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Clinical Observations

Recurrent Fat Embolic Strokes in a Patient With Duchenne Muscular Dystrophy With Long Bone Fractures and a Patent Foramen Ovale

Christopher J. Bugnitz MD *, Linda H. Cripe MD, Warren D. Lo MD, Kevin M. Flanigan MD

The Heart Center, Nationwide Children's Hospital, Columbus, Ohio

ABSTRACT

BACKGROUND: Individuals with Duchenne muscular dystrophy have an increased risk of long bone fractures. Such fractures are sometimes associated with brain dysfunction due to fat embolism syndrome, although this syndrome has seldom been documented in muscular dystrophy patients. **PATIENT DESCRIPTION:** We describe a child with Duchenne muscular dystrophy who developed fat embolism syndrome with neurological dysfunction following multiple long bone fractures. He experienced recurrent cerebral infarctions that probably resulted from embolization through a patent foramen ovale. The patent foramen ovale was closed by an occluder device in the cardiac catheterization laboratory, and he did not experience further infarctions. **CONCLUSIONS:** Fat embolism with ischemic cerebral infarction can occur in individuals with Duchenne muscular dystrophy following long bone fractures. In this setting it is important to identify and close atrial level shunts in order to prevent additional infarctions.

Keywords: Duchenne muscular dystrophy, fat embolism syndrome, atrial septal defect, patent foramen ovale, cardiomyopathy, stroke

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Introduction

Fat embolism syndrome is a potentially life-threatening complication of long bone or pelvic fractures. It can involve multiple organs, but the classic symptom triad of fat embolism syndrome includes respiratory distress, cerebral dysfunction, and petechial rash associated with thrombocytopenia.[1,2] The syndrome is rare in children, with a reported incidence that is 100 times less than in adults.[3] A recent analysis of data from the National Hospital Discharge Survey identified no cases of fat embolism syndrome in individuals less than ten years of age with isolated fractures but a rate of fat embolism syndrome of 0.37% in children aged ten to 19 years of age and adults aged 20 to 39 years with isolated fractures.[4] However, fat embolism syndrome has been identified as a rare but devastating cause of morbidity and mortality in individuals with Duchenne muscular dystrophy (DMD). To date, there have been three published series summarizing a total of 14 patients with DMD who developed fat embolism syndrome after minor trauma.[5-7] Fractures are common in individuals with DMD for a variety of reasons including the osteoporosis associated with severe neuromuscular disease.[5,6,8] Patients with DMD are at high risk for fractures during falls associated with daily care.

We present a boy with DMD who developed fat embolism syndrome with recurrent cerebral infarctions related to the presence of a persistent patent foramen ovale (PFO). Following closure of the PFO by an atrial septal occluder device, the child had no new embolic infarctions.

Patient Description

This 13-year-old nonambulatory boy with genetically confirmed DMD was in his usual state of health before this incident. He had never been treated with corticosteroids and had used a wheelchair full-time since age eight years. At his previous clinic visit, he was noted to

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* Communications should be addressed to: Dr. Bugnitz; The Heart Center, Nationwide Children's Hospital; 700 Children's Drive; Columbus, Ohio 43205-2664.

E-mail address: Christopher.Bugnitz@nationwidechildrens.org

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have 4/5 ankle plantarflexion; 3/5 strength in neck flexion, wrist flexion, and wrist extension; 2/5 strength in elbow flexion and extension and knee flexion; 1/5 strength in shoulder abduction, knee extension, and ankle dorsiflexion. Mild dextroscoliosis was noted. He did not use nocturnal ventilatory support but pulmonary function testing suggested a mildly restrictive pattern. Echocardiography and cardiac magnetic resonance imaging (MRI) one month before admission demonstrated normal function with a left ventricular ejection fraction of 69% and no atrial level shunting. A dual-energy x-ray absorptiometry scan at age 13 years showed a z score of -3 for the whole body bone mineral density.

Because he was unable to bear weight, he was lifted for transfer to the toilet. During one such transfer, he was dropped, resulting in immediate bilateral leg pain. His initial radiographs demonstrated fractures of his left proximal tibia, left distal femur, and right proximal tibia. Within hours of the injury, his mental status rapidly deteriorated. Upon arrival at our institution, he was minimally responsive with a Glasgow coma score of 9. Naloxone was given without response, and he was placed on biphasic positive airway pressure secondary to respiratory failure. His complete blood count demonstrated a leukocytosis. A chest radiograph and chest computed tomography (CT) scan showed no evidence of lung disease. A cranial CT scan did not show hemorrhage, midline shift, or mass effect.

He was admitted to the pediatric intensive care unit, where he was somnolent, lacked spontaneous eye opening, and exhibited no response to simple commands or purposeful movement. Global withdrawal to pain and local withdrawal to pain in the left arm, equally reactive pupils with slight constriction and sluggish response, somewhat disconjugate gaze, increased bilateral leg tone. He exhibited a left Babinski sign but otherwise had no apparent reflex asymmetry (albeit difficult to examine due to increased tone). The differential diagnosis at that time included fat embolism syndrome, thromboembolic stroke, seizures, and infectious encephalopathy.

