Neonatal Care for Ichthyosis or Epidermal Differentiation Disorders

CME Series on Epidermal Differentiation Disorders — Presented by Dr. Jinia El-Feghaly

---

00:00

Welcome to Neonatal Care for ichthyosis or Epidermal Differentiation Disorders, part of First's CME series on EDDs or ichthyosis.

00:09

Presenting this topic is Dr. Jinia El-Feghaly. Dr. El-Feghaly is chief of pediatric dermatology at Golisano Children's Hospital, part of the University of Rochester Medical Center.

00:20

She is also an adjunct professor at the American University of Beirut. She cares for infants and children with complex genetic skin conditions, including EDD and has published on the acute management of newborns with collodion membrane.

00:35

Today we are going to speak about neonatal care for patients who suffer of ichthyosis or EDS.

00:45

I do not have any relevant financial relationships to disclose for the stock. Our learning objectives for today recognize the various aspects of the care for a neonate with a suspected EDD, Implement appropriate acute management for a neonate with a suspected EDD, Support the family throughout the journey of neonatal care for a suspected EDD. Please do not reproduce these images.

01:16

A group of multidisciplinary specialists has recently joined forces to build a consensus based guideline for the care of the neonate with ichthyosis or epidermal differentiation disorders. In the next slides, we are going to go over the major points of this consensus, and we are going to share all of this information with you so we can support our patients.

01:48

First, what is a baby with congenital ichthyosis or EDD? What does that mean? It is a very important congenital disease to recognize as soon as possible. Early recognition is vital. Congenital ichthyoses are rare. They are genetic skin disorders that present with scaling,

erythema or redness, and profound skin barrier dysfunction. Many of these present with what is known as the collodion membrane.

02:24

This is a tiny, shiny, translucent, parchment like skin layer that in cases the newborn, as seen in the image here.

02:34

These patients will have a high risk neonatal course, which frequently mandates immediate NICU admission and urgent dermatologic consultation. Therefore, we have core pediatric priorities to address in the slides, and it's very important to also know who to reach out to in these situations. The tautness of the skin directly risks the restricted chest excursion, ectropion, eclabium, severe fluid loss, temperature dysregulation, and systemic sepsis as well.

03:18

So how to make the diagnosis? To be able to make the diagnosis of a baby with collodion membrane, or a newborn with a suspected EDD we need to obtain a very detailed, comprehensive family history to construct a pedigree and determine potential inheritance pattern, including the possibility of consanguinity. We also need to begin the evaluation based on a predominant clinical feature that we notice upon our skin exam, as well as other features that we note.

03:53

These changes that are phenotypic help us further delineate a specific genotype and guide our next steps. Initiating genetic testing as soon as possible is crucial because it also helps guide our management plan and the therapies that we are looking into. Genetic counselors are very helpful in these conditions, as well as our geneticist teams that can that can help us in further supporting the family, the parents, as well as ordering the genetic testing that is needed.

04:33

Please remember, routine histology is not diagnostic for most EDDs. That means we do not have to biopsy all these babies to make the diagnosis. This is a clinical diagnosis. Neonatologist and or the primary health care team should initiate consultation with dermatology, typically pediatric dermatology, if available at your center or in the area where your medical center is, genetics and others as needed

05:04

based on the history, the clinical exam to guide further diagnostic and therapeutic plans.

05:15

What is the next step? What is the environment that we need to prepare for these babies? Place the neonate with suspected EDD in a humidified incubator. The question that always comes up is what level of humidity should we have? Well, typically we will say it is about 40% up to 80% humidity. Maintain a constant environmental temperature within the isolate and encourage parental contact.

05:46

if the neonate is stable and of sufficient gestational age. This is very important to help the child grow developmentally well, because the connection with the parents is very important from the beginning. It helps create a better relationship with the family also between the patients and their parents. Implement protocols for initiating and weaning humidity based on severity and other factors.

06:14

Neonatologist are typically very well aware of how to take care of these little humidity and temperature regulations. Follow standard unit protocol for weaning to a crib that depends on your institutions protocols. Avoid dressing in clothing or swaddling in the isolate until the collodion membrane is shed. And this is a very important point. We frequently get questions about this topic because a lot of times we feel that we want to dress these babies, but technically we should not be over dressing in in clothing or swaddling the kids so that they can get enough humidity and for their collodion membrane to shed properly.

