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EDITORIAL

WILEY Congenital Heart Disease

New intriguing subsets of pediatric hyperlipidemia patients

As we are caring for more children with hyperlipidemia, it is becoming increasingly obvious that there are certain clinical subsets that demand more attention and investigation. With the epidemic of childhood obesity we are currently experiencing, there are more and more children coming in with acquired hyperlipidemia in addition to genetic forms.

One of the biggest issues facing us in our lipid patients are the heterozygotes with familial hypercholesterolemia. The heterozygotes far outnumber the homozygotes and yet there is still much to learn about this group of patients. There is no question that the heterozygote develops early coronary disease in their 30s and 40s. The real question is, when does that coronary disease begin to develop and there are no real answers so far in the literature. If one knew for example that coronary artery disease almost never occurs under age 20, then the evaluation and intense investigation would occur more in young adulthood. If, however, there are signs of early coronary disease in the early to mid-teens, then pediatric cardiologists and pediatric hyperlipidemia specialists would need to become much more actively involved in the evaluation, diagnosis, and treatment of early coronary disease in this population. It is critical to set a new paradigm for the heterozygotes to determine when they actually begin to develop coronary artery disease and how best to evaluate that. Should a cohort of patients all have CT scans or MRIs at a predetermined age, how best is it to evaluate that information at an early age?

In addition, there are now groups of patients that we see with very low and very high HDL cholesterol levels. The patients with HDL cholesterol levels under 20 and those over 70 or 80 comprise a very interesting group of patients. These patients will need long-term follow-up to see if the low HDL patients are really at increased risk for early coronary disease and if the high HDL patients are protected from coronary disease. These studies involve long-tern follow-up and categorization of their patient populations but to define those patient populations currently is an important undertaking and should be done.

Another fascinating group of patients are those that come in with morbid obesity that have very low lipid levels, particularly very low cholesterol, LDL, and triglyceride levels. These patients are unique and provide another subset of lipid patients that need further investigation. Why, in the face of morbid obesity, are their lipids so low and what characterizes this group of patients clinically?

Another subset of patients are those with elevated LPa. LPa is clearly a risk factor for early coronary disease but our data suggest that it also may be a risk factor for idiopathic stroke. In addition to that, the interplay between low HDL cholesterol levels and LPa is important in this stroke population. One of the problems is there is very little that one can do treatment-wise with an elevated LPa, other than niacin. However, niacin is ineffective in many patients. We have seen, however, some patients respond to diet and exercise and that bears further investigation.

As we see more and more of these various subsets of our lipid patients, we need to get to clinically characterize these patients and have a primary base of understanding of what they all look like clinically, as we follow them over time. Keeping these patients in the medical system and following them over extended periods of time is going to be important, but it is also important to know what the clinical subsets look like today, as we progress into the future. Clearly, the more lipid patients we see, the more we find that there are subsets that merit significant further clinical investigation.

