Vulvar Care and Sexuality in Epidermal Differentiation Disorders

CME Series on Epidermal Differentiation Disorders — Presented by Dr. Nessa Aghazadeh

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Welcome to Vulvar Care and Sexuality in Epidermal Differentiation Disorders, part of First's CME series on EDDs or ichthyosis. Presenting this topic is Dr. Nessa Aghazadeh. Dr. Aghazadeh is an assistant professor of dermatology at Mayo Clinic in Rochester, where her practice and research focus on genetic skin diseases, pediatric dermatology and women's dermatology.

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Thank you so much. I am more than happy today to be talking with this wonderful audience about vulvar care and sexuality in epidermal differentiation disorders. I do not have any relevant financial disclosures. So the objective for today's talks is really to recognize the women's health conservation specific to people with EDD or ichthyosis. A lot of the features and phenotypes that relate to epidermal differentiation disorders have been recognized.

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They have been explored, they have been researched. But some aspects that really, truly relate to women's health, particularly vulvar vaginal symptoms and how they affect the sexual and intimacy part of our relationships, have really not been recognized or has not been talked about as much. So really, this is truly a topic that we want to bring forward because, you know, a lot of women with EDD do feel and live these experiences, but they may not necessarily share it with their dermatologists.

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And, you know, sometimes women with EDD they may just see their dermatologist once a year, but they might see their family physician or their women's health doctor or gynecologist a lot more commonly. So that's why we feel it's really, really important for the community of all physicians to come together for this important conversation and discussion to be opened and to be had with our patients.

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I also want to thank all of the women with EDD who have gracefully let us use their pictures and wanted to support this mission. So as you see many of these faces who are absolutely champions in putting this subject forward.

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So as we briefly talked about vulvar disease and ichthyosis is common, and it's really not reported or recognized as much as we would like it to be. Well, when we talk about ichthyosis we talk about a disorder or heritage theory disorder that affects every part of our skin. It is quite easy and quite recognizable for for patients to talk about how their skin feels, or what the symptoms of their skin are on their face, on their arms, on their legs.

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But, you know, it is very uncommon that we hear that conversation, that how the changes of EDD or ichthyosis really affect our vulvar vaginal and anal rectal area. The reality is vulvar skin is also skin, and it can be affected in exactly the same way as any part of our skin can be affected with ichthyosis. So in women with ichthyosis vulvar skin can have scaling, can have fissures.

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It can itch, it can crack. Also the area, anatomically, as we're going to talk about and physiologically is a little bit more thinner and more sensitive than our body parts. So it just really makes sense to look into and search and ask about these symptoms. This is not an uncommon problem in women's health and in people suffering from chronic diseases in general.

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About 23%, about one fourth of people with chronic skin conditions not only are living with that chronic skin condition, but they also report that because of that chronic skin condition, they do have trouble with sexual and intimate relationship. So that is also another important aspect of taking a comprehensive and wholesome care of women and all people really affected by ichthyosis.

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Interestingly, when they survey patients about these symptoms and about these presentations, they really are waiting for their clinicians to open that conversation because it seems like even a simple question, a simple inquisition and curiousity opens the door and allows them to have that conversation and makes it feel like a safe environment to share that experience. So it's really upon us as clinicians to make sure we include that simple question, within our normal visits and part of our physical examinations, as we talked about, this care not only extends from the clinician’s room, but also goes to literature.

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There's almost no published literature that studies the vulvar vaginal involvement in women with EDD so we don't really know what women are suffering, both from a clinical standpoint and from a research standpoint. So I really want to here talk a little bit about why the vulvar skin and the anatomy of the anal genital area is different from the other skin surfaces, and why this makes it

more common for this area to actually be affected with particular symptoms when a person does have ichthyosis.

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So first of all, our bulbous skin is thinner in the stratum corneum, which is the top layer of the skin of the keratinized skin. And it has a weaker barrier. We also have to note that in women, as we have the labia majora and the labia minora, there is a gradual transition from keratinocytes, keratinized skin, which is the area that has the hair, it's the hair bearing labia majora.

