Adult Consent to Take Part in a Research Study

Name of Study: An Open-Label and Long-Term Extension Study to Evaluate the Efficacy and Safety of Ustekinumab in the Treatment of Patients with Ichthyoses.
Drug and Funding Support Provided by: Janssen Scientific Affairs, LLC (Company)
Name of Researcher (referred to as the study doctor): Amy S. Paller, M.D.

This consent form describes a research study for which you might qualify at Ann & Robert H. Lurie Children's Hospital of Chicago ("Lurie Children's") and NU/NMH. Research studies help us learn more about conditions and develop new treatments. Taking part in a research study is voluntary. It is your choice to take part in this research study. Please read this consent form and ask questions about anything you do not understand. You may talk to others such as your family or healthcare providers before you decide to take part in this study. The study staff will also explain the study to you and answer any questions that you may have. Your decision will not affect your regular care.

Key Study Information:

What are the purpose and goals of this study?
You are being asked for your consent to take part in this study because you have been diagnosed with ichthyosis (severe and lasting problems with dry skin). This research is being done to look at safety and how well ustekinumab works in the treatment of individuals with ichthyosis. Ustekinumab is an investigational drug. "Investigational" means this drug has not been approved by the United States Food and Drug Administration (FDA) for treatment of ichthyosis.

Ichthyosis is a lifelong genetic disorder that may cause thickening, scaling and inflammation (redness, heat, swelling and pain) of the skin. Ichthyosis is commonly associated with impaired quality of life, social exclusion due to an altered appearance, itchiness, discomfort, and functional limitations. Therapy for ichthyosis is supportive, focused on clearing skin scales, and is often time-consuming for patients. Currently, there are no standard therapies based on what causes ichthyosis.

The investigational drug used in this study is called ustekinumab. Ustekinumab is a monoclonal antibody. An antibody is a type of protein that helps protect the body against foreign materials such as germs. A monoclonal antibody is a laboratory-made substance that recognizes a specific region of a protein. Ustekinumab regulates proteins in the body called IL-12 and IL-23. Abnormal regulation of IL-12 and IL-23 has been associated with a variety of autoimmune diseases (when your body attacks normal cells) including psoriasis. Recent studies have shown that a specific protein called interleukin-23 (IL-23) is increased in the skin of ichthyosis. IL-23 is known to be increased in another skin disorder with redness and scaling called psoriasis. Reducing IL-23 levels in skin with ustekinumab has been found to markedly improve psoriasis.

This study seeks to understand if ustekinumab works better than standard treatment for ichthyosis. It also seeks to learn about the safety of ustekinumab in those with ichthyosis.

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Stamped by: EJK

We expect about 15-20 people here at Lurie Children's Hospital/Northwestern University will be in this research study.

What are the main procedures of this study?
We expect that you will be in this research study for about 2 years (25 months). There will be a screening period, an initial 13 month open-label (OL) phase in which you will receive ustekinumab, followed by a 12 month long-term extension (LTE) to assure long-term safety during which you will continue to receive ustekinumab. You will have approximately 15 visits to the study site.

Each subject will receive ustekinumab at Baseline (Week 0) and Month 1 (Week 4), Month 3 (Week 12), Month 5 (Week 20), Month 7 (Week 28), Month 9 (Week 36), and Month 11 (Week 44). During the LTE, subjects will receive injections every 8 weeks for one year: Month 13 (Week 52), Month 15 (Week 60), Month 17 (Week 68), Month 19 (Week 76), Month 21 (Week 84), and Month 23 (Week 92). Subjects will come back in for a follow-up visit at Month 25 (Week 96) for an end of study visit (no drug administration).

What are the important risks and side effects from this study?
The most common (≥ 3%) harmful reactions associated with ustekinumab in patients being treated for psoriasis were nasopharyngitis swelling of the nasal passages and the back of the throat), upper respiratory tract infection, headache, and fatigue.

What are the benefits from this study?
Your ichthyosis may improve from taking part in this study. The information learned from this study may also help others in the future with ichthyosis.

