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Optimizing Patient Management Strategies

Announcer:
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Dr. Vissing:
This is CME on ReachMD. My name is John Vissing, and I'm going to talk to you about the daily management of patients with limb-girdle muscular dystrophy type R9.

So one important issue for these patients – one is the skeletal muscle issue, which is very often the most apparent thing – but that's the cardiac issues. And patients can develop a quite severe cardiomyopathy and need treatment. So typically, these patients are followed at the cardiologist. If you have completely normal findings in the patients, you don't necessarily have to follow up a patient every single year. You can space it out 2 or 3 years. But once you find something abnormal, you would typically go to a yearly follow-up for these patients.

The cardiomyopathy these patients develop is typically not as severe as we see it, for instance, into Duchenne and Becker muscular dystrophy. So the number of heart transplants are much lower than you will see in Becker muscular dystrophy patients.

Another important issue that also affects these patients is the respiratory function, and especially if you have a childhood onset, you are in need of respiratory help already in your late teens and invasive ventilation typically when you reach the age of 30.But milder patients should also be followed with respiratory checks. And in our hands, and I think in most other hands, when you reach an FVC of 60% of predicted, you should refer the patient to a respiratory unit where the patients can also have a sleep study to monitor for nocturnal hypoventilation, which calls, then, for some assisted ventilatory support for these patients.

Another important issue here is how much should these patients exercise? In the old days, this was thought to be detrimental to patients, but we know now from exercise studies that this is, in fact, quite important that they exercise, and we typically prescribe aerobic exercise for these patients.

Another and perhaps a little bit overlooked thing is the psychological support for these patients. I think we are sort of short of psychologists many places, but imagine getting a diagnosis when you are 26 and you are in the middle of your education, planning to get children and family, and then all of a sudden, you're hit by a genetic disease. This is really a big crisis for a person, and I think in many, we are disregarding how big this crisis is. And we are here at our place doing now a study with our crisis psychologists where we map patients' well-being psychologically, and then we offer them group cognitive therapy for 3 years or we follow them for 3 years.And hopefully we will be able to know exactly whether this is any help for the patients.

Otherwise, obviously, patients, like other patients with muscle wasting diseases, are in need of help in assistive devices for walking, also help if they need a wheelchair. When should you go into a wheelchair?You shouldn't try and fight and fight and fight as long as you can. When you start falling too much, this is probably the time when you should consider going into a wheelchair.

Genetic counseling is quite easy, in most cases, for these patients. It's a recessive disease, and if your partner is not related, the likelihood of you passing this on to your children is very small, except maybe if you live in Scandinavia, where the carrier frequency is 1 in 150. There, you could consider testing your spouse. But otherwise, it would not be a great concern.

Hopefully, you can use some of these tips in your own clinical practice and thank you for listening.

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