07:03

Phototherapy for neonatal hyperbilirubinemia, if required, is safe for the neonates covered in emollient.

07:15

What about bathing and moisturization?

07:20

Delay the first bath for more than 24 hours after birth to retain the vernix caseosa, and bathe at least every 2 to 3 days thereafter. This is very important to clean the skin from all the bacteria, particularly being in a high humidity environment. Use non irritating fragrance free soap free cleansers. Avoid mechanical removal of the collodion membrane as it will shed spontaneously in the first weeks of life.

07:50

It is crucial to respect cultural bathing practices while ensuring a safe care for the newborn. Educate parents on proper techniques for bathing and maintaining a warm environment. Pat the skin - do not scratch or rub. Use thicker, more occlusive moisturizers versus creams. Typically,

more occlusive moisturizers are more oily and help the skin get better moisturization versus the creams that are usually more white have a lower moisturizing factor compared to the occlusive moisturizers. In these situations,

08:29

these occlusive moisturizers are highly recommended. Petrolatum and glycerin based products are safe. Reapply the most in a visible layer several times a day, particularly after bathing. Monitor for potential contact dermatitis. Some people also know that as allergic contact dermatitis or irritant contact dermatitis, and suspect a potential trigger based on the distribution of the reaction. Please avoid,

09:01

or use with caution,

09:03

Topical salicylic acid, alpha hydroxy acids, vitamin D3 analogs, corticosteroids, and other anti-inflammatory agents due to the risk of systemic absorption and toxicity. We will not go into more details regarding these specific toxicities. This is beyond our talk today. But please, this is a very important point to remember for many neonates, particularly neonates who have a skin barrier dysfunction.

09:33

As you can see in these images here, both these newborns who have ichthyosis with erythroderma a lot of erythema and redness and scaling are very well moisturized with the shining that you can see, on the skin. And this is the goal. A visible layer should be seen of an ointment and an occlusive moisturizer on the skin of these infants.

10:01

What about feeding and nutritional support? Well, because of the eclabium that we can see here in this picture, a child who has neonatal EDDs cannot really close their mouth properly. That not only affects breathing, but it also affects their ability to feed. And in these cases, this very tight skin is impeding our ability to really feed this child.

10:31

How can we take care of that and support the growth of the child? If the parent is interested in breastfeeding, please encourage that. And if not, we can also encourage breast milk conservation. If the child is not able to to breastfeed at that moment, include a lactation consultant and or speech swallow therapist as needed to support oral feeding.

10:58

Monitor for the ability to breast or bottle feed neonates with eclabium. Use high calorie foods and provide specific protocols for feeding, guided by early consultation and close follow up with nutritionists, it is particularly important to have a high caloric foods because kids who have EDDs have a higher tendency to lose a lot of their energy because of

11:25

this function that we are seeing in the skin. If needed, supplemental feeding can used. We can use an NG, an OG, or a G tube, feeding or parenteral nutrition if needed. Increased caloric and protein intake is often required. As I mentioned already, with the highest levels of nutritional supplementation necessary in erythrodermic neonates or the neonates who have a lot of redness on their skin.

11:59

Pain and itch management. This is very important point because a lot of times we are not able to really assess these little changes and symptoms that the newborn is feeling. However, there are certain signs and symptoms of pain and itch that we are able to see while we are examining the child, some of which are shown in the vital signs, some of them in the child being irritable.

12:29

Early engagement of a pediatric or perinatal supportive care team is recommended. Involving the family in the decision making regarding pain and pruritus management is also crucial. Providing sensory tools such as music and soothing sounds for neurodevelopmental support and distraction is a very important point in this situation. And how can we do that? By involving child life support and the institution, particularly for full term neonates.

13:04

Because the neurodevelopment of these kids is important as they grow in the NICU and stay in the NICU for a couple of weeks, sometimes months, identifying pain and pruritus through the vital signs, through the other symptoms that we can see in the newborn is also important. And neonatologists and NICU nurses are pretty familiar with identifying signs of pain and discomfort in a child.

