

Collodion baby

Januar 2015



Soroken C, Scerba F, Calza AM, Karam O, Neonatal Intensive Care Unit, University Hospitals of Geneva, Switzerland

Title figure: Wikipedia

Collodion baby (CB) is rare, estimated to occur once in 50'000 to 100'000 deliveries (1). Less than 300 cases of collodion babies (CB) have been reported in the literature. Here, we present a CB with favorable outcome.

This female patient was born at 37 weeks of gestation via vaginal delivery after an uneventful pregnancy. She was the first child of the family, and the parents were not consanguineous. The mother had a medical history of skin disorders: desquamative erythrodermia in infancy and Lyell syndrome secondary to the administration of various medications (non-steroidal anti-inflammatory drugs, myorelaxants).

Postnatal adaptation was excellent with Apgar scores of 9, 10, 10 at 1, 5 and 10 minutes, respectively. The first impression was that the newborn was covered by a thin parchment-like glossy skin (Fig. 1). The face showed discrete bilateral ectropion, absence of eye brows and small eyelids (Fig. 2). In addition, there was sparse hair, discrete eclabion, wrinkled ears, and skin splitting localized in flexural areas (Fig. 3). Mobility of all joints was preserved. There were no extracutaneous symptoms except for intrauterine growth restriction: birth weight was 2320 g (P 5 – 10), head circumference 33 cm (P 10 – 25), length 43 cm (< P3).

Although sucking was preserved, an umbilical venous catheter was inserted in order to ensure proper caloric intake. Parenteral alimentation was initiated in parallel to enteral feeding. Complete and active feeding was achieved by day 10 of life with appropriate postnatal weight gain.

The infant was placed in a humidified incubator with 90% humidity and close monitoring of body temperature. Frequent monitoring of electrolytes did not reveal any abnormalities, suggesting near to normal transdermal water losses. She was able to be transferred from the incubator to an open crib at one week of life.

Our dermatologists suggested applying skin emollients (petroleum jelly alternating with dexpanthenol every 3 hours). The first bath was given at one week of life. Almost complete shedding of the collodion membrane was observed after two weeks, leaving generalized thin white scaling, with desquamation involving palms, soles and scalp (Fig. 4). On day 12 of life, aseptic pustules were observed on the neck and scalp (Fig. 5). With regards to the ectropion, ophthalmologic examination revealed no signs of keratitis or infection, and artificial tear drops were initiated.

In search of an underlying condition, extensive investigations were undertaken. There was no evidence

of a metabolic disorder. Abdominal, renal and cerebral ultrasounds as well as a chest X-ray were normal. Hair analysis did not reveal the pathognomonic feature of Netherton syndrome, i.e. trichorrhexis invaginata (bamboo hair) (2, 3). Since this symptom is not regularly detected, its absence could not exclude this disease entity with certainty (3).

Given the fact that the baby was otherwise stable, and because the phenotype of the underlying diagnosis often becomes evident only after weeks or months of follow-up, we decided to wait and observe the evolution without further investigations (i.e., a skin biopsy was not done, blood for genetic testing was sampled but not analyzed). The patient was discharged at two weeks of life.

Two weeks later, at one month of life, she was seen by the dermatologists. By then, the glossy thin membrane had disappeared completely, leaving some large scales on the inferior part of the back and around the contact point of the diapers. The volar aspects of the palms and soles remained intact as well as the nails. Daily creaming with dexpanthenol was prescribed. At two months of life, the daily moisturizing was discontinued given the fact that the skin was perfectly normal. This evolution confirmed the diagnosis of self-healing collodion baby (SHCB).



Fig. 1

Collodion membrane with fine scaling.





Fig. 2

Discrete bilateral ectropion, absence of eye brows and small eyelids, sparse hair, discrete eclabion.





Fig. 3

Skin splitting over flexural areas of the joints.



Fig. 4

Skin desquamation at the age of two weeks.



Fig. 5

Aseptic pustules observed on day 12 of life.

Although the collodion membrane is only an evanescent phenomenon in the newborn, neonatal complications can occur in 45% of all CBs, leading to a mortality rate of up to 11% in the first few weeks of life (2). Common complications are marked temperature instability (hypothermia), increased insensible water loss predisposing to hypernatremic dehydration, cutaneous infections and septicemia (1–4).

Given the disruption of the skin barrier, the initial management of these patients includes:

- Nursing of the newborn in a highly humidified incubator and frequent monitoring of electrolyte balance
- Application of greasy emollients several times a day
- Close supervision for signs of cutaneous or systemic infections
- Ophthalmologic evaluation and follow-up

The diagnosis of CB is made clinically; it is characterized by a shiny tough transparent membrane, resembling cellophane wrapping stretched over the skin. The neonatal presentation differs greatly from the later mature phenotype because of the differences between the wet, intrauterine environment and the dry, postnatal environment.

