

D.R.E.S.S. Syndrome

QUICK FACTS

Drug: Caused by many drug classes, most commonly:

- Anticonvulsants: Phenytoin, carbamazepine, phenobarbital, lamotrigine.
- Antibiotics: Vancomycin, minocycline, sulfamethoxazole/trimethoprim (bactrim), amoxicillin, dapsone.
- Other: Abacavir, allopurinol, sulfasalazine, olanzapine

Reaction: A delayed, severe hypersensitivity response to a medication that may be life-threatening.

Eosinophilia: An excess of eosinophils (type of white blood cell involved in allergic reactions) often found in the blood of DRESS patients. It's a helpful diagnostic marker but is not necessary for diagnosis. Atypical lymphocytes are also common.

Systemic: DRESS affects the whole body! It may involve the skin, multiple internal organs, blood abnormalities, and the nervous system. The liver is most common organ affected, but also the kidneys, lungs and heart. Blood abnormalities may include eosinophilia, atypical lymphocytes, leukocytosis, elevated liver enzymes and herpes virus reactivation (most notably: HHV-6).

Symptoms: May occur days to weeks after starting a medication and even after a drug is stopped. Initial symptoms of fever and rash are often followed by facial swelling, blood abnormalities and organ injury.

First signs usually include:

- Skin eruption/rash
- Fever (38 to 40C)/(100 to 104F)
- Facial swelling (edema)
- Enlarged lymph nodes

HHV-6 VIRUS ASSOCIATION

Human herpesvirus 6 (among other herpes viruses) has been shown to play an important role in DRESS. This virus, which lays dormant in almost everyone, has been demonstrated to reactivate in over 50% of DRESS patients. Those with viral reactivation may have a more severe course of illness.

DIAGNOSIS AND TREATMENT

It is most important to identify and discontinue the offending drug. However, DRESS is tricky in that it can continue to get worse even after stopping the drug. It is imperative to diagnose DRESS quickly and follow up with proper testing. Due to the progressive nature of the illness, certain conditions like eosinophilia and HHV-6 reactivation may not show up initially in blood test. Retesting for these at the right time could make a significant difference in diagnosis and treatment.

Well-defined criteria (RegiSCAR) has been developed to aid in a scoring system for diagnosis.

Treatment will vary based on confirmation of symptoms but may include oral steroids, IVIG and antivirals.

COMPLICATIONS

Seemingly successful, initial treatment of symptoms are not necessarily an indication that the patient is home free. Many complications of DRESS can develop weeks to many months later, including:

- Serious flares during steroid taper
- Myocarditis
- Organ Injury: Liver, Kidney and Lung
- Autoimmune disorders: Type 1 diabetes and thyroiditis
- Recurrent viral reactivations
- A flare in symptoms caused by another medication (can be related or unrelated to original offending medication).