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ARTICLE

Effectiveness of a Clinical Pathway for the Emergency Treatment of Patients With Inborn Errors of Metabolism

Dina J. Zand, MD[a,b], Kathleen M. Brown, MD[a,c], Uta Lichter-Konecki, MD, PD[a,b], Joyce K. Campbell, RN, MSN[d], Vesta Salehi, MD[e], James M. Chamberlain, MD[a,c]

[a]Department of Pediatrics, George Washington University School of Medicine and Health Sciences, Washington, DC; Divisions of [b]Genetics and Metabolism and [c]Emergency Medicine and [d]Center for Hospital-Based Specialties, Children's National Medical Center, Washington, DC; [e]Department of Pediatrics, Weill Cornell Medical College, New York, New York

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What's Known on This Subject

Prompt and effective care is crucial for patients with inborn errors of metabolism because delays may result in poor outcomes, including severe metabolic acidosis, hyperammonemia, cerebral edema, and death. High-energy infusions are used to prevent catabolism and complications.

What This Study Adds

Measures of timeliness and effectiveness showed improvement after implementation of an ED pathway for patients with inborn errors of metabolism. Therefore, a clinical pathway can improve the emergency care of patients with inborn errors of metabolism.

ABSTRACT

OBJECTIVE. The goal was to measure the effectiveness of a clinical pathway for the emergency department care of patients with inborn errors of metabolism.

METHODS. Two years after the implementation of a multidisciplinary clinical pathway for patients with inborn errors of metabolism in our urban, academic, pediatric emergency department, we compared measures of timeliness and effectiveness for patients treated before the pathway with the same measures for patients treated after implementation of the pathway. Measures of timeliness included time to room, time to doctor, time to glucose infusion, and total emergency department length of stay. Measures of clinical effectiveness included the proportion of patients receiving adequate glucose infusions, proportion of patients admitted, inpatient length of stay, and proportion of patients requiring PICU admission.

RESULTS. A total of 214 emergency department visits for patients with inborn errors of metabolism were analyzed, 90 before and 124 after initiation of the pathway. All measures of timeliness of care except total emergency department length of stay demonstrated significant improvement in comparisons of values before and after initiation of the pathway. Measures of clinical effectiveness also demonstrated significant improvements after initiation of the pathway. There was improvement in the proportion of patients who received adequate glucose infusions, with a decrease in the proportion of patients who required admission to the PICU. Emergency department length of stay, inpatient length of stay, and the proportion of patients admitted to the hospital were not affected.

CONCLUSIONS. Most measures of timeliness and 2 measures of effectiveness showed improvement after implementation of an emergency department pathway for patients with inborn errors of metabolism. Therefore, a clinical pathway can improve the emergency care of patients with inborn errors of metabolism. *Pediatrics* 2008;122:1191–1195

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Key Words
quality of health care, critical pathways, emergency medical services, inborn errors of metabolism

Abbreviations
ED— emergency department
IEM—inborn error of metabolism
PRISA II—Pediatric Risk of Admission II

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Address correspondence to James M. Chamberlain, MD, Division of Emergency Medicine, Children's National Medical Center, 111 Michigan Ave, NW, Washington, DC 20010. E-mail: jchamber@cnmc.org

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THE INSTITUTE OF Medicine has identified timeliness and effectiveness as 2 important components of quality health care.[1] Prompt effective care is crucial for patients with inborn errors of metabolism (IEMs) who present with seemingly minor illnesses, because delays may result in poor outcomes, including severe metabolic acidosis, hyperammonemia, cerebral edema, and death.[2–4]

Historically, the standard of care for emergency treatment of children with IEMs at our institution has been the provision of an emergency letter for the family to present to the emergency department (ED) at arrival. This summary contained the child's diagnosis and specific recommendations for immediate intervention, evaluation, and stabilization, as well as contact information for metabolic specialists. It has been our experience, however, that the delivery of timely care often is compromised by several family-related factors, including lost emergency letters, lack of communication by family members with pediatric metabolic staff members before arrival at the ED, and cultural and

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language barriers. ED factors such as unpredictable volume surges and resultant relative physician understaffing also contribute to delays. Furthermore, routine ED processes, such as obtaining a history and performing a physical examination, may delay the immediate administration of glucose to stop catabolism, which often is the most crucial immediate intervention.

