SPECIAL NEEDS: REALIZING POTENTIAL

A New Era for Treatment of Muscular Dystrophy

Diagnosing, Treating MD

When muscular dystrophy is suspected, a neuromuscular specialist can be extremely helpful to families and patients. Diagnostic and management considerations for DMD are expected to be published later this year by the Centers for Disease Control and Prevention.

There are no drugs currently approved for use in MD. Prednisone has proven efficacy in DMD, with a dose of 0.75 mg/kg per day considered an appropriate starting dose. Injections of intravenous injections of dystrophin, located on the X chromosome at Xp21. Between 2 and 4 years of age, children may begin to show signs of leg weakness or have some mildly delayed motor milestones. Parents may notice they have difficulty running or jumping or keeping up with their peers.

Care of both pediatric and adult muscular dystrophy patients is highly variable across the United States. The optimal management of MD is multidisciplinary, as the disease affects so many systems and aspects of a child’s life. Even though we think of muscular dystrophy as a genetic disease, there are a cognitive component, respiratory and cardiac issues, and other issues that are important to address early on. Getting one health care provider to coordinate care is particularly helpful to families and patients. Diagnostic and management considerations for DMD are expected to be published later this year by the Centers for Disease Control and Prevention.

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Study: CT Scans Pose Risks for Pediatric Trauma Patients

Baltimore — CT scans are responsible for 91% of total radiation exposure in pediatric trauma patients, even though they usually imaging do not undergo imaging studies and can be used to detect or diagnose a variety of conditions. In these patients, computed tomography (CT) scans are commonly used to assess injuries, and the radiation dose associated with these scans is a concern.

"Each provider has to decide: What’s the benefit of getting the pictures that we get?" said Dr. Marissa A. Brunetti, an intensivist at Johns Hopkins Hospital, Baltimore.

"Especially with CT, if a patient is critical and the imaging study means the difference between life and death, then there are times when imaging studies are done out of convenience or in place of other imaging modalities [like ultrasound] that could get pictures that are similar without radiation exposure."

In a presentation at the annual meeting of the Pediatric Academic Societies, Dr. Brunetti reported on 729 patients aged 14 years and younger who underwent CT scans at the emergency department at her hospital over a 1-year period. Transfer patients were excluded from the analysis, as were any follow-up imaging studies. In total, 4,157 CT scans and 4,663 radiographic studies were conducted on these patients during that period.

The average radiation dose for each patient was 12.8 millisieverts (mSv), with a high of 73.5 mSv. "The average dose from the environment is about 3 mSv per year, so that’s more than four times the annual dose," said Dr. Brunetti.

Stratified by type of injury, the 178 patients whose trauma resulted from a motor vehicle accident registered the greatest radiation exposure, with an average of 18.6 mSv. Pedestrians struck by cars had the second highest level of radiation, 15.6 mSv.

Part of the reason possibly unnecessary imaging studies are done, said Dr. Brunetti, is that “pediatricians and providers don’t know the doses that these images impose. In the emergency department, and even in the trauma center, the calculations of the highest doses, and emphasis on keeping these studies to a minimum where possible, could lower patients’ total radiation exposure.

Another problem lies with transfer patients. Although the study did not look at transfers to the hospital, an audience member pointed out that “there is this idea that the trauma patient is going to die, and we need to do whatever it takes to save their life.”

Dr. Brunetti said that she had no disclosures or conflicts of interest to report.