Details of Taking Part in this Study:
If I agree to take part in this study, what would I need to do?
You will be asked to come to these study sites for the study visits. Any visits that do not require an injection of the study drug may occur in the Northbrook location per your request. All study visits requiring an injection will need to occur at the downtown location.

- Ann & Robert H. Lurie Children's Hospital of Chicago, 225 E. Chicago Ave, Chicago, IL 60611
  - Outpatient Clinic, 3rd Floor; or
  - Clinical Research Unit (CRU), 19th Floor
- Ann & Robert H. Lurie Children's Hospital of Chicago Outpatient Center in Northbrook, 1131 Techny Rd., Northbrook, IL 60062

The below tables outline the study procedures that will be conducted at each visit.

	Screening	Baseline	Month 1	Month 3	Month 5	Month 7	Month 9	Month 11	Month 13
	Week -4	Week 0	Week 4	Week 12	Week 20	Week 28	Week 36	Week 44	Week 52/ OL end
Procedures	**Visit 1**	**Visit 2**	**Visit 3**	**Visit 4**	**Visit 5**	**Visit 6**	**Visit 7**	**Visit 8**	**Visit 9**
Consent	X								
Demographics	X								
Inclusion/Exclusion	X	X							
Medical History	X	X	X	X	X	X	X	X	X
Concomitant Meds	X	X	X	X	X	X	X	X	X
Adverse Event Assessment		X	X	X	X	X	X	X	X
Full complete physical exam, including skin exam	X	X							X
Full skin exam			X	X	X	X	X	X	
Vital Signs (incl. weight)	X	X	X	X	X	X	X	X	X
Height	X								X
Severity evaluations	X	X	X	X	X	X	X	X	X
Venipuncture	X								X
Saliva collection	X								
Blood collection	X								X
Tape Strip collection		X				X			X
Microbiome collection		X				X			X
Skin water loss measurements		X	X	X	X	X	X	X	X
Pain and Itch scores		X	X	X	X	X	X	X	X
PROMIS Itch (if ≥2 on NRS, otherwise subject does not need to complete)		X				X			X
Patient reported outcomes		X				X			X
Subject severity self-assessment		X	X	X	X	X	X	X	X
Photography		X				X			X
QuantiFERON®-TB gold	X								X
Hematology and Chemistry testing	X								X
Hep B/C, HIV testing	X								
Urine HCG (if applicable)	X	X	X	X	X	X	X	X	X
Injection		X	X	X	X	X	X	X	

	Month 13	Month 15	Month 17	Month 19	Month 21	Month 23	Month 25
	Week 52/LTE Start**	Week 60	Week 68	Week 76	Week 84	Week 92	Week 96/ End of LTE
Procedures	Visit 9	Visit 10	Visit 11	Visit 12	Visit 13	Visit 14	Visit 15
Consent							
Demographics							
Inclusion/Exclusion							
Medical History	X	X	X	X	X	X	X
Concomitant Meds	X	X	X	X	X	X	X
Adverse Event Assessment	X	X	X	X	X	X	X
Full complete physical exam, including skin	X						X
Full skin exam		X	X	X	X	X	
Vital Signs (incl. weight)	X	X	X	X	X	X	X
Height	X						X
Severity evaluations	X	X	X	X	X	X	X
Venipuncture	X			X			X
Saliva collection							
Blood collection	X						X
Tape Strip collection	X			X			X
Microbiome collection	X			X			X
Skin water loss measurements	X	X	X	X	X	X	X
Pain and Itch Scores	X	X	X	X	X	X	X
PROMIS Itch (if ≥2 on NRS, otherwise subject does not need to complete)	X			X			X
Patient reported outcomes	X			X			X
Subject severity self-assessment	X			X			X
Photography	X						X
QuantiFERON®-TB gold	X						X
Hematology and Chemistry testing	X						X
Hep B/C, HIV testing							
Urine HCG(if applicable)	X	X	X	X	X	X	
Injection	X	X	X	X	X	X	

Screening/Baseline Visit

The Screening and Baseline visit procedures may be combined on the same day if you meet all screening eligibility criteria. If you have not had genetic testing (research or a CLIA-approved lab), you will also be asked to enroll in Dr. Paller's "Screening for the Genetic Basis of Dermatologic Disease" IRB# 2009-13803 and have testing sent to Dr. Keith Choate at Yale University. If the clinical diagnosis is ichthyosis / ichthyotic disorder, having a known gene mutation is not required before randomization and entry into the study, however it is required that genetic testing is performed before end of study.