His brain MRI showed numerous symmetric punctate foci of restricted diffusion within the centrum semiovale, corona radiata, caudates, putamina, thalami, and the pons and cerebellar white matter, consistent with emboli (Figure). These lesions were suggestive of fat embolization given the recent fracture. The initial transthoracic echocardiogram demonstrated no evidence of intracardiac vegetations or thrombosis. There was also no evidence of an intracardiac shunt by two-dimensional imaging or routine color Doppler. Right ventricular pressure was normal, and there was normal left ventricular systolic function. An electroencephalogram revealed diffuse background slowing but no epileptiform discharges.

On the second hospital day, his right hemiparesis acutely worsened and he experienced a two-minute seizure with clonic jerking of all extremities, bilateral eye flutter and squinting (right more than left), and lip smacking. It seemed to begin as right facial twitching and then generalized. He was given a loading dose of levetiracetam (1500 mg or approximately 20 mg/kg) and then started on twice-daily oral dosing (500 mg/dose). A epeat cranial MRI on hospital day four demonstrated new embolic infarctions within the subcortical white matter of the posterior left and right frontal lobes, left parietal lobe, and left temporal lobe (Figure). Doppler ultrasonology of his legs, abdomen, and neck did not detect deep vein thromboses. The new emboli on cranial MRI raised concern for the presence of an intracardiac shunt. A repeat transthoracic echocardiogram and contrast study with agitated saline revealed right to left shunting at the atrial level shunt consistent with a PFO. Because this atrial communication may have allowed emboli to shunt from the systemic venous to the systemic arterial circulation, his PFO was closed.

A 23-mm atrial septal occlude device (Cribiform Septal Occluder) was placed across the atrial septum without complication. He was

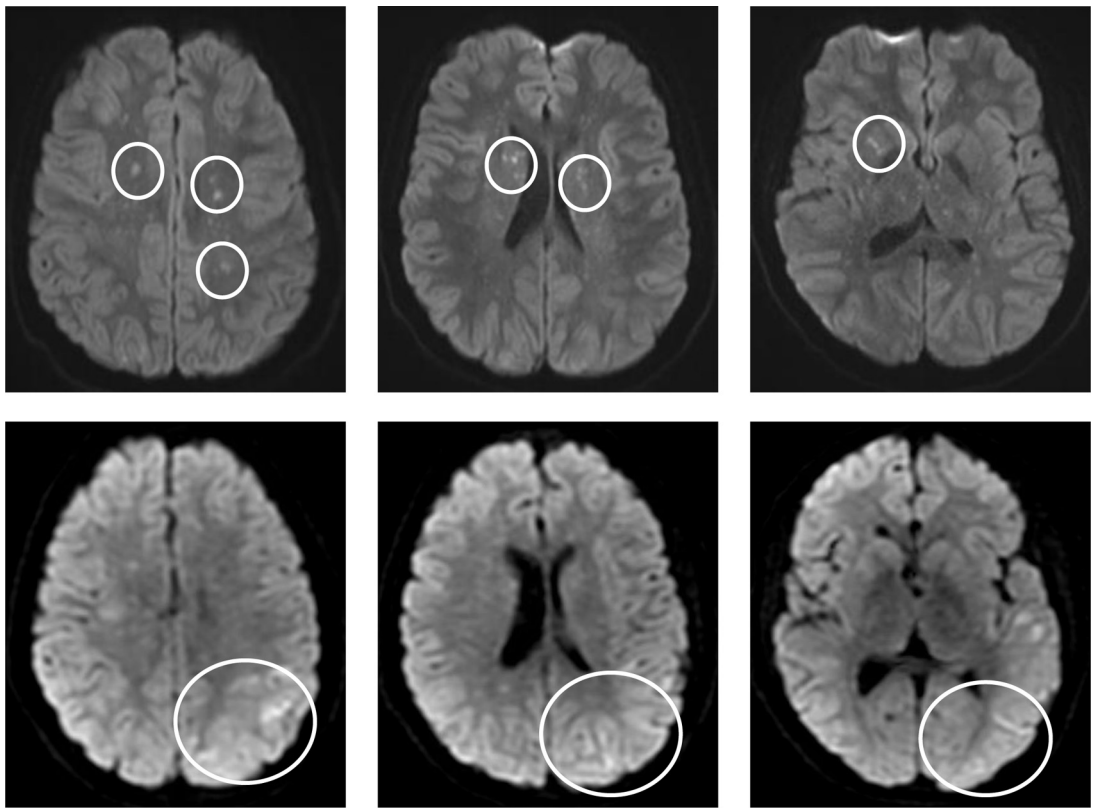

FIGURE.
(Top panel; initial MRI) Diffusion-weighted images demonstrate numerous punctate foci (white circles) of restricted diffusion symmetrically within the centrum semiovale, corona radiata, caudates, putamina, thalami, pons, and cerebellar white matter. (Bottom panel; four days later) Previously noted diffusion changes resolving. New areas of restricted diffusion (white circles) are present within the cortex and subcortical white matter of the left mid-temporal lobe, left parietal lobe, and left occipital lobe.