13:35

Finally, apply emollients often to minimize potential discomfort from dry skin and fissuring.

13:47

Ocular considerations. Consult with ophthalmology when eye abnormalities are present, particularly ectropion, as seen in this figure here, corneal opacities, conjunctival injection or epiphora. Please use preservative free lubricating eye ointments as frequently as every hour for

ectropion. Because of ectropion, the eyes tend to dry a lot and therefore the importance of lubricating eye ointments. Culture eye discharge when seen, particularly if it is yellow or green.

14:25

What about the auditory considerations? Well, a lot of babies who are born with EDDs can have built up of skin, particularly in the external meatus. Review the results of hearing screening, given the hearing loss in many forms of EDDs is also important. If you identify ear or hearing abnormalities, please consult ENT or otolaryngology. Otolaryngology team typically removes the ear canal cerumen and debris, particularly when they build up in the ears, hence the importance of this

15:06

Lines:

15:09

In these cases, particularly when in the NICU, the nurses and the NICU neonatologists are pretty familiar with a lot of lines being needed to support the growth and the care of these infants. Adhesives often adhere poorly to the neonatal EDD skin and can induce injury, particularly when we are applying a lot of emollients. And this is a very important point.

15:35

As you can see in this image here, a lot of times we can use some extra or additional ways to help stick these lines in their place for a longer period of time. A very common tool is using the arms or the sides to help with that. We prefer non or minimally adherent dressings on the skin. Secure any lines or tubes by affixing tape to the dressing material rather than to the skin, and this is another important tip.

16:10

Central venous umbilical or peripheral line access is important in more severely affected neonates. Some forms of EDDs are associated with early respiratory issues and may require invasive or noninvasive ventilation.

16:28

Monitoring. How to monitor these babies. Most infants with EDDs require enhanced observation, including consideration of NICU admission. Base the frequency of temperature, weight, fluid and electrolyte balance monitoring on prematurity and clinical needs. We cannot have one rule for all newborns with these conditions. It is very important to have the clinical

16:59
thinking and the critical thinking in the NICU with the big experts in neonatal care to be able to support these newborns.

17:10
The frequency really changes based upon many factors. This is why there is not one set or a frequency or factors that we could really delineate here, but the things that we really should keep an eye on are the temperature, the weight, the fluid and electrolyte balance. Because the skin, again is dissipating a lot of heat because of the skin barrier.

17:34
This function and these newborns are not really able to regulate their temperature properly. Include assessment of serum proteins, albumin and electrolyte levels, and initial monitoring of fluid balance, and evaluate ongoing fluid status through urine output and serial weight measurements. As we do for a lot of our premature babies in the NICU.

18:01
What about infections? Because of the disrupted epidermal barrier, there is a higher risk for bacterial and fungal skin infections, particularly staff Pseudomonas and Candida. Despite the importance of frequent emollient and humidification for these babies, the practices of particularly these frequent emollients and the humidification increase the risk of these specific microbes. We need to consider infection and a differential diagnosis of temperature instability, apnea, bradycardia, tachycardia, respiratory distress, and other common clinical and lab signs of neonatal infection.

18:45
What are the skin lesions that could raise our suspicion for skin infection? Pustules and purulence, vesicles, erosions and crusting. And a very important sign is increase in erythema and odor. Please culture the skin and skin wounds as clinically indicated. And if you suspect an infection, collect these cultures as soon as possible. Complete standard of care surveillance cultures on admission and as per protocol based on your institution.

19:22
You can see in this image here a few signs of infection, particularly the increase in erythema in the armpit right here. Please use antibiotics only if clinical signs of infection are present, and select broad spectrum antibiotics based on the clinical suspicion. IV antibiotics are not always recommended, but are appropriate for skin infections and necessary for widespread infection as needed.

19:51
Whenever we use topical antibiotics, we have to be cautious of the risk of absorption as well, so they should be used only for localized infections in well appearing neonates.

20:05
There are additional considerations in neonates with Keratinopathies, or Epidermolytic EDDs. And the ABCA-12-nEDD or Harlequin ichthyosis, as well as other procedural considerations. I will not go over this in this talk. Typically, in these situations, we should really have a dermatologist as part of our team to be able to take care of these patients.

