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### Challenges in the Management of Difficult-to-Treat HAE

Announcer:

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Dr. Buttgereit:

This is CME on ReachMD. I'm Dr. Thomas Buttgereit, and I'm not alone today. Here with me is Dr. Anna Valerieva. Our topic today is challenges in the treatment and management of difficult-to-treat HAE.

So, Anna, I think that it's a very challenging question I will ask you now. What do you understand by the definition of difficult-to-treat HAE?

Dr. Valerieva:

Thank you, Thomas. I think this is a great and really challenging question because we really do not have a clear definition on the topic of difficult-to-treat HAE. From my perspective I would argue that difficult-to-treat HAE is not always severe HAE. So difficult- to-treat HAE surely, are the patients with severe HAE who tend to have severe and frequent HAE attacks, and they might be put on prophylaxis. But for me, difficult-to-treat HAE are those patients who would have breakthrough attacks, and they will have, also, challenges in recognizing the attacks, or challenges in treating those acute breakthrough attacks under prophylaxis.

But I think difficult-to-treat HAE might be, also, patients who tend to have certain comorbidities, and those comorbidities might also be influencing the course of HAE. So something in my approach, usually, is to check the patients with difficult-to-treat HAE for underlying diseases and to control those diseases, and whether they might have some benefit from optimizing their patient care with regards to another disease.

Difficult-to-treat HAE, I would also add, are the patients who tend to not recognize the HAE attacks. I don't know if you would also agree from your practice, that no matter how much we discuss with the patients from time to time, I tend to have a patient in the clinic that do not recognize the abdominal attacks or are not always sure whether this is an HAE attack, or something related to gastritis or some other abdominal crisis. And this is indeed a true challenge in our HAE practice on a daily basis, that we really do lack the biomarkers that can help in such situations, to recognize whether this is an HAE attack or something else.

I also would say that difficult-to-treat HAE are the patients who have low compliance to the disease therapies. And from my practice, I always see some patients that try to negotiate the need for a blanket treatment under acute circumstances. Or the patients who tend to

skip their prophylactic agents, and this is increasing the risk for having a breakthrough attack.

Dr. Buttgereit:

Yeah, really, really, a lot of points you mentioned already, what you understand under this for difficult-to-treat HAE. So I think, really, it has a lot to do with compliance, with the nature of the disease itself, but also, with the treatments we have available to treat HAE. And we know that we have modern treatment options available, and many of them work perfectly, but there are still patients who face attacks, are not happy with their treatment because they still have the symptoms of HAE.

So the next question would be, then, Anna, for you. Are there any guideline recommendations on how to treat these patients with difficult-to-treat HAE?

Dr. Valerieva:

So as far as I'm aware, the guidelines are not really clear on defining what is difficult-to-treat HAE, and we really do lack specific recommendations in those patients. So this is something that we, as HAE specialists, should be looking forward to defining how to better manage patients with difficult-to-treat HAE.

Dr. Buttgereit:

Yeah, I can completely understand this, dear Anna. So I think it's a work in progress.

So maybe from my point of view, many years ago, we had a different definition on how to treat HAE, what is difficult-to-treat HAE. So with the modern treatments we have now available, we have probably another more different definition of what is difficult-to-treat HAE. And I think future guidelines should then, more focus on this, especially on the patient populations who still face attacks. What could be the reasons, and what could be the measures, too. Yeah, find out what the reason for the attacks is, and especially, what the recommendations and how to treat and manage these patients are.

So unfortunately, our time is up now. So, Anna, it was great to have you here in this episode. Thanks for the great discussion, and I will hand over the final words to you.

Dr. Valerieva:

Thank you, Thomas. I think something that I would like to also underline here, is for patients with difficult-to-treat HAE, as HAE specialists, we should always be supportive to our patients and if we face such patients, maybe it is up to us to schedule more frequent visits, to always have a plan, and try to guide the patient to finding a solution.

Because we have the more therapies, we have different options, and I think this is something that we can add on to their treatment.

Dr. Buttgereit:

Great. We hope this information was helpful for you and useful for your practice.

Announcer:

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