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And then as we move to labia minora, the top surface keratin is lost. So we go into the mucosal membrane, which has different characteristics as compared to keratinized skin. The vulvar vaginal area is also very rich in different glands - sebaceous glands, eccrine glands, apocrine glands. This area also has a higher absorption rate of both topicals and, you know, different medications that may be

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applied, over the skin in this are. Anatomically,

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The area is very prone to occlusion by underwear by also moisture and contact with urine and menstrual blood. And also there's a lot of mechanical friction constantly from walking activity, from sports, from intercourse and from clothing. So all of these things together actually just bring that, problem and question forward as to these particular anatomic and physiologic features of the vulvar vaginal skin, how they interplay in women who have EDD and how they actually present these symptoms.

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So as to that question, we have listed a number of common vulvar symptoms and the mechanism that women with ichthyosis could be affected by these symptoms as part of their disease. So as we know ichthyosis really impairs the barrier function of the skin. And because of that, there's increased transepidermal loss of water. The water, natural water or moisture in this thing cannot be retained.

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That causes the skin in the vulvar area as well as other places in the in the body, to become scaly and dry fissures which are deep and thin cracks in the skin or mucosal membranes can happen, particularly in the perineal area or around the opening of the vaginal introitus. They can cause quite a bit of pain, and sometimes there is secondary bacterial infection or cornification over those fissures, which really exacerbates the problem.

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Itching is a very common symptom in the vulvar area

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in women with EDD which actually happens because of a variety of reasons, including dryness, added inflammation because of the constant activation of the inflammatory pathways, and also because of nerve irritation and finally redness or visible erythema in the area can also happen because of the constant inflammation.

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Odor can be a very disturbing problem in some women with ichthyosis. It is really important to know why this problem happens, because it is truly because of all the trapped scale that cannot normally shed itself because of the hereditary EDD or ichthyosis and the trapped scale actually bring the trapping of moisture and bacteria that cause malodor, it is a really, really important point to emphasize with women and patients with ichthyosis that it is not due to bad hygiene, it is not something they did to cause the odor.

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But it can be a problem that people are very aware of and very self-conscious about, and quite bothersome for people who are affected by it. Recurrent infections can also be a problem whether these are on the skin surface, such as bacterial infections or more related to the vulvar vaginal area, such as yeast infections, bacterial vaginosis. This is mainly caused because of the problem in the barrier of the skin and natural antimicrobial pathways that are often compromised in people with ichthyosis.

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There are also some different subtypes of ichthyosis that have particular immune dysfunction that may predispose infections even more than other subtypes, and that is always important to note that these symptoms really, really vary across different subtypes, genetic subtypes of ichthyosis, and also from one woman to another.

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Dyspareunia is another common problem in the disorders of vulvar vaginal area, which is actually defined by pain with penetrative intercourse or penetrative sex. And this also can have multiple reasons.

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As we talked about, fissures in the opening of the vagina can be a cause. Erosions or little actually surface blisters or opening of the mucosal membrane or even the skin can cause pain within intercourse. And finally dryness and reduced lubrication can also cause pain with

penetrative intercourse. Vulvodynia is a chronic pain that affects the vulvar area, whereas you don't really see any contributing skin lesion.

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So in the anatomic and and physical examination part, everything looks fine. But the women who are affected by vulvodynia actually report constant pain. Sometimes the pain is really constant throughout. But things like sitting, actually, wearing tight underwear or clothing really make the pain worse. Unfortunately, vulvodynia happens as a result of many different disorders that affect the vulvar vaginal area.

