

Know the Facts About DRESS Syndrome

Helpful answers to common questions
for DRESS patients, their loved ones,
and medical providers



DRESS SYNDROME FOUNDATION

Know the Facts About DRESS Syndrome

DRESS Syndrome is a confusing, difficult, and often isolating experience. We're glad you found this resource.

As a relatively unknown condition, DRESS patients often don't know what they have until well into their symptoms – sometimes, at life-critical stages. And medical teams are often working hard to understand a little-known condition with limited resources to help them.

We created this booklet to help DRESS patients, their loved ones, and medical providers have important and helpful answers to common questions about this disease. This content highlights the top questions we're often helping patients and their medical teams understand. You'll find details about what DRESS Syndrome is, how to diagnose and treat it, and what to be aware of.

Our goal is to empower you with an easy resource that unwinds some of the complexities of this illness. We hope this information helps you better know about DRESS Syndrome and how you can overcome this condition. And should you need perspectives or other support at any point, we're here to help.

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Know the Facts About DRESS Syndrome

1. What is DRESS Syndrome?

DRESS stands for Drug Reaction with Eosinophilia and Systemic Symptoms. The disease is also known as Drug Induced Hypersensitivity Syndrome (DIHS).

DRESS is a severe, cutaneous, adverse, delayed reaction from mainly taking one of over 50 medications. "Cutaneous" means it's related to the skin, and "adverse" means harmful. The condition usually affects people's skin, blood, and organs.

The medical community considers DRESS to be rare, in part because the condition is under-diagnosed and not reported as often as it occurs. Today's estimates show that DRESS occurs in approximately 1 in every 1,000 to 10,000 drug exposures. People die in 10% of cases – with liver failure being the most common reason. To date, FAERS has 1,609 reported cases of people dying from DRESS Syndrome (as of 7/1/24). FAERS stands for FDA Adverse Event Reporting System.

FOLLOW THIS EQUATION WHEN SUSPECTING DRESS SYNDROME

$$\text{MEDICATION}^* + \text{DELAYED REACTION OF FEVER \& RASH} = \text{DRESS SYNDROME}$$

Contact your doctor immediately if you suspect DRESS!

* within 2 to 8 weeks of first starting medicine

2. Who gets DRESS?

Anyone – regardless of age, gender, or racial background – who takes medication can get DRESS. Developing DRESS Syndrome involves an association of drugs, viruses, and the immune system. The most vulnerable people include those:



With conditions that require taking high-risk DRESS drugs



That carry a genetic predisposition to develop DRESS from specific drugs, meaning their genes may make them more at risk

3. What drugs often cause DRESS?

Many different known medications cause DRESS reactions. And as new drugs come to the market, we're learning of more medications that can cause DRESS. Five drugs are reported to cause 50% of DRESS cases and 56% of reported deaths (marked with an asterisk [*] in the lists below). The most common DRESS-causing drugs are:

Antibiotics

- Amoxicillin
- Dapsone
- Minocycline*
- Sulfamethoxazole/Trimethoprim (Bactrim)*
- Vancomycin*

Anticonvulsants

- Carbamazepine*
- Lamotrigine*
- Phenobarbital
- Phenytoin
- Valproic acid
- Zonisamide

Other

- Abacavir
- Allopurinol*
- Mexiletine
- Olanzapine
- Sulfasalazine



For a deeper list of drugs that cause DRESS, visit our website:

[DRESSsyndrome.foundation.org/drugs-associated-with-dress](https://dresssyndrome.foundation.org/drugs-associated-with-dress)



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4. What are the symptoms?

DRESS affects the whole body!

Patients experience a delayed, severe hypersensitivity (extreme physical sensitivity) response to a medication that may threaten their lives. This exaggerated immune response shows symptoms over time and may become much worse during your illness.

When Symptoms Start

Reactions typically occur within 2 to 8 weeks of starting treatment with certain drugs. But, some drugs may cause a reaction within a quicker or longer timeframe. And you may continue to experience symptoms after you stop taking the medicine that's causing your reaction.