Urine pregnancy test (subjects of childbearing potential only) will be completed prior to any baseline study procedures and ustekinumab injection.

Medical and Medication History

The study doctor or designated study team member will collect information regarding your relevant medical history/current medical conditions at every study visit as indicated in the above tables. All collected information will be documented. When possible, diagnoses (rather than symptoms) will be recorded. Your history regarding your ichthyosis will also be obtained including date of diagnosis and prior treatments. Medications including current medications and those taken within six months preceding enrollment will be recorded.

While enrolled in the study, you may not use any topical agent for the treatment of ichthyosis during the first 6 months other than bland emollients. After the 6-month time point, the following keratolytics, including but not limited to agents containing tazarotene, salicylic acid, alpha-hydroxyacids, and urea, can be added to the regimen as per your study doctor's discretion.

Vital Signs and Height/Weight

Vital signs, including blood pressure, pulse, respiratory rate, temperature, and weight will be collected at every visit. Height will be recorded at the first visit, the end of the open label phase (OL) and the end of the long-term extension (LTE).

Complete Physical Examination & Skin Examination

A complete physical examination will be performed by a physician at the screening and baseline visits, as well as the end of the open label phase and the end of the long-term extension. This will involve an evaluation of the major body systems. A full skin exam (which will involve evaluation only of the skin) will occur at baseline and then at every visit at which there is study drug administration, as indicated in the above tables.

Safety Labs and Pregnancy Testing

Clinical lab evaluations (blood tests) at screening visit include hematology and chemistry panels, QuantiFERON gold, Hepatitis B, Hepatitis C and HIV testing to be resulted at Lurie Children's Hospital. Approximately 15 ml, about 3 teaspoons, of blood will be drawn by a licensed phlebotomist per hospital requirements. Hematology/chemistry panels and QuantiFERON gold will also be drawn at visits for Months 13 and 25.

If you are female and of child-bearing potential (i.e. female subjects who have experienced their first menstrual period) you will be tested for pregnancy at the screening and baseline visit. Urine HCG testing will be obtained. Anyone with a confirmed positive pregnancy test during screening is not eligible for participation in this study. In addition, if you are a female of child-bearing potential, you will have urine HCG testing prior to every administration of the study drug.

Severity Evaluations - IIS and Ichthyosis Severity Self-Assessment

Physician severity evaluations will be performed at every study visit. A severity assessment will be collected which will include the Ichthyosis Severity Index (ISS). In addition, you will be asked to complete a severity self-assessment at every visit. These are surveys.

TEWL and Tape Strips

Transepidermal Water Loss (TEWL) skin measurements will be performed on the nondominant arm to assess the skin barrier at all study visits. TEWL measurements address the level of moisture that is lost from the skin (how dry the skin is). This procedure involves placing a small measurement device on your skin for a few seconds to get a reading. The measurements will be repeated up to four times, with each time having the device read measurements in triplicate.

Tape strips will be also be obtained from your skin at Baseline, Month 7, Month 13, Month 19, and Month 25. Tape stripping is painless and involves the application and removal of small circular tape discs to your skin. Tape stripping will be performed on your upper arm using circular 2x2 or 3x3 cm transparent tape strips. Up to 16 tape strips will be serially extracted on each skin site. You will be asked not to apply emollient or moisturizer for at least 24 hours prior to tape stripping. The tape strips will be stored on site at the Paller Laboratory at Northwestern University. Tape strips will be used for biomarker analysis.