In response to the observed delays, we implemented a multidisciplinary pathway in the ED to expedite the treatment of patients with IEMs. This pathway stresses early recognition, rapid nurse-initiated treatment with a high-energy dextrose infusion to prevent catabolism, and consultation with experts in metabolic diseases as soon as possible.

We performed the current study to measure the effectiveness of this clinical pathway for patients with IEMs. We hypothesized that the pathway would be associated with improvements in measures of the timeliness and effectiveness of ED care.

TABLE 1 **Patient Characteristics Before and After Implementation of the Metabolic Pathway**

	Before	After	*P*
Patients with amino acidopathies, *n*	27	48	
Patients with organic acidopathies, *n*	27	33	
Patients with fatty acid oxidation disorders, *n*	9	16	
Patients with urea cycle disorders, *n*	28	27	.32
Age, mean ± SD, mo	41.1 ± 132.1	73.0 ± 162.5	.13
Predicted probability of admission from PRISA II score, mean ± SD	0.32 ± 0.27	0.34 ± 0.28	.57
Blood urea nitrogen level, mean ± SD, mg/dL	10.9 ± 6.2	13.1 ± 7.3	.03[a]
Serum bicarbonate level, mean ± SD, mmol/L	20.8 ± 3.4	20.1 ± 3.5	.22

Diagnoses included maple syrup urine disease, isovaleric aciduria, ornithine transcarbamylase deficiency, citrullinemia, arginosuccinic aciduria, carnitine palmitoyltransferase deficiency, argininemia, propionic aciduria, methylmalonic aciduria, short-chain acyl-coenzyme A dehydrogenase deficiency, medium-chain acyl-coenzyme A dehydrogenase deficiency, and long-chain hydroxyacyl-coenzyme A dehydrogenase deficiency.

[a] Statistically significant difference.

METHODS

Setting

The study was performed at a large, inner-city, academic, freestanding, tertiary pediatric hospital. The ED sees >70 000 children per year and is the major referral center for the metropolitan region.

Development of the Pathway

The pathway was developed by a multidisciplinary team, including representatives from ED nursing, ED physicians, metabolic physicians, ED management, and ED informatics. By using the qualitative methods of process flowcharts and key informant interviews, the committee identified the following recurrent barriers to timeliness and effectiveness: (1) failure of families to bring the emergency letter, (2) language barriers, (3) family-related cultural barriers (eg, passive rather than aggressive personalities), (4) relative nonavailability of ED physicians because of attendance to other patients, (5) medical cultural barriers (concerns of ED nurses about calling non-ED physicians for consultations), and (6) nonavailability of appropriate intravenous fluids in the ED.

The committee implemented the following changes in processes to address these barriers: (1) identification of all patients with IEMs in the ED patient-tracking software, with a special flag visible at patient sign-in (which eliminated the need for the family to bring the medical summary and reduced the impact of language barriers); (2) immediate telephone contact from the metabolic physician on call to the ED attending physician when a family was directed to go to the ED; (3) education of IEM families by the metabolic team about the need for aggressive ED therapy and the need to speak up regarding any concerns about delays; (4) strict enrollment criteria, to promote initiation of the pathway by ED nurses without ED physician permission; (5) strict instructions for ED nurses to call the metabolic specialist at patient arrival; and (6) education and standard written instructions to facilitate ED nurse preparation of an initial 10% dextrose solution with appropriate electrolytes. Positive verbal feedback for both the initial telephone calls and adherence to the pathway was an important component in reinforcing the changes in patient management. In addition, e-mail was used to provide feedback to ED staff members when care was exceptionally good or when improvements were necessary.