What Symptoms Develop

Reactions can be on the outside and inside of your body. And not everyone experiences every symptom. So, it's important to pay attention to how your reaction changes.



External

The syndrome may affect your skin and include the following symptoms:

- Fever
- Rash that typically starts on upper body and progresses to lower body
- Peeling skin
- Facial swelling (edema)
- Swollen lymph nodes



Internal

DRESS can also affect you on the inside of your body:

- Hurt multiple internal organs, like liver (most common), heart, kidneys, and lungs
- Create blood abnormalities
- Damage your nervous system (brain, spinal cord, nerves)
- Cause other less common conditions, like:
 - Gastrointestinal (digestive system issues)
 - Mucositis (inflamed mouth or gut)
 - Neurologic (nerves/nervous system)
 - Ocular (eyes/vision)
 - Rheumatic (joints, tendons, ligaments, bones)

CHECKLIST: COMMON INITIAL DRESS SYNDROME SYMPTOMS

- FEVER (38° TO 40°C)/(100° TO 104°F)
- SKIN ERUPTION/RASH
- FACIAL SWELLING
- SWOLLEN LYMPH NODES
- GENERAL SENSE OF FEELING UNWELL



Have symptoms? Act now!

If you're taking medication and develop any symptoms, do the following:

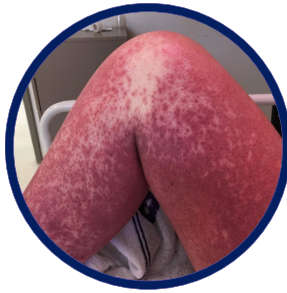
1. Blame the drug – refer to the list in *Question 3: What drugs commonly cause DRESS?* (page 1).
2. Seek medical attention immediately.
3. Work with your doctors to stop taking the medication that you suspect is causing your reaction.
4. Insist that doctors seriously consider and test for DRESS Syndrome.

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5. What does DRESS Syndrome look like?

Rash

Rashes happen in nearly all DRESS cases and often start on the trunk (chest, abdomen, chest, back). The rashes can spread to cover over half of your body. Morbilliform eruption (looks like the measles) is most common. You may also get pustules (small puss-filled blisters), blisters, and target lesions.



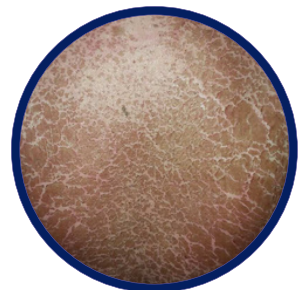
Edema (Swelling)

Swelling of the face and ears is common in 75% of DRESS cases – and can be a helpful sign in diagnosis. In many cases, the swelling can be severe and change how people look. Other parts of the body can also become very swollen.



Peeling Skin

Your skin may become exfoliative (start shedding) or peel off in patches. The skin can also peel in what seem like sheets.



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6. How do you diagnose DRESS Syndrome?

Diagnosing DRESS is tricky, because it can seem like other illnesses and symptoms can worsen over time. This progressive condition (happens in different stages) can develop even after you stop taking the medicine that you think is causing your reaction. Diagnosing DRESS requires your doctor's judgement of your clinical symptoms and laboratory findings.

Testing Requirements and Criteria

Doctors can follow well-defined criteria and a scoring systems for a diagnosis called, RegiSCAR and J-SCAR.

Your doctor will take a drug history to try and identify the medication that caused your DRESS Syndrome. They may also perform many tests to rule out other conditions and evaluate if you have DRESS.



For more information about diagnostic criteria, visit our website:

Dresssyndromefoundation.org/regiscar-diagnosing-dress



Common Tests

Depending on your symptoms, diagnostics may include tests on your:

- Blood
- Bone marrow
- Organs/organ biopsies
- Skin biopsy
- Urine



Additional Tests

You may also need the following tests to determine a diagnosis and the state of your reaction:

- CT scan
- MRI
- Echocardiograph (EC)
- Ultrasound
- Echocardiogram (ECC)
- X-ray



Important Note About Blood Abnormalities

You'll need a variety of blood tests to identify if you're experiencing DRESS.