Microbiome Sampling

Microbiome sampling will be done at baseline, Month 7, Month 13, Month 19, and Month 25. The microbiome is the collection of organisms living in and on our bodies. This sampling will involve swabbing your skin with three separate swabs in three different areas on your nondominant upper outer arm. You will be asked not to shower or apply emollient or moisturizer for at least 24 hours prior to sampling.

Photography

Full body photography (including face) will be done at Baseline, Month 7, Month 13, Month 19, and Month 25. Photos may also be taken at additional visits if your ichthyosis severity changes outside of these mentioned time points. Photos will be labeled using your study ID as well as your visit number and date of the photo. Photos will be uploaded to a secure, research server maintained by Northwestern University within the Department of Dermatology and deleted from the camera memory card as soon as possible.

Blood Samples for Biomarkers

A 15.0mL, about 3 teaspoons, blood sample will be collected at Screening, Month 13 and Month 25. Blood will be drawn by a licensed phlebotomist per hospital requirements and numbing solution may be applied to your skin prior to the blood draw to reduce pain. Your samples will

be labeled with your study ID, study visit number, and visit date and stored at Northwestern University. Samples will be banked at Northwestern University for future analysis.

Patient Reported Outcomes

You will be asked to complete several patient reported quality of life measures that will take approximately 15-20 minutes to complete. These will include the following surveys:

-Dermatology Life Quality Index (DLQI) OR Children's Dermatology Life Quality Index (CDLQI)

-Ichthyosis Financial and Time Burden (IFTB)

-Stigma questionnaire

Collected at Month 0, Month 7, Month 13, Month 19, and Month 25

-Itch and Pain Numerical Rating Scale

Collected at all visits

-PROMIS Itch

PROMIS Itch survey will be conducted at Month 0, Month 7, Month 13, Month 19, and Month 25 if the subject scores ≥ 2 on the Itch NRS.

Open Label and Long Term Extension Rollover

You will have visits at Screening, Baseline, and Months 1, 3, 5, 7, 9, 11 and 13 for the open label phase (OL). Should you and the investigator find ustekinumab to be beneficial, you may have the option to continue to the Long-Term Extension (LTE) with in-person study visits every 8 weeks (Months 15, 17, 19, 21, 23 and 25). The final in-person study visit will be at Month 25, with the final administration of study drug at Month 23.

What are the risks, side effects, or discomforts related to the study?

You may have some side effects and discomfort while in this research. If you have any side effects, you should tell the study doctor or staff as soon as possible. They will monitor you closely. This research may also have risks or side effects that are not well known or understood at this time.

Risks related to ustekinumab:

The possible discomforts, side effects and risks related to ustekinumab treatment are not all known. Most side effects are not serious. Some may be serious and may require treatment or additional testing. This section describes how frequently side effects occurred in subjects who were treated with ustekinumab. In this section, the following terms are used:

- Very common: affects more than 1 user in 10
- Common: affects 1 to10 users in 100
- Uncommon: affects 1 to 10 users in 1,000
- Rare: affects 1 to 10 users in 10,000
- Very rare: affects less than 1 in 10,000

Very Common:

- None

Common:

- Infection of the throat or airways or sinus
- Sore throat
- Feeling tired
- Redness and pain at drug injection site
- Back, joint or muscle pain
- Vomiting
- Headache
- Dizziness
- Diarrhea
- Nausea
- Itchiness

Uncommon:

- Swelling, itching, hardness, bleeding, bruising and irritation where the injection is given.
- Shingles (a painful rash)
- Depression
- Inflammation of tissue under the skin. Signs include warmth, swelling, redness and pain
- Nasal congestion
- A form of psoriasis with raised bumps on the skin that are filled with pus
- Allergic reactions including rash or raised, itchy bumps
- Tooth infections
- Acne
- Feeling weak
- Vaginal yeast infection
- Chest infection

Rare:

