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### Who Are the Key Players for a Multidisciplinary Approach to Diagnosis and Management of Non-Advanced Systemic Mastocytosis?

#### Announcer:

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#### Dr. Akin:

Hello, I'm Dr. Cem Akin. I'm a Professor of Medicine at the University of Michigan. And in this episode, I would like to talk about the key players for a multidisciplinary approach to diagnosis and management of non-advanced systemic mastocytosis.

This is a disorder that often requires management by multiple specialists, because when mast cells get activated and cause the symptoms of this disease, this activation could occur in a number of different organ systems, including cardiovascular, skin, digestive, respiratory, and even musculoskeletal and neurologic symptoms. And when we look at our patients how - in terms of how they present with non-advanced systemic mastocytosis, about 70 to 80% of these patients present with skin lesions called urticaria pigmentosa, and this is the older term, in the new literature, you might also see make your papular cutaneous mastocytosis. So, these patients have fixed hyperpigmented lesions that occur in their trunk and extremities. And they usually present to a dermatologist because they have a skin lesion that does not go away and they may urticate and may become itchy and redder if they are exposed to heat, temperature changes, or friction, but they always stay in the same spot. And the dermatologists – experienced dermatologists – usually diagnose this just by morphologic appearance of the lesions, but if there is any doubt, a biopsy can be taken and dermatopathologist can factor in terms of the diagnosis.

And the rest of the patients present with anaphylaxis, flushing, or other signs of mast cell activation. And these patients may not have skin lesions and, so one has to maintain a high level of suspicion to diagnose these patients when they present with anaphylaxis, flushing, or other symptoms. And of course, the specialist – the major specialist – in diagnosing these patients is allergists, because that's, by definition, who deals with anaphylaxis and allergic-type symptomatology. But these patients can also present to a gastroenterologist because of abdominal symptoms like diarrhea, bloating, nausea, or even endocrinologists sometimes see these patients as the first point of contact because of their flushing, and sometimes severe osteoporosis.

Now, when a patient presents with anaphylaxis, the question becomes which of these patients presenting with anaphylaxis need to be worked up for systemic mastocytosis? Because obviously, not all patients with anaphylaxis will have mastocytosis as an underlying cause. So this is a table to kind of give you some guidance on what patients might need a systemic mastocytosis workup and a bone marrow biopsy referral. So this score was developed by our Spanish colleagues, and it's called the REMA score. And it looks at gender, clinical symptoms, and tryptase levels, and if the – and then assigns different points based on these variables. If the patient is male, they get +1, females get -1 point, and absence of urticaria and angioedema get +1, urticaria and angioedema, if they are present during anaphylactic episodes get -2, and presyncopal or syncopal episodes get +3. And tryptase levels, if they are less than 15, they get -1, and if they are greater than 25, they get +2. So at the end, you add up all of these pluses and minuses. And if your score is 2 or higher, then your patient will have a high probability of having systemic mastocytosis as the underlying diagnosis of their mast cell activation or

anaphylactic symptoms, and that patient should be referred to a bone marrow biopsy for diagnostic procedure.

So – and that's when the hematologists' and hematopathologists' expertise become very important because they are the specialists who perform the bone marrow biopsy and diagnose it based on the criteria. And so they are often indispensable in initial diagnosis of systemic mastocytosis, and they are also – they also manage the advanced varieties of systemic mastocytosis presenting with hematologic disorders. Allergists and immunologists can be involved in general management of the patient as well as mast cell activation symptoms and venom allergy, treatment with venom

Immunotherapy. And the gastroenterologists can perform endoscopies to figure out whether the gastrointestinal symptoms are due to mast cell infiltration or simply due to mast cell mediator release. And finally, the endocrinologists are important partners in management of osteoporosis.

When considering a referral, because of mastocytosis being a rare disease, there is often lack of localized expertise, and these two websites could be very helpful in figuring out where the expertise is located. In the United States, we have the American Initiative and Mast Cell Diseases website, and in the Europe, ECRM website provides useful information on that to refer these patients.

I hope I was able to convey the complexity of symptomatology and the number of specialists that may encounter these patients in this episode, and I thank you for your attention.

**Announcer:**

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