Typical results of blood that's not acting normally include:

- Atypical lymphocytes (white blood cells that are not normal)
- Eosinophilia (an increase of eosinophils, a type of white blood cell. This condition **is not always present or necessary** to diagnose DRESS!)
- Herpes virus reactivation (a virus that awakens in your body, most common is HHV-6)
- Elevated (raised) liver enzymes
- Leukocytosis (high white blood cell count)

Be aware!

Hemophagocytic lymphohistiocytosis (HLH) may occur in severe DRESS, which is a rare blood disorder that causes life-threatening inflammation in the body.

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Common Initial and Repeating Blood Tests

Your doctors may have to continue doing blood tests to correctly diagnose and monitor your symptoms, including:

- Complete Blood Count (CBC) with differential
- Liver Function (LFT)
- Kidney Function: BUN and creatinine
- Heart (Cardiac) Function: Troponin T
- Endocrine: TSH, free T4
- Rheumatologic: CK
- Viral qPCR: (HHV-6, HHV-7, EBV and CMV)



The Link Between Herpesviruses and DRESS Syndrome

Human herpesviruses, particularly HHV-6 (which lies dormant in most people since childhood) reactivates (awakens) in over 50% of DRESS patients. Those with viral reactivation may experience a worse case of DRESS and may require antiviral treatment. As a result, HHV-6 could be an important clue in diagnosis and treatment.

Viral testing is done with blood labs, and doctors may need to test multiple times. Positive results may not show until several weeks after your first DRESS symptoms.

Other herpesviruses can reactivate in DRESS in sequential order. These include:

- Epstein-Barr (EBV)
- Human herpesvirus-7 (HHV-7)
- Cytomegalovirus (CMV)



Important Note About First Blood Tests

Since reactions can evolve, some conditions like eosinophilia and HHV-6 reactivation may not show up at first in a blood test.

Retesting for these markers during your illness could significantly improve your diagnosis and treatment.

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7. How do you treat DRESS Syndrome?

The most important first step in managing DRESS is — with the help of your doctor — to immediately stop all drugs that could be causing it. From there, start supportive care.

While no large clinical trial exists to support treating DRESS Syndrome, a global medical community of DRESS experts has proposed best practices on how to diagnose and treat patients. They based their suggestions on literature and their clinical experiences. The following information shares the most effective treatment to date and focuses on three treatment phases.



Most Commonly Accepted Core Treatment for DRESS Syndrome

Phase 1: First Treatment

1. Stop the suspected drug.

First, you'll need to work with your doctors to identify and stop the drug they think is causing the reaction. This means taking a thorough history (at least 6-weeks out from first symptoms) of your prescribed and over-the-counter medications, and any recreational drugs, contrast media dye (used in CT scans and imaging), and vaccines. If you're on many medicines at the time of a reaction, identifying the offending drug can be difficult. So, you may need to stop all medicine that isn't vital to your health. Make sure your medical team documents your suspected medications in your medical records.

Be Aware!

Treatment varies by person and condition, since people can have different reactions.

2. Provide supportive care.

You should receive individual attention to reduce the pain and stress your symptoms cause. This care can mean pain management, counseling — basically any action not focused on curing DRESS that helps you and your family get through the experience.

3. Treat with systemic and topical steroids.

Next, you can receive the following steroid treatment:

- Systemic corticosteroids (typically 1 - 2 mg/kg/day of prednisone or the equivalent of another steroid)
- Topical steroids

These steroids may be necessary when DRESS is affecting your organs. Taking them could decrease your chance of organ failure, symptom flare-ups, and autoimmune complications.

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4. Give steroid-sparing medications and additional medications, if needed.

This treatment step helps to lessen the effects of corticosteroids if you can't tolerate these drugs and when steroids alone aren't enough to control symptoms.