- Serious allergic reactions, which could be life-threatening (including low blood pressure, trouble breathing, swollen face, lips, mouth and/or throat)
- A form of psoriasis with redness and scaling of a much larger area of your skin or your entire body (erythrodermic psoriasis)
- In rare cases, symptoms such as cough, shortness of breath, and fever may also be a sign of an allergic lung reaction to ustekinumab
- Inflammation of small blood vessels, which can lead to a skin rash with small red or purple bumps, fever, or joint pain (vasculitis)

Infections
Ustekinumab is a drug that may change how your body fights infections. Serious infections requiring hospitalization for medical observation and /or treatment have been seen in ustekinumab studies. Some of these infections have also been life threatening.

Tell your study doctor if you have a new infection, if an infection keeps coming back, or if you have any signs of infection such as:

- fever
- chills
- headache
- coughing
- congestion
- chest tightness
- shortness of breath
- flu-like symptoms
- nausea
- vomiting
- diarrhea
- increased frequency or burning while passing urine
- redness warmth, tenderness orswelling of skin or joint
- cold sores
- new or worsening of pain in any location
- weight loss
- tiredness
- night sweats

It is unknown if ustekinumab may stop you from developing a fever if you do have an infection, and therefore hide that you have one.

Fungal infections have been reported in subjects taking ustekinumab. Some of these fungal infections can be serious and involve internal organs. You should find out from your study doctor which fungal infections are common where you live or travel and what symptoms they cause. Tell your study doctor and family physician right away if you develop symptoms of such illnesses.

Subjects who receive ustekinumab may also be at a greater risk for certain serious infections such as tuberculosis. Tell your study doctor if you have ever had tuberculosis or anybody in your family has ever had tuberculosis or if you come in contact with someone who has tuberculosis. Tell your study doctor if you develop:

- a cough that does not go away
- coughing up blood
- shortness of breath
- fever
- night sweats
- weight loss

Cancer
Cancers have been reported in subjects who have received ustekinumab but it is unknown whether taking ustekinumab has increased their risk for developing cancer. Because ustekinumab may suppress your immune system, it is possible that it may increase your risk of developing cancer, including skin cancers. Subjects who have been diagnosed with psoriasis have a higher

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chance of developing skin cancers. Tell your doctor if you have any new or changing skin lesions.

It is known that people who have had inflammatory diseases (such as, Crohn's disease, Rheumatoid Arthritis, Ulcerative Colitis etc.) for a long time and who use immunosuppressive therapies (such as, azathioprine, methotrexate etc.) for a long time have a higher risk of developing cancer. These people get cancer of the lymph nodes more often than other people.

Infusion Reactions, Injection Site Reactions and Allergic Reactions
Ustekinumab may cause an allergic reaction in some subjects. These reactions are usually mild to moderate. The following can be symptoms of an allergic reaction:

- fever
- chills
- hives
- rash
- swelling
- itching
- headache
- flushing
- nausea
- light-headedness
- chest pain or tightness
- wheezing
- difficulty breathing or swallowing
- decrease or increase in blood pressure
- anaphylaxis (life threatening allergic reaction)

Serious allergic reactions have been reported in subjects taking ustekinumab and can be life threatening. If this happens during the infusion, the infusion will be stopped. Signs of a serious allergic reaction include skin rash, swollen face, mouth, lips, and/or throat, and trouble breathing. Tell your doctor or get emergency medical help right away if you have an allergic reaction. If you experience a serious reaction to an injection or an infusion, you will not receive any more study treatments.

If you have an infusion reaction or an allergic reaction at the doctor's office, additional necessary treatment will be provided immediately. Your study doctor may give you an antihistamine (medication used to treat allergic symptoms such as hay fever) or other medications used for treating an allergy. Antihistamines can make you sleepy, so please use caution when driving a car or operating machinery.

Another type of allergic reaction has occurred in some subjects 1-14 days after receiving some similar medications. The symptoms of this type of allergic reaction may include fever, rash, muscle aches and joint pain.

