Transcript Details

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An Expert Perspective on Polygenic Autoinflammatory Diseases

Announcer:
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This episode of Living Rheum, titled An Expert Perspective on Polygenic Autoinflammatory Diseases," is sponsored by Novartis US Clinical Development and Medical Affairs. The host and speaker have been compensated for their time. This program is intended for health care professionals.

Here's your host, Dr Jason Liebowitz.

Dr Liebowitz:
For pediatric patients, an unusual collection of symptoms may be a sign of a single inflammatory disease. What do we need to know about these types of disease? This is ReachMD, and I'm Dr Jason Liebowitz. Joining me to discuss everything we need to know about polygenic autoinflammatory diseases is Dr Bella Mehta. Dr Mehta is an assistant attending physician at the Hospital for Special Surgery and an assistant professor of medicine at the Weill Cornell Medical College in New York City. She specializes in research and care of patients with different rheumatic conditions, including Still's disease. Dr Mehta, thanks for being here today.

Dr Mehta:
Thank you so much for inviting me, and thank you so much for your kind introduction, Dr Liebowitz. Looking forward to discuss with you about this.

Dr Liebowitz:
Dr Mehta, can you start by defining polygenic autoinflammatory diseases for our listeners? How does this collection of diseases typically present?

Dr Mehta:
In general, autoinflammatory disorders are characterized by seemingly unprovoked episodes of systemic or organ-specific inflammation, in the absence of high-titer autoantibodies or self-reactive lymphocyte populations. Unlike monogenic autoinflammatory diseases, such as familial Mediterranean fever, or FMF, and cryopyrin-associated periodic syndromes, or CAPS, which are due to a single gene mutation, polygenic autoinflammatory diseases are due to multiple genetic factors, and that includes conditions which are most common which are systemic juvenile idiopathic arthritis or sJIA (the other name for it is also Still's disease), Crohn's disease, and ulcerative colitis. Type 2 diabetes is also an autoinflammatory disease, in addition to its metabolic dysregulation.

Dr Liebowitz:
As we know, Still's disease and systemic juvenile idiopathic arthritis share many of the same clinical features. How similar are these 2 diseases?

Dr Mehta:
Systemic JIA, or systemic juvenile idiopathic arthritis, does in fact present with many of the same clinical features as Still's disease. In fact, we consider sJIA, which is the short form, and the adult-onset Still's disease basically the same disease just in different age groups and it's the same spectrum. The clinical features involved include quotidian fevers, rash, sore throat, hepatosplenomegaly, lymphadenopathy, and sometimes serositis.Complications that can arise from Still's disease include macrophage activation syndrome, which is often life-threatening and important to recognize early on. Some of the other complications, mainly seen in the inpatient setting because patients are very sick, pulmonary arterial hypertension, interstitial lung disease, and alveolar proteinosis. Systemic JIA is important to recognize early on because when they get into complications like MAS, sometimes it needs very quick and aggressive

treatments, and if they're not treated properly and early on, the mortality can be very, very high in these patients. There is, actually, just because sJIA and Still's disease is basically the same spectrum, there's a movement to standardize the name of the disease so that it is the same across the spectrum and it's easier from a treatment perspective and understanding the pathophysiology perspective.

Dr Liebowitz:
Thank you so much for that very helpful discussion. Now let's shift our focus from pediatric to adult care. How should we manage this transition of care for patients?

Dr Mehta:
Often patients with autoinflammatory disease, such as Still's disease, have a disease burden which spans over many decades. So, the need to transition and have a plan to transition from a pediatric to an adult not only rheumatologist, but pediatric to adult providers, should be done meticulously with a well hand-off system. Sometimes these patients have severe systemic manifestations which require specific management. Also, some of these patients may have aggressive chemotherapy agents or something done early on, and it is important to hand off that to the adult rheumatologist too. These management strategies need to be clearly detailed, and during transition of care patients having high-risk of being lost to follow-up, and thus, involving social workers in the transition may be required. Having patients adjust from a family-centered interaction in the pediatric population to a patient-centered interaction in the adult population is also important. And sometimes, just given the age of the patients, pediatric and adult rheumatologists have different ways and goals of approaching these patients. And it's important for the patients to also somehow see the transition and recognize that.

Some of these patients can also, require multiple specialty physicians treating them. So, years when a lot of flares happen, when patients are going from pediatric to adult, they are seeing multiple providers, like say pulmonology, rheumatology, maybe nephrology, and when they now need to see all of the new providers, it is often overwhelming for these patients. So again, I think you bring up a really good point that transition of care is very, very important in these patients.

Dr Liebowitz:
Well, with those final thoughts in mind, I want to thank my guest for helping us better understand polygenic autoinflammatory diseases. Dr Mehta, it was a great pleasure speaking with you today.

Dr Mehta:
Likewise, Dr Liebowitz. It was great to speak with you today, and I think it is very important to take the big takeaways from this is that Still's disease needs to be recognized early. Some of the symptoms and signs can be tricky and thus rheumatologists need to be on the lookout for it. And it is important to have adequate transitions of care, especially when transitioning from the pediatric to the adult rheumatologists and the adult physicians.

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