

EMERGENCY CARE POLICY AND A RARE CONDITION: THE IMPACT OF TECHNOLOGY



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After I testified at a Congressional subcommittee in support of the Emergency Medical Services for Children (EMS-C) reauthorization in June, a listener reached out to contact me. She had originally tuned in to the livestream to listen to a physician's testimony in support of another house bill, the "Newborn Screening Saves Lives" reauthorization act. This mother shared the story of her son Travis, who had isolated congenital asplenia (ICA). Having never heard of ICA I went to the website in her e-mail (www.team4travis.org) to learn more.¹ I had the opportunity to talk to this engaging mother who has risen above the loss of her own child to create an organization whose mission is to end asplenia mortality. The goals of T.E.A.M. 4 Travis are to increase awareness of ICA, collaborate with researchers to prevent ICA deaths, educate the medical and nursing communities about ICA, provide education and support to families with a child diagnosed with ICA, and to preserve memories of Travis' life.¹ Technology that would provide newborn screening and diagnosis of ICA is another focus of T.E.A.M. 4 Travis.

Children born with ICA have no spleen, a splenic remnant, or a nonfunctioning spleen, resulting in an immunocompromised child. These children are at significant risk for sepsis from infections most commonly caused by *Streptococcus pneumoniae*.² The incidence of ICA is not well

known, estimated at 1 in 2 million births.² However, ICA incidence may be underestimated. The diagnosis is often made postmortem. About 40% of the cases of ICA are caused by a genetic mutation of ribosomal protein SA. Identification of children with ICA is difficult as there is not currently a newborn screen for ICA. Would an ultrasound after birth to evaluate for the presence of a spleen be a viable technologic method to diagnose ICA? As emergency nurses, we are acutely aware of the importance of the spleen's role in immunity. Patients who have had a splenectomy are usually educated on the importance of sharing this important surgical history information with health care providers.

In the emergency department, especially a pediatric emergency department, the emergency nurse is cognizant of many chronic pediatric illnesses that require special and often emergent interventions. Parents of children with clinical conditions such as neutropenic fevers or long-chain acyl-coenzyme A dehydrogenase deficiency (LCAD) notify emergency department staff on arrival of their child's clinical condition to receive prompt, often lifesaving care. Protocols are in place in many emergency departments to facilitate the management of these and other acute and chronic pediatric emergencies.

Parents of children with ICA are often unaware of their child's condition. A very real concern is that a fever could be an ominous clinical presentation for the child with ICA. Emergency nurses can be desensitized to pediatric fevers, following protocols at triage for fever management while reassuring parents. In the child with ICA, a lack of knowledge regarding asplenia or splenic malfunction can be fatal. It is not possible today to determine at triage if the child with a fever has ICA, but with advances in technology ICA may be a condition that emergency nurses will have protocols to aggressively manage in the future.

Emergency department patients, especially children, often present with clinical conditions that are the result of a genetic mutation. Emergency nurses tend to focus on ferreting out life threatening clinical presentations and intervening to facilitate optimal outcomes. It is important for emergency nurses to consider that fever in a neonate or young child may require more aggressive interventions to reduce mortality from sepsis due to lack of an intact immune system in the presence of ICA.

ICA and the associated mortality from sepsis is a clinical issue we need to have knowledge of for optimal interventions and outcomes. Unfortunately, ICA is a

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life-threatening clinical condition that requires technology for early diagnosis to ensure correct management. Emergency nurses should consider collaboration with perinatal nurse colleagues to advocate for mandatory fetal anatomy scans after 20 weeks gestation and in neonates, which could identify ICA or other life-threatening anomalies. Supporting legislation that would include ICA in the mandatory newborn screening panel is another strategy to help diagnose this life-threatening condition.

I remain committed to the purpose of my original congressional testimony in June, reauthorization of the EMS-C act, which has helped improve pediatric emergency care across the United States. I learned so much that day from the other three experts testifying. Emergency nurses should support legislation that requires adding ICA to the

list of mandated screening tests for all newborns prior to their leaving the hospital. I am grateful to Travis' mom for taking the initiative to educate me about ICA. Emergency nurses are lifelong learners and if sharing what I learned about ICA makes a difference for even one child or family in the future, it would be beneficial.

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