Antibodies to ustekinumab
Sometimes the body can make special antibodies that may increase the risk of an allergic reaction to either ustekinumab or other antibody medicines. If you have an allergic reaction, you may not be able to have these types of medications in the future. You should always tell your doctors that you have been treated with human antibodies in this study.

Latex allergy
The needle cover for the prefilled syringe that contains study drug contains dry natural rubber (a form of latex). This may cause allergic reactions in people who are sensitive to latex. Please tell your study doctor if you have ever had an allergic reaction to latex.

Cardiac and Vascular
Heart attacks and strokes have been reported in subjects who have received ustekinumab. These events have rarely resulted in death. It is unknown whether taking ustekinumab increases your risk for developing these events.

People who have psoriasis, and certain other inflammatory diseases, have a higher risk of having heart attacks. These people have heart attacks more often than other people. Seek medical care immediately if you develop;

- chest pain or discomfort
- trouble breathing
- irregular heartbeats
- dizziness
- loss of balance
- new numbness or weakness
- visual or speech changes

Vaccination
Vaccines are made to help protect people from certain illnesses. Some vaccines are made from live bacteria or live viruses. You cannot receive most kinds of live vaccines (for example, FluMist™, varicella/chicken pox) during the study or for 3 months after the last study injection. Another kind of live vaccine is BCG, which is a vaccine against tuberculosis. You cannot receive a BCG vaccine during this study or for 16 weeks after the last study injection. You could get sick from these kinds of vaccines while on ustekinumab. If you do get a live vaccination during this study, you must tell your study doctor immediately.

Tell your study doctor if anyone living in your home needs a live vaccine. Some viruses used in live vaccines can spread from a close contact (someone living in your home) to people with a weakened immune system.

Other kinds of vaccines, like tetanus,flu shots, and COVID vaccines that are not live are allowed. It is not known if ustekinumab may interfere with them from working. Tell your study doctor before getting any vaccine while you are in this study.

Other Therapies
Tell your doctor if you are receiving treatments that weaken the immune system while using ustekinumab (for example, oral steroid medicines). These treatment combinations have not each been studied with ustekinumab, so it is unknown if they could possibly increase the risk of diseases related to a weakened immune system.

Allergy Immunotherapy (Allergy Injections)
Tell your study doctor if you have ever had or are now getting allergy injections. Ustekinumab may affect your response to allergy injections.

Other Risks

Two cases of a very rare disease of the brain, posterior reversible encephalopathy syndrome (PRES), also known as Reversible Posterior Leukoencephalopathy Syndrome (RPLS), were reported in clinical studies with Ustekinumab. PRES is generally reversible and is not caused by an infection. It is unknown whether taking Ustekinumab increases your risk of developing PRES/RPLS.

Symptoms of this condition are:

- headache
- seizures
- confusion
- loss of eyesight

Tell your study doctor if you experience any of these symptoms. There may be other discomforts or risks to you from this study that are not yet known. Your study doctor and staff will ask you about any side affects you may have at every visit. If you have any problems, you should let your study doctor know right away.

Tell your doctors or dentist that you are or have been in a study where anti IL-12 and anti-IL-23 is the study drug. This is important if you have any surgery, dental procedures, or receive treatment for any other medical condition.

Risks related to study procedures and tests:

Blood Draws Risks
You may have discomfort or pain when blood is collected. You may feel faint or pass out. There is a risk of infection, bleeding, or bruising at the puncture site. You may develop a small scar at the puncture site where several blood samples are taken.

Confidentiality:
There is a risk of loss of confidentiality. However, steps will be taken to maintain confidentiality. Only authorized study staff will have access to your information.

Risks related to pregnancy:
The effects of the study drug on the reproductive system (sperm, eggs) or to an unborn baby are unknown. It is important that when in this study, participants do not become or make their partner pregnant. Those who are pregnant or nursing will not be allowed to take part in this study. Everyone in this study must agree to use a reliable method of birth control or not have sexual intercourse during the study. The study doctor/staff will talk about this with you and provide information on the best methods of birth control.