Study Design

We performed an analysis comparing measures of timeliness and effectiveness before and after implementation of the pathway. The decision to study the effects of the pathway was made 2 years after implementation; therefore, all data were collected retrospectively, and ED staff members were unaware of the study. The study was limited to children with IEMs that placed them at high risk for catabolism, and the diagnoses included amino acidopathies, organic acidurias, fatty acid oxidation disorders, and urea cycle disorders (Table 1).

Data

Patient ED event times (ie, triage, brought to room, seen by physician, and disposition) were recorded through the ED tracking software (LogiCare 1.6.0.32; LogiCare, Eau Claire, WI). Treatment times, hospital length-of-stay, and other clinical characteristics were abstracted through chart review.

Outcomes

Measures of timeliness included the following time intervals: (1) time from triage to room (the time from initial assessment by the triage nurse to the time of placement in a treatment room), (2) time from triage to doctor (the time from initial assessment by the triage nurse to the time of initial assessment by any physician), (3) time from triage to glucose administration (the time from initial assessment by the triage nurse to the time of initiation of 10% dextrose infusion), and (4) ED length of stay (the time from initial assessment by the triage nurse to the time of either admission or discharge). For

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TABLE 2 **Measures of Timeliness and Effectiveness Before and After Implementation of the Metabolic Pathway**

	Before	After	*P*
Triage to room time, mean ± SD, min	11.4 ± 47.9	2.0 ± 8.9	.04[a]
Triage to doctor time, mean ± SD, min	37.8 ± 41.1	15.8 ± 20.9	<.001[a]
Triage to glucose time, mean ± SD, min	153.5 ± 89.0	96.5 ± 63.2	<.001[a]
ED length of stay, mean ± SD, h	5.9 ± 2.4	6.5 ± 3.2	.16
Inpatient length of stay, mean ± SD, h	96.9 ± 105.0	195.5 ± 100.2	.49
Proportion of patients receiving adequate dextrose infusion, %	63.3	87.1	<.001[a]
Proportion of patients admitted, %	62.2	68.5	.34
Proportion of patients admitted to PICU, %	16.7	7.3	.03[a]
Patients returning to the ED within 72 hours after initial evaluation, *n/N*	1/34	3/39	.37

[a] Statistically significant difference.

the purposes of this study, transfer to the observation unit for >12 hours was considered an admission.

Measures of clinical effectiveness included the proportion of patients with appropriate provision of high-energy dextrose infusion (10% dextrose solution administered at a rate ≥130% of the maintenance intravenous fluid rate), the proportion of patients admitted to the hospital, the inpatient length of stay, and the proportion of patients admitted to the PICU. For discharged patients, we compared the rates of unscheduled returns within 72 hours.

We used Pediatric Risk of Admission II (PRISA II) scores[5] to measure the severity of illness in the prepathway and postpathway groups. We also compared 2 components of the PRISA II scores, that is, the total serum bicarbonate level (a measure of acidosis) and the serum urea nitrogen level (a measure of dehydration), between the 2 groups. All analyses were performed by using SPSS for Windows 16 (SPSS, Chicago, IL). The study was exempt from institutional review board review because it involved review of extant medical records without the use of patient identifiers.

RESULTS

Data for 214 ED visits by patients with IEMs were analyzed, including 90 before and 124 after initiation of the pathway. Table 2 presents the characteristics of the 2 study groups. There were no significant differences between the groups with respect to age, illness severity (PRISA II scores), or acidosis (serum bicarbonate levels). Postpathway patients had higher mean serum urea nitrogen levels.

All measures of timeliness of care demonstrated significant improvement in comparisons of the prepathway and postpathway groups, except for the total ED length of stay (Fig 1). The time from triage to room improved from 11.4 minutes to 2 minutes, and the time from triage to doctor improved from 37.8 to 15.8 minutes. The time from triage to administration of dextrose improved from 153.5 to 96.5 minutes.

