Leclercia adecarboxylata Sepsis and Cerebral Herniation

Karen Sethi,1 Eric M. Barker,2 Leon A. Metlay,3 Mary T. Caserta,2 and Louis Eugene Daugherty1
Divisions of 1Critical Care and 2Infectious Diseases, Department of Pediatrics, and 3Department of Pathology, University of Rochester School of Medicine and Dentistry, New York

Corresponding Author: Karen Sethi, MD, 601 Elmwood Ave, Box 667, Rochester, NY 14642. E-mail: karen_sethi@urmc.rochester.edu

Received May 8, 2012; accepted September 13, 2012; electronically published January 27, 2013.

Leclercia adecarboxylata, a gram-negative bacillus of the Enterobacteriaceae family, is rarely identified as a pathogen in humans. We describe a fatal case of *L. adecarboxylata* sepsis in a child. This is the first reported pediatric death associated with infection due to *L. adecarboxylata*.

Key words. cerebral herniation; *Leclercia adecarboxylata*; sepsis.

*Leclercia adecarboxylata*, a motile gram-negative (GN) bacillus of the *Enterobacteriaceae* family, is a rare human pathogen [1]. We treated a child with *L. adecarboxylata* sepsis discovered post mortem. The patient ultimately died of a secondary complication due to disseminated intravascular coagulation with brainstem hemorrhage and resultant cerebral herniation. This is the first case reported in the literature a pediatric mortality associated with *L. adecarboxylata* infection in either immunocompromised or healthy individuals.

CASE REPORT

A 5-year-old boy with a significant past medical history of colonic neuropathy and pseudo-obstruction presented with a seizure to the emergency department, and the patient had a recent history of low grade fevers. This young boy had a history of constipation and severe reflux, which had been extensively evaluated since 1 year of age. He was diagnosed with intestinal inertia on the basis of intrinsic intestinal neuropathy. The patient’s status was post ileostomy repair due to prolapse with gastrostomy tube placement 18 months earlier. He had been receiving total parenteral nutrition and fluid replacement intermittently over the previous 2 years via an indwelling tunneled central catheter. The replacement fluid consisted of lactated ringsers (2 L/day intravenously) in addition to replacement for ostomy output (>2000 mL/day). He was treated daily with cephalexin, nitazoxanide, or amoxicillin/clavulanic acid, rotated every 2 weeks for small bowel bacterial overgrowth. Other pertinent past medical history included central line-associated blood stream infections with *Enterococcus* species and *Escherichia coli*.

On the day of admission, the patient was seen by his primary care provider for evaluation of persistent low grade fevers for 3 weeks. The child had been previously well other than a description of increasing ostomy output, increased clumsiness, and subjective reports of his “legs feeling funny” over a few days. While traveling home in the car, the child suffered a 2-minute, generalized tonic-clonic seizure and became unresponsive. The patient was brought to the emergency department where he was noted to be pale and was difficult to arouse with altered mental status (Glasgow Coma Score of 9). His pupils were equal, round, and reactive to light; his pulses were thready diffusely and he demonstrated poor perfusion with delayed capillary refill of 3–5 seconds. He was tachycardic with a heart rate of 140 beats per minute and had perioral cyanosis. His initial blood pressure was 56/32 mmHg; his oxygen blood saturation was 96%; and he had a temperature of 40.2°C, which was obtained via temporal artery thermometer. He was treated with oxygen and fluid resuscitation via his central venous catheter while peripheral intravenous access was obtained. Due to the unknown etiology of the fever and mental status changes, the patient was given ceftriaxone empirically after a blood culture was obtained from the central venous catheter. Laboratory values were significant for leukopenia, neutropenia, and mild dehydration.

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A chest radiograph showed mild increased vascular congestion. The patient was given lorazepam for possible subclinical seizure activity, after which he began vocalizing with more purposeful movements. He was noted to have additional seizure activity, and a neurology consultation was obtained. His examination was significant for 2-mm pupils bilaterally, which were sluggish to react. Corneal and gag reflexes were intact, and he was able to withdraw to pain in all extremities. There was no evidence of trauma.

As the patient was being prepared for imaging, he developed sonorous respirations with worsening perfusion, hypoxia, and hematemesis. He was intubated via rapid sequence induction with etomidate, succinylcholine, and midazolam, and he was noted to have a 90-second period of ventricular tachycardia that self-resolved. A computerized tomography scan demonstrated acute hemorrhage with blood in the lateral ventricles, third ventricle, quadrigeminal plate cistern, and fourth ventricle. The fourth ventricle was compressed and the temporal horns of the lateral ventricles were prominent, indicating hydrocephalus. There was effacement of the suprasellar cistern and herniation of the cerebellar tonsils. The patient was transferred to the pediatric intensive care unit (PICU), and neurosurgery evaluated him for possible ventriculostomy placement versus surgical decompression. While in the PICU, the patient became hypotensive and bradycardic and required an epinephrine infusion. His pupils were noted to be asymmetric, with the left pupil dilated in comparison to the right. He was treated with intravenous mannitol, dexamethasone, increased ventilation, and fluid resuscitation. He was also noted to have significant oozing with central line and arterial line placement.

