Cardiac Neonatal Lupus Risk Factors for Death

Fetal risk factors linked with mortality included hematologic hepatic lupus and early delivery.

BY SHARON WORCESTER

FROM THE ANNUAL MEETING OF THE AMERICAN COLLEGE OF RHEUMATOLOGY

ATLANTA – The overall case fatality rate in cardiac neonatal lupus is nearly 18%, according to a review of data from the Research Registry for Neonatal Lupus.

Of 325 children enrolled in the large U.S.-based registry before October 2010, 57 (18%) died; 30% died in utero, 30% died during the neonatal period, 14% died between 1 and 6 months of age, and 26% died after 6 months of age, Dr. Peter M. Izmirly reported at the meeting.

Of the deaths, 42 were cardiac related – most often a result of complications from cardiomyopathy, 6 were due to infectious complications, and 8 were a result of unknown causes. One pregnancy was terminated electively, said Dr. Izmirly of New York University, New York.

"There was a significantly higher case fatality rate in minorities, compared with Caucasians," Dr. Izmirly said, noting that 14% of white children with cardiac neonatal lupus died, compared with 28% of children belonging to minority groups.

The study, which was conducted in an effort to update morality data on cardiac neonatal lupus and to thereby improve evidence-based counseling of anti-Ro/La positive mothers whose babies are at increased risk of cardiac neonatal lupus, identified fetal and maternal risk factors for death in affected babies.

Significant fetal risk factors for death were associated hematologic hepatic neonatal lupus (present in 27% vs. 7% of deceased vs. living babies), earlier gestational age at detection (detection occurred at 21.8 vs. 23.4 weeks in deceased vs. living babies), delivery prior to 37 weeks' gestation (delivery occurred prior to 37 weeks in 69% vs. 42% of deceased vs. living babies), and earlier gestational week of delivery (delivery occurred at 34.2 weeks vs. 36.9 weeks in deceased vs. living babies), Dr. Izmirly said.

Fetal risk factors not found to be associated with mortality were female sex, associated neonatal lupus rash, cesarean section delivery, and year of birth.

Fetal echocardiographic risk factors associated with mortality were lower ventricular rate nadir (rate was 50.2 vs. 53.6 in deceased vs. living babies), and the presence of endocardial fibroelastosis (which occurred in 30.25% vs. 4.3% of deceased vs. living babies), dilated cardiomyopathy (which occurred in 32.6% vs. 8.6% of deceased vs. living babies), hydrops (which occurred in 57.4% vs. 3.4% of deceased vs. living babies), and valvular disease (which occurred in18.2% vs. 4.8% of deceased vs. living babies).

VITALS

Major Finding: Of 325 children enrolled in the Research Registry for Neonatal Lupus before October 2010, 57 (18%) died; 30% died in utero, 30% died during the neonatal period, 14% died between 1 and 6 months of age, and 26% died after 6 months of age.

Data Source: A retrospective analysis of data from a large U.S.-based cohort.

Disclosures: Dr. Izmirly had no disclosures.

Fetal echocardiographic factors not associated with mortality were ventricular rate detection, atrial septal defect, ventricular septal defect, and patent ductus arteriosus.

Only one maternal risk factor – a maternal diagnosis of systemic lupus erythematosus or Sjögren's syndrome – showed a trend toward significance in terms of risk for fetal death. Diagnosis occurred in 56% of women whose babies died, vs. 43% of those whose babies were living.

Maternal age, maternal anti-La antibodies, maternal anti–52-kD Ro antibodies, and use of nonfluorinated steroids, fluorinated steroids, terbutaline, or hydroxychloroquine were not associated with increased risk of fetal mortality.

As for morbidity in affected children, 70% required pacing – including 1% in utero, 53% in the neonatal period, 12% between ages 1 and 6 months, and 32% after age 6 months. The timing was unknown in 3% of cases.

Also, four children required cardiac transplantation, including one child who required two transplants, Dr. Izmirly said.

