

Code Blue Sclera: A Girl with Osteogenesis Imperfecta

In a Hospital Room

Most decisions in medicine come down to weighing risks and benefits, as did the code I was involved with last week. From early in my medical training I was taught to heed the physician's doctrine, "Do no harm." And yet, if I had run a full code on this particular patient, I would have surely killed her.

This 2-year-old girl had been admitted to the hospital just a few days earlier. At that time, she had the most expressive eyes with faintly blue sclera and a look that cried out, "Love me strongly, help me softly." Whereas a child without osteogenesis imperfecta may have been better able to handle a simple respiratory virus, this patient clearly could not. Because of her inability to normally assemble collagen for her bones, ligaments, sclera, and teeth, she had countless fractures, skeletal deformity, and functional impairment. Moreover, she had many family members who loved her with all their might, but could not hug her with any might at all.

During my patient's early hospital course, her work of breathing was excessive and there was no way to effectively deliver chest physical therapy. I worried that even her coughing bouts might cause more fractures. She soon grew increasingly tired and then acutely lethargic. Remembering the ABCs of resuscitation, we brought her airway under control with a cautious and gentle but potentially fatal endotracheal intubation in order to mechanically provide positive pressure ventilation. However, she developed bradycardia, and shortly thereafter, as her blood pressure dropped to 0 mm Hg, a code was called.

Each person on the medical team at my patient's bedside had to summon up a huge degree of restraint. I had to hold myself back from manually forcing the blood to circulate around her little body. The code team was desperate to give chest compressions to a young girl whose thoracic cavity—already misshapen from innumerable rib fractures acquired over a short lifetime of delicate handling—would never sustain even the mildest of compressions. But the code team was acutely aware that just one chest compression for this patient would have been her last.

With maximal medical therapy, the patient rallied. Chest compressions were not given nor was defibrillation delivered. Receiving high-dose inotropics and pressors, she remains iatrogenically paralyzed with a neuromuscular blocking agent. At this point, I have yet to see what function she will recover, if she can ever be weaned off the cardiorespiratory support and extubated.

After the "code blue sclera," the code team watched the patient's mother convey her love by delicately stroking the fingers and short disfigured limbs of her daughter. There was, and still is, a lingering concern about how the next event will be handled by the code team and how this patient's friable bones in her frail body will handle the code team's next resuscitative efforts.

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