The patient was emergently taken to the operating room for decompressive craniotomy due to clinical evidence of brainstem herniation. While in the operating room, the case was aborted because of multiple arrests requiring resuscitation, severe coagulopathy (INR 11.7), and grim prognosis. Upon return to the PICU, the patient developed pulseless electrical activity. Resuscitation was terminated per the mother’s request, given the poor chance of survival.

Autopsy demonstrated diffuse cerebral edema, subarachnoid hemorrhage, and cerebellar tonsillar herniation bilaterally. Widespread petechiae were noted on the internal organs, consistent with coagulopathy. A mitral valve abnormality was also noted on gross pathology (Figure 1), and the blood culture obtained from the central catheter in the emergency department grew L. adecarboxylata identified by a VITEK 2 automated microbiological system (BioMérieux, Inc, Durham, NC). In addition, post mortem quantitative blood cultures obtained from the inferior vena cava grew L. adecarboxylata, too numerous to count. Both isolates were sensitive to all antibiotics tested, including aminoglycosides, penicillins, carbapenems, first-, third-, and fourth-generation cephalosporins, quinolones, and trimethoprim sulfamethoxazole. At the time of illness, the child had completed a course of amoxicillin/clavulanic acid 4 days prior and by report was compliant with antibiotic medications.

**DISCUSSION**

*Leclercia adecarboxylata* is a member of the *Enterobacteriaceae* family, regarded as normal flora in the gut of animals, and has been isolated from human stool and environmental sources. It was first described by Leclerc [2] in 1962 as *Escherichia adecarboxylata* and formerly was known as Enteric group 41. It has been suggested that *L. adecarboxylata* is an opportunistic human pathogen that phenotypically resembles *E.coli*, and, when isolated, it is often part of a polymicrobial infection. This synergism may enhance the virulence of the organism [3]. In addition, *L. adecarboxylata* has been identified during episodes of bacteremia associated with centrally placed catheters [4, 5] in immunosuppressed hosts [6]. Sepsis, septic arthritis, diarrhea, peritonitis, and cholecystitis have also been described [7].

In our patient, the initial blood culture was positive only for *L. adecarboxylata* without evidence of coinfection. There is only 1 other case report of *L. adecarboxylata* infection in an otherwise immunocompetent host without coinfection [4].

To date, there are no reports in the literature of *L. adecarboxylata* infections associated with death. We postulate that this child’s death was due to sepsis with
**L. adecarboxylata.** We are unsure whether disseminated intravascular coagulation led to intracranial hemorrhage and herniation, or alternatively, if the brainstem hemorrhage and herniation were secondary to emboli from a mitral valve vegetation, as has been previously reported [8]. This patient did have predisposing risk factors for endocarditis, including a long-term central catheter, a poorly characterized gastrointestinal abnormality, and previous antibiotic use that may have selected for this opportunistic pathogen, as previously suggested by Myers et al [8], although the pathology from his valve was not specific for endocarditis due to limited sample. However, endocarditis seems unlikely given his isolate’s susceptibility to penicillins and the recent course of amoxicillin/clavulanate. Despite his history of chronic antibiotic use, the organism was sensitive to all antibiotics tested, in keeping with previous reports [9] but raising the question of medication adherence or the possibility that the *L. adecarboxylata* isolated in the blood culture was due to contamination. We do not think this is likely because both blood cultures obtained from the patient grew only this organism; the first culture before parenteral antibiotics were given and the second with a high organism burden. In addition, post mortem quantitative blood cultures grew *L. adecarboxylata*, too numerous to count, obtained as a central blood sample through a large bore needle in the inferior vena cava and not through the central line. As dictated by the University of Rochester microbiology laboratory protocol, any blood culture that flags positive and demonstrates GN bacilli is subsequently subcultured to blood, chocolate, and MacConkey agars. The organism from this patient grew out as lactose-fermenting, GN bacilli and was found to be indole positive. It was then set up in a BioMérieux VITEK 2 GN card (47 biochemicals). The additional key tests that were positive are malonate utilization, glucose and adonitol fermentation with confirmation of lactose fermentation. The key tests that were negative were ornithine and lysine decarboxylation, citrate, H2S production, and sorbitol fermentation. Each of the 4 isolates tested (pre and post mortem) had the same exact combination of results, and the margin of confidence of this identification is 99%.

Due to the limited information available regarding *L. adecarboxylata*, we believe it is important to describe this case to increase the awareness of this organism and expand the recognized clinical manifestations and potential severity of infection with *L. adecarboxylata*.

**Acknowledgments**

**Potential conflict of interest.** All authors: No reported conflicts.

All authors have submitted the ICMJE Form for Disclosure of Potential Conflicts of Interest. Conflicts that the editors consider relevant to the content of the manuscript have been disclosed.

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