Pregnancy testing may be required at some visits while you are in this study. During the course of the study, if there is any chance you may be pregnant (late or missed menstrual period), please contact the study doctor right away. If you become pregnant when taking part in this study, you may be asked to stop the study. However, follow-up visits to review the effects on you and/or the baby may be needed.

It's also important that you tell your baby's doctor and other health care professionals that you received treatment with Ustekinumab while pregnant, because a 6-month waiting period following birth may be recommended before the administration of a live vaccine (like BCG and rotavirus) to the baby.

Risks related to genetic testing:
Genetic information is like a fingerprint and it is unique for each person. Many of the risks linked with genetic testing include the loss of confidentiality and privacy. There is a risk that

someone could trace the genetic data back to you. This could happen even if the genetic data is stored without your name or other information that can identify you. This risk may increase in the future as people come up with new ways of tracing information.

Many people are worried that the results of genetic testing might be held against them. There are federal and state laws to protect people. The federal law is the Genetic Information Nondiscrimination Act (GINA). The state law is the Genetic Information Privacy Act (GIPA). GINA and GIPA generally make it unlawful for health insurance companies, group health plans, and employers to discriminate based on genetic information. This law does not protect against discrimination by companies that sell life, disability, or long-term care insurance. Ask the study doctor for more information on these laws.

What other options do I have?
You do not have to participate in this study if you do not want to. The alternative is to not take part in this research study. If you choose not to participate in this study, it will not impact your medical treatment for ichthyosis in any way.

What if my study doctor or I do not think I should stay in the study?
You can stop taking part in this study at any time. Your decision will not affect your regular care.

Returning Study Results:
The study team will not return study results to you. You still have a right to request a copy of your medical record.

The overall results of the study will be published; however, personal information that could identify you will not be used or made available.

Important New Information:
We will tell you if we learn new information that may make you change your mind about being in this study.

Planned Sharing of Your Information:
If you agree to take part in this study, you also give permission for the use and sharing of your information. This permission lasts until the study is completed.

The information that may be collected and shared will include your:

- Personal and health information
- Past and present medical records
- Records from study visits and phone calls

The study staff, including Lurie Children's employees and Medical Staff, Northwestern Memorial Hospital (NMH) and Northwestern University (NU) may use your information and samples for this study and share it with:

- The study company, Janssen, and those working with the company.
- Northwestern University
- The Institutional Review Board (the committee that is in charge of protecting the rights of all adults and children who take part in research studies).
- Your other providers and their staff directly involved in your care, if your provider is a part of the Lurie Children's electronic health information exchange.
- The Office of Human Research Protections (OHRP), the Food and Drug Administration (FDA), or other government offices.

These are the only people to which we will give your information. We cannot guarantee that those listed above will not share it with others without your permission.

Your name will not be included in any written or verbal reports of study results.

What if I decide not to give permission to use and give out my information?
If you decide not to allow the release your information, you will not be able to take part in this study. If you give permission to the use of your information, you can withdraw it at any time. Your request should be in writing and sent to the study doctor. The study team can still use any information collected before you tell them to stop.

Can I review or get a copy my study information?
You cannot see your study records while the study is ongoing. However, any testing that relates to your medical care will be put in your medical record. You still have a right to request a copy of your medical record and tests related to regular medical care that is given during the same time as the study.

Will my information or samples be used in future research studies?
The data and samples collected from your participation in the study will be analyzed with those of all other study participants. The Study Doctor will use your data for future decision making, such as to plan other studies, to better understand the disease including diagnosis and associated health problems, as well as to understand how ichthyosis drugs work in the body.

In this research we will be looking at the DNA/RNA in your cells. Your sample will be identified by their subject number. Only the study doctor and site staff can link the number to you. During this time, your sample will be stored securely at Northwestern University.

<u>Costs Related to this Study</u>:
The study sponsor will pay for the tests, procedures, and/or medications for the study.