Additional Treatments Patients May Need

The following antivirals and steroid-sparing medications may be necessary. Taking them could decrease your chance of organ failure, symptom flare-ups, and autoimmune complications.

- Antivirals (in cases with viral reactivation)
- Cyclosporine
- Intravenous (IVIG)
- Interleukin-5 axis inhibitors
- Janus kinase inhibitors
- Mycophenolate mofetil
- Rituximab



To read research about DRESS treatment, visit our website:

Dresssyndrome.foundation.org/dress-syndrome-medical-research-and-papers

Phase 2: Ongoing Treatment

1. Repeat blood tests.

If your symptoms are severe or you don't respond well to initial treatments, you may need repeating blood tests (at least twice a week). You may also need to test for viral reactivations that could be complicating your recovery.

2. Taper steroids gradually.

As your skin and systemic symptoms improve, a gradual taper of systemic corticosteroids and/or steroid-sparing therapies will begin.

3. Manage side effects from treatments.

During this time, your medical team should monitor and manage any side effects you may have from treatments. These side effects can include ones that result from taking steroids.



Important Note About Weaning Steroids

Weaning your steroids slowly is a very important step, with a minimum of 6 to 12 weeks (some patients will need a longer course).

Otherwise, you risk having symptoms return — and possible organ failure!

For more details on how weaning too quickly can affect the heart, read *Important Note About Myocarditis* in *Question 8: What complications can develop from DRESS?* (page 8).

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Phase 3: Long-Term Treatment

1. Monitor symptoms.

Your medical team will need to monitor your symptoms for one year and potentially longer depending on your illness. They'll want to look out for the following critical reactions:

- Organ dysfunction
- Autoimmune conditions
- Viral reactivation
- Psychiatric conditions

2. Follow up with an Allergist-Immunologist.

To help you manage any future medications, connect with an Allergist-Immunologist. They may be able to help better identify your DRESS-causing drug through skin and genetic testing. Currently, some DRESS-causing drugs have a known genetic association. The Allergist-Immunologist could help you identify if your genes make you more at risk for DRESS with a specific drug.



Find an allergist at:
Allergist.aaaai.org/find/

8. What complications can result from DRESS?

Some people may receive quick diagnosis and treatment, and avoid some of the most severe symptoms. However, a crucial characteristic about DRESS Syndrome is that a first treatment that seemed successful doesn't always mean lasting recovery. Long-term complications like organ failure and autoimmune diseases are reported to occur in up to 16% of DRESS cases. (continued on next page)



Important Note About Myocarditis

DRESS-related myocarditis (heart inflammation) is an often under-recognized outcome of this condition that people can die from. It can show up both immediately and months after you first develop a fever and rash – and months after your laboratory abnormalities improve. Make sure that your medical team follows your heart throughout your illness and recovery. When suspecting myocarditis, doctors may do the following tests:

- Blood labs to check heart enzymes
- Echocardiogram
- Troponin
- Creatinine kinase
- MRI

Myocarditis can occur when corticosteroid treatment is too rapidly reduced.

You can develop two forms of myocarditis:

1. Hypersensitivity myocarditis, which often doesn't need treatment and is responsive to therapy
2. Acute necrotizing eosinophilic myocarditis (ANEM), which can result in high risk of death (up to 50%)

Patients may develop:

- Chest pain
- Hypotension (very low blood pressure)
- Dyspnea (difficulty/labored breathing)
- Tachycardia (abnormal, increased heart rate)

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Many DRESS complications can develop weeks to months or even a few years after your first symptoms. These long-term complications include but are not limited to:

Rise in Symptoms

- DRESS symptoms flare up (suddenly worsen again) if your steroids are lowered too fast
- Another medication causes symptoms (the drug may or may not be in the same class as your original DRESS-causing medication)
- Viral reactivations develop, often herpes family viruses



Organ Injury

- Organ injury, such as to the liver, kidney, and lungs
- Heart injury/Myocarditis (occurs in 13% of cases)



Auto-Immune Disorders

You may develop autoimmune disorders that typically occur months to years after you first develop DRESS, such as:

- Alopecia areata
- Fulminant + Type 1 diabetes
- Hemolytic anemia
- Myocarditis
- Thyroiditis
- Systemic lupus erythematosus



Psychiatric Conditions

You may develop a psychiatric response from your DRESS experience, like:

- Anxiety
- Depression
- PTSD

Continue to Monitor Your Health

For these reasons, it's very important that your medical team continues tracking your health, even if you think you've completely recovered from DRESS Syndrome.