Measures of clinical effectiveness also demonstrated significant improvements after initiation of the pathway (Fig 2). There was improvement in the proportion of patients who received adequate glucose administration, with a decrease in the proportion of patients who required admission to the PICU. PICU admissions decreased from 16.7% to 7.8% (odds ratio: 3.24; 95% confidence interval: 1.33–7.8) after implementation of

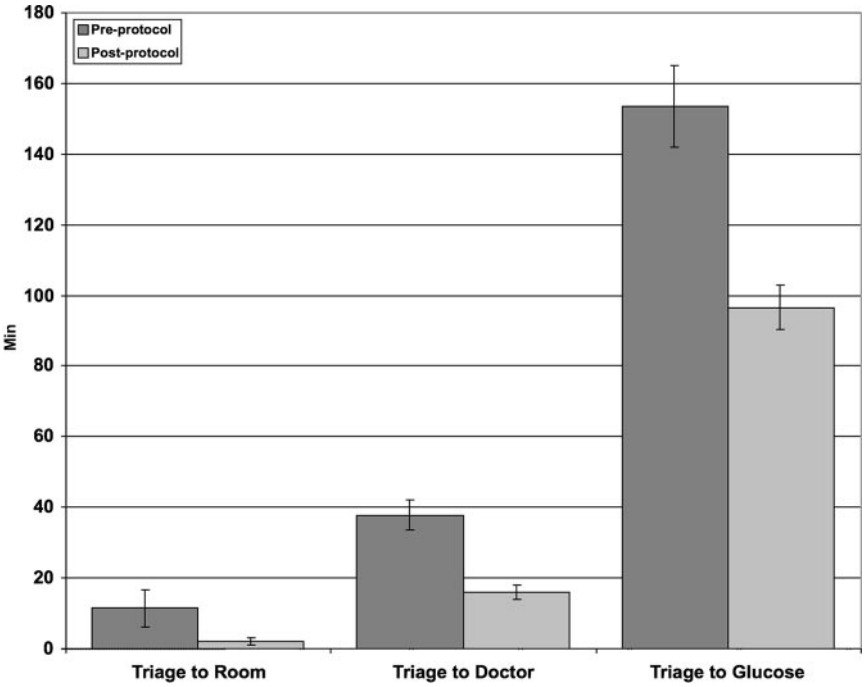

FIGURE 1
Measures of timeliness of care. Displayed are times from ED triage to placement in a room, from ED triage to evaluation by a doctor, and from ED triage to initiation of glucose therapy.

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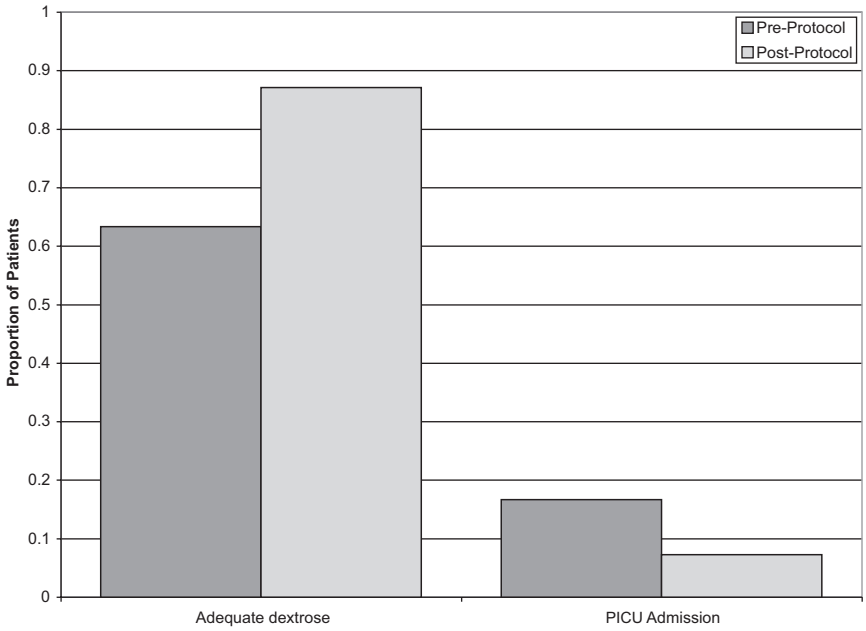

FIGURE 2
Measures of clinical effectiveness. Displayed are the proportion of admitted patients who received adequate dextrose infusions and the proportion of patients who required PICU admission.

the pathway. The severity analysis demonstrated that this decrease was not a result of patients being less ill at presentation; the PRISA II scores were similar in the 2 groups. In fact, the average serum urea nitrogen level was higher in the postpathway group. There was no difference in the proportion of patients admitted to the hospital, no difference in inpatient length of stay, and no difference in the proportion of discharged patients returning within 72 hours (Table 2).