20:35
Technically, it's always better to have a dermatologist with us for any form of EDDs,

20:41
but these ones require more advanced care.

20:45
For many families, having a baby with EDDs is a new thing, or is a first time thing. For others, they've had a sibling or another family member with EDDs and they're more familiar with it. It is very important for us as health care providers to support these families throughout this process and help them with understanding the disease and the condition.

21:13
It is true that a lot of times, these babies have to stay in the NICU for the first few weeks to the first few months of life. However, they grow to become great people with the support of the healthcare providers, their families, and a lot of organizations out there, particularly First who can provide these families with resources to support them at a social level, at the financial level as well at the medical level.

21:46
So we need to ensure that the parents and the caregivers receive comprehensive social, psychosocial and complex care management support throughout the stay and the hospital and throughout this neonatal period. Engaging supportive care teams from the get go is very important to facilitate this holistic, longitudinal, long term care and the shared decision making. Involving the families and the caregivers and the care decisions by including them on a daily basis in the medical rounds and the multidisciplinary discussions, is very important for them to first understand what's going on, familiarize themselves with this condition, and also able to feel that they are there for their child and supportive of their child.

22:42

Respecting integrating the family's cultural values is of utmost importance, as we've discussed before, and that is something that we value a lot as health care providers. Encourage early parent infant bonding as clinically appropriate from day one, if possible. And remember that a lot of times parents might not really know a lot, might not ask the questions. So we need to be proactive in approaching them to support them as well throughout this process.

23:21

Discharge planning. The moment has come for the patient to go home. How can we support the patient and their families going home with a lot of things to do? Providing this comprehensive training for the caregivers on the ongoing care needs is very important, and the people who can provide the support are, of course, the NICU team as well as the pediatric dermatology team.

23:50

A lot of times we will need extra resources and we will need extra support for these families. First can offer a lot of these resources as well. Connecting these families with other families, with kids who have EDDs or with patients who have EDDs, who are now older children or even adults. Helping the caregivers prepare the home for their child is important.

24:19

An example would be having a humidifier at home or having the right crib. Bathing and moisturization - what soaps to use, what moisturizers to use, which one is better? Feeding and nutritional support. What is the best way to feed the baby? How often should we feed the baby? When should we be worried? When should we not be worried? Medication management if any medications are prescribed.

24:46

Surveillance for infection and the ability of the parents to recognize the signs and symptoms of any other comorbidities or clinical changes. Multidisciplinary follow up can feel like a lot. Many of these kids will not only need to follow up with dermatology, which is crucial, of course, but also with ophthalmology, ENT, speech therapy, etc. Prior to discharge, assess the caregivers abilities to manage care effectively at home.

25:24

I have seen a lot of babies with collodion membrane, a lot of newborns with EDDs and I had the privilege to care for these newborns. After a long stay in the NICU and after they got much better and their collodion membrane sheds, it is very important to assess the ability of their family members to take care of them.

25:48
What I typically do is that I go to the room with them and go over all the details, and then I ask them to do the care in front of me 1 or 2 times before they go home, and kind of fine tune the little things and help them throughout the process. Ensure that all the educational tools, clear written materials, return precautions, and the lists of resources are available to the caregivers, and they are reviewed before discharge.

26:22
Typically, all these materials and resources are reviewed with the families by the healthcare providers. Could be by the physician, the dermatologist, particularly the nurses as well. I usually try to provide these written materials in the language of the patients and their families, so that we have a real, written material that they can understand. Sometimes, we also use illustrations. First has a lot of resources, has a lot of articles, a lot of videos, and a lot of experts that could support these families as well as the patients and their families that are a great connection for your patients.

27:13
Finally, ensure that follow up appointments are scheduled with the primary care team, the dermatologist, and other specialists as needed.

27:24
Finally, feel free to reach out to colleagues with expertise in EDDs. There is a physician finder option on First Foundation website where you can find a physician in your region who has more expertise with EDDs You can also reach out by email to all these experts as needed, so that we can support these children wherever they are.

27:54
I hope with this, we were able to review this clinical care in the NICU for newborns with EDDs. Please feel free to reach out to us whenever you need help. Thank you.

28:12