The better you understand how your health is progressing, the better you can monitor for long-term complications. You'll also better inform decisions to avoid a repeat of symptoms – and stay healthy.



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9. What should you do after you are discharged from the hospital?

After your medical team discharges (releases) you from the hospital, it's important to pay close attention to your health. You'll want to take certain steps to be sure you support your ongoing recovery and to prevent potential relapses or complications.

Here are some key recommendations:



Maintain Good Health Practices

Focus on your overall health and wellbeing. Eat healthy, drink enough water, get enough sleep, and exercise. Be sure to avoid too much time in the sun, which can make your skin sensitive.



Go to Follow-Up Care

Attend all scheduled follow-up appointments with your healthcare providers. These visits are very important for monitoring your recovery progress. They also will check your organ function (such as liver and kidneys) and adjust any ongoing medications. DRESS can take many weeks to months to fully resolve, and you may see a number of specialist during this time. Keep your general practitioner well informed of your progress.



Avoid Medication That May Be Harmful to You

Avoid the medicine that triggered your DRESS Syndrome in the future. If your medical team added a drug that caused your reaction, make sure they tell you which specific drug or drug classes you should avoid. It's very important to communicate this information to all healthcare providers in the future to prevent accidental exposure!



Continue Education and Awareness

Educate yourself about DRESS Syndrome and its potential triggers. Be vigilant about reading labels on medications. Inform healthcare providers about your history of drug allergies or reactions. Every step can make a difference in your health.

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Monitor for Signs of Relapse

Be aware of any new symptoms that could indicate a relapse or ongoing complications. Contact your healthcare provider immediately if you experience any symptoms such as rash, fever, swollen lymph nodes, or organ-specific reactions. Also, be on the lookout for signs of heart issues (myocarditis) like chest pain, shortness of breath, high heart rate, dizziness, and vomiting.



Consider Using Medical ID

Medical ID is an alert bracelet that carries your medical details card. This information lists your history of DRESS Syndrome and the medicine(s) that affected you. For emergencies, this ID can very helpful in your treatment.



Join Support Groups

Consider joining a support group or receiving counseling if you feel you need it. Coping with a severe drug reaction and a relatively unknown condition like DRESS Syndrome can be emotionally challenging. Connecting with others who have experienced similar situations can be helpful.



Join our Facebook support group, The DRESS Syndrome Community:

@D.R.E.S.S.SyndromeCommunity



Remember, your complete recovery could takes months or longer – everyone is different. We encourage you to be patient with yourself and surround yourself with a support network who can further help you foster positivity and wellbeing for years to come!



Continue to Track Your Symptoms

Be sure you and your medical team watch your symptoms for at least a year after starting your initial treatment. If you notice symptoms coming back or getting worse, get medical help immediately.

Be diligent – and check in with your medical providers often!

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Track Your Symptoms

Once symptoms start, DRESS moves quickly. And every day with DRESS Syndrome can be different. If you suspect DRESS, document every reaction. Take pictures to show what's happening to your medical team.

Use this page to help you keep track of your symptoms. Every detail counts.

Know the Facts About DRESS Syndrome



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DRESS SYNDROME FOUNDATION

We're here to help!

The DRESS Syndrome Foundation is a 501(c)(3) nonprofit based in Richmond, Virginia, USA. We're a collaborative network of patients, families, researchers, and physicians dedicated to supporting people worldwide who are experiencing DRESS Syndrome. We also educate the public and medical communities about the syndrome, and advocate for advanced research and treatment.

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