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But essentially when there is a trigger in the vulvar vaginal area and the nerves are constantly irritated, this can cause a residual level of neuropathic pain that can persist even though the original trigger or the original lesion has resolved, and that can be more of a chronic pain situation for some women who are dealing with EDD and ichthyosis. All right, as we talked about, not all of these symptoms happen in all women with ichthyosis. Of course,

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One, not all, but one of the distinguishing features of the statement of different types of EDDs are the genetic subtype. So based on different genetic, you know, particular features that could affect the vulvar vaginal areas are the ones that we will be discussing here. So women who are affected with epidermolytic ichthyosis generally caused by keratin 1 and keratin 10 variants typically have more of a loss of the mechanical strength of the skin.

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And these are women who are more prone to having blisters, erosions, and ulcers in the areas with friction and areas with mechanical trauma. So we might have open wounds, healing wounds, and also with time, specially trapped hyperkeratotic skin which can cause the malodor and super infection along with maceration. Women with Netherton syndrome, which goes by SPINK5 variants, actually do have severe barrier dysfunction and also might have different degrees of the immune dysregulation.

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These are women who oftentimes have more severe itching, which can of course affect the vulvar vaginal area. And also they might be at increased risk of different types of infections in the vulvar vaginal area, including, for example, genital warts. People with more spectrum of lamellar ichthyosis NCIE might have more issues with redness, inflammation and also depending on their clinical phenotype and severity, they might have built up of platelet scale.

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Again, it is important to note that the features in the vulvar vaginal area may or may not exactly reflect the involvement of skin in other areas, for example, in the vulvar vaginal area, as we discussed, because there is more occlusion and there's more moisture, sometimes the build up of scale actually can be less in these fold and wrinkles of surfaces as compared to other places, but that is again, not constant among all the women who are affected by these diseases.

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Okay. So what are the practical, guidance and practical counseling that we can give our patients once we see women with ichthyosis especially in primary care setting, in women's health setting as family physicians and also as primary care doctors, how can we support these women? How can we provide counseling? As we talked about, I think one of the most important things is to ask directly.

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So like we talked about, this is an uncomfortable conversation to have for most patients. And it turns out for most physicians too and that's something that we need to internalize and actually accept but then overcome. So most patients will not spontaneously report their vulvar symptoms to you unless they are specifically asked. And that is what the patients really want us as clinicians to do during our visits.

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So a single screening question, even if it is built in as part of our review of system, when we see people with EDD, would be sufficient. And that could be something like do you have itching, do you have cracking, or do you have any discomfort in your genital area? That really opens up the conversation and as we talked about, really turns the visit and the clinical environment into a safe space to discuss if there are ongoing or undiscovered issues that women with EDD are suffering from with regards to vulvar vaginal or even sexual dysfunction.

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So back to the very practical advice that we can share with our patients. So regardless of what the symptoms are and the vulvar vaginal areas, there are simple tips and there are simple care items that really could help or aggravate the symptoms and vulvar vaginal area. So these are important to share with our patients. In general, anything that has scents and fragrances is not for the vulvar vaginal area, soaps, bubble baths, fragrance washes.

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they do not belong in the vulvar care. Right? Vaginal douches are really not helpful. They really actually disturb the normal microbiome of the vulvar vaginal area. The women's vulvar vaginal area has a perfect natural way of keeping its microbiome healthy and, vaginal douche and

things like that, feminine washes actually do disturb the natural composition of the bacterial microbes in that area that really needs to be maintained.

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So we want to avoid over washing and over soaps. So tight, or unbreathable or synthetic underwear, they really make every type of vulvar vaginal symptom worse. There are days that are going to be difficult, and if there are any symptoms or underlying conditions, some days that are really hot, sweaty, lots of friction and activity involved, they might flare up some of the symptoms.

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For women, an important consideration with regards to ichthyosis and the skin care or keratolytics. So we use keratolytics, which are chemical products that we are commonly applied to most skin surfaces. In people with ichthyosis to help with the gentle exfoliation of the thick skin. Now, while keratolytics are really great addition to the general skin care of people with EDD or ichthyosis, we have to use caution when we are, trying to actually incorporating as part of vulvar vaginal care.

