreased mortality with this compound. In our study, this duration was longer in spite of using the same therapy. Its cause may be related to supportive care, the occurrence of cutaneous infections and prematurity.

In this study, the mortality rate was 25 percent and the major complications were hypernatremia, cutaneous infection and sepsis. Four of the seven patients with infections were preterm. Aside from the primary disorder, prematurity can also a predisposing factor for infection and mortality.

Supportive care is of primary importance in the management of the collodion baby. These patients are best managed in a humidified incubator, with special attention given to the prevention of temperature instability, cutaneous infection, sepsis, and fluid and electrolyte imbalance. The skin should be covered with vaseline and gauze to prevent irritation and water loss.

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