"Cardiac neonatal lupus is associated with substantial mortality, which is predicted by slower heart rates and echocardiographic abnormalities consistent with antibody-associated disease beyond the AV node," Dr. Izmirly concluded, adding that the disparity in outcomes between whites and minorities suggests that attention should be given to "an inherent difference in organ response to injury, and/or access to care." ■

Endovascular Aortic Repair Aids Some With Marfan

BY DAMIAN McNAMARA

FROM THE ANNUAL MEETING OF THE SOUTHERN ASSOCIATION FOR VASCULAR SURGERY

NAPLES, FLA. – Some patients with Marfan syndrome benefit from endovascular aortic repair, according to a review of 16 patients who were treated at the University of Florida, Gainesville, between January 2000 and June 2010.

Most patients had a history of multiple interventions, which points to the complexity of Marfan syndrome and a need to follow patients closely over the long term, Dr. Alyson L. Waterman said at the meeting.

Dr. Waterman and her associates reviewed their experience with endovascular abdominal aorta repair (EVAR) and thoracic endovascular aorta repair (TEVAR) for Marfan syndrome patients at the University of Florida.

After a median of 8.3 months, seven patients had clinical and radiographic evidence of successful treatment. Another six patients were primary treatment failures (for example, they had a type I endovascular leak or had persistent false lumen flow), and two patients had a secondary treatment failure, that is, initial success followed by proximal or distal aortic failure unrelated to the original intervention site. The remaining patient was lost to follow-up.

"Results are sobering but support [the concept that] some Marfan syndrome patients benefit from endovascular therapy," said Dr. Waterman of the department of surgery at the University of Florida, Gainesville.

The main causes of death in Marfan syndrome are aortic dissection and rupture, Dr. Waterman said. She pointed out that open surgery is still required for repair of the ascending aorta, but endovascular intervention is an option for descending thoracic and abdominal aorta repair for those with this connective-tissue disorder.

The 16 Marfan syndrome patients in the series underwent a total of 19 relevant procedures (1 EVAR, 15 TEVAR, and 3 combined procedures). The median patient age was 52 years. Three patients had a secondary TEVAR within 28 months of their index intervention.

In all, 15 of the patients had previous surgery (from 17 years to 1 week prior) of the ascending aorta or arch.

Chronic dissection and/or aneurysmal dilation of the descending aorta were the indications for elective intervention in 13 patients. Two acute dissection/malperfusion cases and one anastomotic disruption early after an open surgery led to emergency intervention in three patients.

VITALS

Major Finding: In all, 7 of 16 patients with Marfan syndrome had successful endovascular treatment of their aortas.

Data Source: Review of endovascular repairs at the University of Florida, Gainesville, from 2000 to 2010.

Disclosures: Dr. Waterman and Dr. Endean said they had no relevant disclosures.

Four of the TEVAR procedures required adjunctive endovascular procedures. They comprised one subclavian artery embolization, one vertebral artery stent, one renal artery stent, and one celiac and superior mesenteric artery stent.

All four EVAR procedures required complex adjunctive endovascular procedures involving visceral arteries.

Five patients died during follow-up. Of them, two died perioperatively: the patient who underwent emergent TEVAR for anastomotic disruption, and a patient who required multivisceral revascularization in conjunction with a second TEVAR. Two patients died following discharge: one because of a respiratory failure at 3 months, and the other because of a cardiac arrest after 4 months.

The remaining patient died more than 6 years after EVAR from advanced age (84 years).

"This is clearly a group for whom treatment options are difficult at best," said study discussant Dr. Eric D. Endean of the University of Kentucky, Lexington.

All were deemed poor candidates for open surgery, and they had a history of an average of almost two previous aortic operations. Six (38%) were classified as primary treatment failures, all required open repair, and half died, Dr. Endean commented. "Results are indeed sobering."

"As you point out, diagnosis of Marfan syndrome was based on clinical diagnosis," Dr. Endean said. He asked how confident Dr. Waterman is about the diagnoses in her series.

"That is one of the difficulties with Marfan syndrome; a lot is based on clinical diagnosis alone." Not all patients undergo genetic screening for the fibrillin-1 mutation, she said.

A small population that makes the study "essentially a case series" is a limitation, Dr. Waterman said.

"Open surgery still has a big role in replacement of these aortas because these aortas are not genetically normal," Dr. Waterman said.

However, there is still a place for endovascular therapy and a need to identify which patients are likely to benefit, she explained.

Dr. Waterman added that the cardiologists at University of Florida also treat Marfan syndrome patients, and they have yet to look at these patient populations at the two institutions combined. ■