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Most of the keratolytic topical products are actually too strong for the sensitive and delicate skin of the vulvar vaginal area, so it is always good to have a consultation or a chat with a dermatologist to see first of all, are they even needed in the vulvar area? And if they are needed, what would be the appropriate type? What would be the appropriate concentration?

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And if it needs to really be used in case of fixed scale buildup, we'd like to really limit it to that keratinized surface, which is mainly the labia majora, and try to avoid the mucosal membranes, which really do not need any keratolytics.

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All right. Again. Now what are the things that we can do to help the health of the vulvar vaginal area? The more water for cleansing the better. So like I said, we like to avoid over washing and over soaps and cleansers in this area generally really soaps are not needed in the vulvar vaginal area for women who have significant, discharge or a significant build up or odor, they may want to use a gentle cleanser. If they would like to use a soap,

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we recommend a very, very gentle cleanser. White, typically, and no scents at all. We also recommend patting very gently, no rubbing of the area.

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So applying petrolatum is one of the most effective, emollient and barriers that we have.

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It is completely safe to use in the vulvar vaginal area. It is very inert. It really doesn't cause any stinging or burning. And it really helps, like I said, not only with retaining the moisture in the area, but also helps as a barrier because it is a repellent to water and moisture. We also recommend 100% cotton underwear. A lot of women actually do prefer to go without different parts of the day or night, and that is completely fine.

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As we said, letting the area breathe with no occlusion actually does help, especially in situations where there is maceration and build up of scale and odor. Again, we mentioned that vaginal douches are not really healthy for the vulvar vaginal care, and one other important aspect of care is for women who have open sores or erosions or ulceration affecting the vulvar skin, or the mucosa.

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Urination can sometimes cause stinging and burning pain because urine is acidic, and when it actually touches the vulvar skin and mucosa, it can cause burning pain. So a couple of tips to address that situation is actually one - hydration. So drinking a lot of water really dilutes the urine and decreases that pH or actually helps ameliorate pH. Second is applying a layer of plain petrolatum right before urination.

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So it acts as a barrier. And the urine, which is water based, does not directly touch the vulvar vaginal surface. And then third, using water to rinse any residual urine to make sure that it does not continue to get exposed to the open erosions or ulcers.

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Another important aspect of addressing itch. I'm talking about pruritus and itch with patient is understanding that each scratch cycle. So each is one of the most common symptoms in general in dermatology and also in particular in vulvar vaginal area, but also in women, in people with ichthyosis, about 80% of patients with different types of EDDs actually report itch, which is a significant factor in quality of life.

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And it really, really affects the day to day functions and aspect of living with ichthyosis, So one of the very complex and curious things about age is that, you know, oftentimes with any itchy dermatosis or affliction of the skin, we have this continuous itch and scratch cycle. So obviously itching is happening in the first place because of the hereditary lack of particular functions,

particular structures of the skin, which cause compensatory activation of inflammatory pathways in the skin, which make the skin become itchy.

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Now the natural response as the itch is transferred via the C fiber nerves from the skin to our central nervous system is to scratch it. So the physical act of scratching, when it happens, it actually causes more skin damage. So whenever there is skin damage, there is again activation of inflammatory pathways towards skin healing. But this inadvertently also activates all of the other inflammatory pathways of the skin, which are already on high alert in a person with ichthyosis.

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So that causes even more itch to happen. And then the more we scratch, the more itching happens. Like I said, this is really called the itch scratch cycle. And understanding this, you know, feedback loop is really important. So we can help support our patient with this particular symptoms. So of course we want to break the cycle.

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And we want to start with the symptom of itching. Right. So obviously first of all avoiding any of the trigger that makes the itch worse in the vulvar vaginal area. These are often heat, sweating, occlusion, you know, tight underwear, all the stuff that we talked about already, also getting help from a dermatologist in the setting of EDD is really important and helpful because now we truly have lots of medications, biologic medications and otherwise, which really can help with that symptom of itching in ichthyosis.