Payment for Taking Part in this Study:
You will be paid $800 for taking part in this study. This will be given in $50 increments as a check mailed to your home after the completion of each study visit. You will only be paid for completed study visits.

You will be asked to fill out a W-9 tax form. This form will collect your name, address, social security or individual taxpayer identification number. It will be used for processing your check or reporting to the IRS if you are paid more than $600 in a calendar year. This reporting is required under federal tax law.

In the Event of Injury:
Contact the study doctor as soon as possible, if you are injured while taking part in this study. Lurie Children's will assist you in finding medical care if needed. You or your insurer may be billed for such treatment.

Your Rights When Taking Part in this Study:
If you agree to take part in this study, you are not giving up any of your legal rights. You can stop participating in this study at any time. Your decision will not affect your regular care.

Additional Information:
Who can answer my questions about this study?
If you have any questions, contact the study doctor, Dr. Amy Paller, at 312-695-3721 or a member of the research team at 312-227-6486 during a workday or 312-695-7787 at night or on weekends.

If you have questions about your rights or if you have a complaint, you can call the IRB Office at (312) 503-7110; or via email at IRB@luriechildrens.org.

A description of this clinical trial will be available on http://www.ClinicalTrials.gov, as required by U.S. law. This website will not include information that can identify you. At most, the website will include a summary of the results. You can search this website at any time.

Please be advised that Dr. Amy Paller is receiving funding support from the company (Janssen) to conduct the study. You may request additional information about this relationship from the study doctor at any time.

You will be given a copy of this consent form. A copy will also be placed in your medical record.

Optional Research:

You can still take part in the study if you do not agree to the optional research below.

What are the purpose and procedures of this optional research?
In addition to study information, portions of samples of your specimens will be banked and stored indefinitely for future use by the research team at Lurie Children's and Northwestern. If the samples are no longer useful for research purposes at some time in the future or if you instruct us to do so, they will be destroyed.

What information or samples will be kept for this optional research?
Samples and data will be labeled only with a code number. This code number will be able to link your study data to the banked sample, but no identifying information will be recorded on the sample itself.

Who will have access to my information or samples for this optional research and how will it be kept private?
The study team at Lurie Children's and Northwestern University will have access to the data and samples in this optional research.

Can I take back (withdraw) my permission for this optional research?
If you would like to withdraw your permission to have your samples banked for future research studies, you can do so by emailing Dr. Paller or her study team.

Please **initial** next to your choice below regarding the optional testing:

Initials	YES – I agree to the storing and future use of my data and samples.

Initials	NO – I do not agree to the storing and future use of my data and samples.

Signatures:

Participant Signature:

By signing this form, I affirm: 1) I have read this form. 2) The research has been explained to me. 3) All of my questions have been answered. 4) I give my consent to take part in this research study.	
Signature of Participant or Legally Authorized Representative (LAR):	Date:
Printed Full Name:	

Signature of Authorized Person Obtaining Consent:

I certify that I have explained the above to the participant/LAR and the signature was obtained voluntarily.	
Signature:	Date:
Printed Full Name:	

Signature of Interpreter/Witness*:

☐ Not applicable, no interpreter used.
I attest that the study information has been presented to the participant/ LAR in their native language.
Signature of Interpreter/Witness:
Printed Full Name or Unique Phone ID/Company Name:

Note to Investigators: When obtaining consent from a non-English speaking participant/LAR

When a study-specific translated consent document is not available, a translated "short form" (available in several languages on the IRB website) may be used, in combination with a verbal presentation of study information (as outlined in this English consent) with the aid of an interpreter.

a. *The consent process must be witnessed by an individual who is fluent in both English and the language understandable the subject. The interpreter may serve as the witness and should sign both the English consent document and short form.*

b. *The participant/LAR should sign the short form (in the language they understand).*

c. *The investigator and/or study staff authorized by the IRB to obtain consent must sign the approved English version of the consent form.*

d. *A copy of both the IRB-approved English consent form (i.e., the summary) and the translated version of the short form must be given to the participant/LAR.*