More than 60% of infants born with a collodion membrane eventually develop ichthyosis (4, 5). The term «ichthyosis», from the Greek word for fish, is

used for those disorders sharing generalized scaling of the skin. Differentiation of ichthyosis subtypes in the neonatal period is difficult. Skin histology in the first few weeks of life is not specific, and therefore not helpful. The final diagnosis emerges after weeks or months of follow-up and depends on the genetic analysis.

In 2009, the first ichthyosis consensus conference was held and established an international nomenclature and classification of inherited ichthyoses: syndromic versus non-syndromic forms (3, 6). Six major distinct clinical subtypes of hereditary autosomal recessive non-syndromic ichthyoses were identified: Harlequin ichthyosis (the most severe form), lamellar ichthyosis (LI), non-bullous congenital ichthyosiform erythroderma (NBIE), epidermolytic ichthyosis (EI), recessive X-linked ichthyosis and ichthyosis vulgaris (IV) (6). Based on this classification, once the clinical subtype is suspected, the results of genetic analyses will help to provide proper treatment and genetic counselling. In severe congenital ichthyosis, DNA-based prenatal diagnosis is possible (3, 7).

More than 60% of affected CB will develop one of the following two subtypes later in life (2, 4, 8): LI (classic clinical findings are large, dark plate-like scales with little or no erythema) or NBIE (fine scale with prominent erythema). In approximately 10–20% of CB cases (as in our case), the ichthyosis phenotype

may improve spontaneously within first three months of life, leaving nearly normal-appearing skin (1–3, 5, 8). This entity has been described as self-healing CB (SHCB) (1, 2, 4, 8). Because many of these patients, when re-examined later in childhood or as adults, have a variable degree of anhidrosis, heat intolerance and mild signs of ichthyosis such as xerosis and fine desquamation, particularly in the axillary and neck region, the term self-improving collodion ichthyosis may be more appropriate (8).

Histological analysis of skin biopsy specimens taken in the first weeks of life usually show nonspecific patterns and are therefore not helpful in differentiating between the various forms of ichthyosis. Some findings on electron microscopy have been reported to help predict whether a baby will ultimately have normal skin or ichthyosis. However, these findings may be misleading (9). To date, mutations in eleven genes have been identified to cause ichthyosis in human patients (6). In the case of SHCB, mutations can be found in the TGM1, ALOXE3 or ALOX12B genes encoding, respectively, for transglutaminase 1 (involved in the cornification of the stratum corneum) and for arachidonate 3 and 12 lipoxygenase (involved in lipid metabolism) (1, 3, 7–10).

CONCLUSION

Ichthyosis is a rare condition that requires significant attention in the neonatal period. Successful management in the newborn period requires an interdisciplinary approach. Families should be offered proper genetic counselling, psychological support and receive information about relevant patient organization and foundations (see: www.ichthyose.ch) (3, 7).

1. Prado R, Ellis LZ, Gamble R, et al. Collodion baby: An update with a focus on practical management. *J Am Acad Dermatol* 2012;67:1362-1374 ([Abstract](#))
2. Chiavérini C. Congenital ichthyosis. *Ann Dermatol Venereol* 2009;136:923-934 ([Abstract](#))
3. Oji V, Tadini G, Akiyama M, et al. Revised nomenclature and classification of inherited ichthyoses: results of the First Ichthyosis Consensus Conference in Sorèze 2009. *J Am Acad Dermatol* 2010;63:607-641 ([Abstract](#))
4. Larrègue M, Ottavy N, Bressieux JM, et al. Collodion baby: 32 new case reports. *Ann Dermatol Venereol* 1986;113:773-785 ([Abstract](#))
5. Aradhya SS, Srinivas SM, Hiremagalore R, Shanmukappa AG. Clinical outcome of collodion baby: a retrospective review. *Indian J Dermatol Venereol Leprol* 2013;79:553
6. Akiyama M. Updated molecular genetics and pathogenesis of ichthyoses. *Nagoya J Med Sci* 2011;73:79-90 ([Abstract](#))
7. Rodríguez-Pazos L, Ginarte M, Vega A, Toribio J. Autosomal recessive congenital ichthyosis. *Actas Dermosifiliogr* 2013;104:270-284 ([Abstract](#))
8. Vahlquist A, Bygum A, Gånemo A, et al. Genotypic and clinical spectrum of self-improving collodion ichthyosis: ALOX12B, ALOXE3, and TGM1 mutations in Scandinavian patients. *J Invest Dermatol* 2010;130:438-443 ([Abstract](#))
9. Harting M, Brunetti-Pierri N, Chan CS, et al. Self-healing collodion membrane and mild non-bullous congenital ichthyosiform erythroderma due to 2 novel mutations in the ALOX12B gene. *Arch Dermatol* Mar 2008;144:351-356 ([Abstract](#))

10. Raghunat M, Hennis HC, Ahvazi B, et al. Self-healing collodion baby: a dynamic phenotype explained by a particular transglutaminase 1 mutation. *J Inv Dermatol* 2003;120:224-228
(Abstract)

SUPPORTED BY



CONTACT

Swiss Society of Neonatology

www.neonet.ch

webmaster@neonet.ch