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And then also it's really important to share with the patients about this itch-scratch cycle. So they're aware and they have that, you know, self-awareness of how scratching is really worse is part of the problem when people are experiencing itching. And even that self-awareness, I think, is really helpful in regulation of the scratching behavior that becomes particularly important in the vulvar vaginal area.

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As, like I said, it's one of the most common in general symptoms of women in vulvar vaginal area. All right. Another general aspect of taking care of women with EDD is emphasizing screening and also vaccination. Of course, these are all part of the general health of women, but in particular in women with ichthyosis like we talked about because of the impaired skin barrier and sometimes the natural, immunomodulatory mechanisms of the skin that are in impaired, there might be increased risk of viral infections, particularly warts in the anal genital area.

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This is even more of a risk in some subtypes of ichthyosis, such as Nethertons disease where the immune function is impaired. As such, all women in general, and particularly those with ichthyosis need to be counseled on the importance of getting HPV vaccination. And even if they are a little bit outside of that age period, ages 27 to 45, we still really strongly recommend having that conversation and encourage taking the vaccine, because the benefits definitely would outweigh the risks in the situation where the skin barriers are already impaired and there's increased risk of all types of infections, including acquisition of viral warts.

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Again, another important is continuing to have routine cervical cancer screening, as recommended by the national screening on their schedule. So we also would advocate, that at least external genital examination is offered to women with ichthyosis when they see their primary care. Visualization of the vulvar area directly really does help with understanding the problem, really seeing what is going on, the degree of erythema, scaling, fissuring, or if there are any additional bacterial yeast or viral infections.

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Sometimes it might be difficult because of the changes of ichthyosis to identify some of the lesions in the area, or for example, to know if a particular hyperkeratotic or thick lesion is a wart, is it pre-cancer, is it, you know, a dysplasia? And in those situations, I think consultation with a dermatologist would be really helpful. And at times considering biopsy to have a diagnostic confirmation.

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And then, as we talked about vulvar care main practices, I really want to emphasize for this slide some of the practices of, you know, how to improve the quality of sexual and intimate life for women with ichthyosis. Like I said, if women with ichthyosis choose to have an intimate and sexual relationship, we should be supportive of that as part of a whole health and care and try to very best to improve that very important, physiologic and psychosocial part of a person's health.

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So with regards to intimacy and intercourse, a lot of, you know, care we already discussed does apply. So making sure the vulvar vaginal area is clean, is well moistured. A petrolatum based barrier cream does really help both prior and also after engaging in intimate or sexual intercourse. Using lubricants also does help because of natural dryness that might happen in the area.

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Preservative free silicon based lubricants are likely the best in this area because again, they have the lowest risk of causing any irritation or any, unpleasant feeling for the already inflamed vulvar vaginal surface. We would like to avoid any of the lubricants or other intimacy products that could have a base of fragrance, alcohol containing product, those that might have propylene glycol.

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Those may actually burn or cause irritation in the area.

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And lastly, I am very happy to say that in coordination and collaboration with First and truly, with the support and help of women with ichthyosis, we are currently doing a study and probably, you know, a survey, study, which is a first of its kind, to truly look into what are the extent of vulvar vaginal symptoms in women with EDD and also study what are the sexual function, what is the sexual function like in women who have ichthyosis.

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So like I said, I am more than grateful for women who have already participated in our surveys. And we are looking for more and more engagement because we truly think this is an area that has been there is a silence that has been going on for longer than it's needed, and we need women's voices. We need to hear women's voices.

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We need to have a conversation, start a conversation about these topics that are understandably difficult, difficult for everyone to talk about. But unless we talk about it. We're not going to find solutions. So thank you again. We welcome referrals for your patients or any woman with ichthyosis who is interested in sharing their experience or lived experience with us.

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We really would like to close this gap in research and in evidence, and bring the voice of women into literature. And the links to the surveys can be found on the First website. And again, we are more than happy and thankful for their collaboration. Thank you so much for listening to this talk, and thank you so much for First Skin Foundation for making it possible.

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