Some Peripartum Cardiomyopathy Can Be Deadly

BY BRUCE JANCIN
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STOCKHOLM – Healthy women who are first-degree relatives of patients with known familial dilated cardiomyopathy should be followed closely during pregnancy and the first 5 months post partum for the emergence of life-threatening peripartum cardiomyopathy.

A significant subset of peripartum cardiomyopathy is the initial manifestation of familial dilated cardiomyopathy. Thus, when women with peripartum cardiomyopathy don’t fully recover left ventricular function, their first-degree family members should undergo screening for presymptomatic dilated cardiomyopathy. Dr. Karin Y. van Spaendonck-Zwarts said.

Dr. van Spaendonck-Zwarts offered three lines of evidence in support of the hypothesis that a subset of peripartum cardiomyopathy is actually a first manifestation of familial dilated cardiomyopathy rather than, as previously assumed, a sporadic event.

One, in reviewing a Dutch cohort of 90 proven dilated cardiomyopathy families, she and her coinvestigators discovered five families, or 6%, had members with peripartum cardiomyopathy. This is far too high a figure to be due to chance.

Peripartum cardiomyopathy is a rare disorder, with an estimated incidence in the United States of 1 case in 4,075 live births, noted Dr. van Spaendonck-Zwarts of University Medical Center, Groningen, the Netherlands.

Second, when the Dutch team reviewed their 10 most recent cases of peripartum cardiomyopathy, they found 3 of the 10 women did not show a full recovery of left ventricular ejection fraction and dimensions. Cardiologic screening of all first-degree relatives of these three women revealed previously undiagnosed cases of dilated cardiomyopathy in all three families.

Third, an in-depth genetic analysis conducted in one of the dilated cardiomyopathy families with a member who had peripartum cardiomyopathy revealed a mutation in TNNC1, the gene encoding cardiac troponin C. This supports the genetic nature of the disease in this particular case of peripartum cardiomyopathy.

“Together, these findings strongly suggest that a subset of peripartum cardiomyopathy is part of the spectrum of familial dilated cardiomyopathy presenting in the peripartum period,” said Dr. van Spaendonck-Zwarts.

If the Dutch findings are confirmed elsewhere, the European Society of Cardiology classification of the cardiomyopathies may need to be amended. The ESC characterizes peripartum cardiomyopathy as a nonfamilial, nongenetic form of dilated cardiomyopathy, she observed.

Dr. van Spaendonck-Zwarts declared she has no relevant financial interests.

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