DISCUSSION

This is the first study to demonstrate the effectiveness of a clinical pathway in improving timeliness and effectiveness of care for children with acute illness. Many clinical pathways were designed to improve efficiency by reducing lengths of inpatient stay and/or costs.[6–11] This is true for both adult[6,9,11] and pediatric[7,10] pathways. To our knowledge, the only study involving patients with IEMs measured the long-term cost-efficiency of newly implemented, expanded, newborn screening.[12] When clinical outcomes are measured, in many cases pathways do not affect[6,7,13–16] or adversely affect[11] patient outcomes. Some clinical pathways for outpatient illnesses have been associated with lower rates of hospitalization.[14,17] An ED-based asthma pathway improved process measures consistent with best practice, including less oxygen use, more metered dose inhaler with spacer use, and an increased proportion of patients receiving oral rather than intravenous corticosteroid treatment. Clinical outcomes were not measured.[18] An inpatient bronchiolitis pathway was associated with lower readmission rates and reduced rates of steroid use.[19]

Given the variable results achieved with previous studies of clinical pathways, we considered it necessary to study the effects of the pathway we created for patients with IEMs. It is important to measure >1 dimension of quality when assessing the effects of process changes. There may be trade-offs, for example, between efficiency and effectiveness. Attempts to improve one dimension may affect others adversely. We were able to demonstrate both improved timeliness and improved clinical effectiveness with the implementation of this pathway. Specifically, all measures of timeliness (arrival to room, arrival to doctor, and arrival to glucose administration) except ED length of stay were improved. Similarly, 2 measures of clinical effectiveness (the proportion of patients receiving adequate glucose infusion and the proportion of patients requiring PICU care) improved after implementation of the pathway.

A significant strength of this study was the ability to control for severity of illness in the before/after comparison. PRISA II has been validated nationally and shows good performance characteristics as a measure of severity of illness.[5] The probabilities of admission predicted by the PRISA II scores in the prepathway group were virtually identical to the probabilities in the postpathway group. Therefore, any differences in outcomes cannot be attributed to differences in baseline severity of illness.

This study has several limitations. First, the design was not a randomized, controlled trial. We think it would have been unethical to randomly assign patients to the nonpathway group. It is important to acknowledge this limitation, because there are often temporal improvements in other aspects of care that affect patient outcomes. We are responsible for the development of the care of patients with IEMs, in both inpatient units and the ED, and we are unaware of any concomitant changes in care. Furthermore, we tested for differences in baseline severity of illness in the comparison groups and we found no differences. Another limitation is that the study of a pathway of care does not allow us to draw inferences about which aspects of the pathway are responsible for improvements. For example, it is not clear from these data whether decreased times to seeing a

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physician represent improved efforts by physicians or empowerment of nurses to demand earlier physician involvement. Finally, the results achieved with the implementation of this pathway may not be achieved at other institutions. In principle, other institutions should be able to improve care delivery through careful analysis and reengineering of processes. However, the magnitude of improvement may vary because specific improvements are inherently unique at each institution. Therefore, pathways may not translate consistently when applied in different settings, and caution should be applied in attempts to generalize these results.[15,20]

CONCLUSIONS

Most measures of timeliness and 2 measures of effectiveness showed improvement after implementation of an ED pathway for patients with IEMs. Therefore, a clinical pathway can improve the emergency care of patients with IEMs. Further process redesign should be implemented and studied to achieve greater improvements in the timeliness of glucose delivery.

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