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HEMATOLOGY: RESEARCH ARTICLE

A proposed algorithm for evaluation and management of pediatric hemophilia patients who present to the emergency department with head trauma

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Abstract

Hemophilia is the deficiency of plasma clotting factor VIII (hemophilia A) or IX (hemophilia B) where management focuses on the prevention and treatment of acute bleeding symptoms and their sequelae. The most concerning risk is for life-threatening bleeding, including intracranial hemorrhage (ICH), which is caused by head trauma. Guidelines exist for the evaluation and management of pediatric head trauma, including the Pediatric Emergency Care Applied Research Network (PECARN) protocol, but limited evidence exists for when hemophilia patients present to the emergency department (ED), specifically with head trauma. Literature is limited regarding ICH and hemophilia, which further supports the culture of uncertainty among providers. The objective of this study is to conduct a retrospective chart review to determine the prevalence and clinical characteristics of ICH, and to describe computed tomography (CT) scan use in hemophilia patients who present to Phoenix Children's Hospital (PCH) ED with head trauma from January 1, 2007 to June 1, 2019. A total of 89 ED visits and 43 patients met inclusion criteria, and prevalence of ICH was determined to be 4% with the patients presenting with varied clinical characteristics and few commonalities. Using these data, we propose a new algorithm to aid clinicians in determining the need for CT scan in pediatric hemophilia patients who present to the ED with head trauma.

KEYWORDS
CT scan, head trauma, hemophilia, intracranial hemorrhage, traumatic brain injury

1 | INTRODUCTION

Hemophilia is caused by the deficiency of plasma-clotting factor VIII (hemophilia A) or IX (hemophilia B). It most often has an X-linked, recessive pattern of inheritance, but it can also occur as a spontaneous mutation. The estimated prevalence of hemophilia in the United States is 12 cases per 100,000 US males for hemophilia A and 3.7 cases per 100,000 US males for hemophilia B.[1,2] Currently, the number of people with hemophilia in the United States is estimated to be about 20,000, based on expected births and deaths since 1994, and may actually be an underestimation of the true rate.[3] Patients with moderate or severe disease may be diagnosed earlier in life due to known family history, whereas patients with mild disease may not present until older ages due to later discovery of bleeding symptoms. Disease severity is based on factor levels, where mild disease is defined as factor levels greater than 5% and less than 40%, moderate is factor levels 1%–5%, and

Abbreviations: AMS, altered mental status; ciTBI, clinically important traumatic brain injury; CT, computed tomography; ED, emergency department; EENT, ears, eyes, nose, and throat; GCS, Glasgow Coma Scale/Score; ICH, intracranial hemorrhage; LOC, loss of consciousness; MOI, mechanism of injury; PE, physical exam; PECARN, Pediatric Emergency Care Applied Research Network; ROS, review of systems; TBI, traumatic brain injury.

severe is less than 1%. Normal levels of factor VIII can range between 50% and 150%.[4] Clinical symptoms include bruising, hematomas, muscle, or joint bleeds, and bleeding after trauma or surgery can occur. Management of hemophilia primarily focuses on the prevention and treatment of acute bleeding symptoms and their sequelae. Most concerning is the risk for serious, life-threatening bleeding, which includes intracranial hemorrhage (ICH).

ICH can occur in pediatric patients secondary to vascular abnormalities, cardiac diseases, malignancies, bleeding disorders, and trauma whether accidental or non-accidental.[5,6] Patients with hemophilia have a higher risk of ICH both spontaneous and secondary to trauma. ICH rates in imaged hemophilia patients is approximately 1%; however, prevalence can range from 2% to 16% following trauma.[7,8] Risk factors for ICH in hemophilia patients include absent or inadequate factor prophylaxis, trauma, HIV, hepatitis C, presence of inhibitors, less than 3 years of age, hypertension, thrombocytopenia, and severe disease.[9–11]

Due to the increased risk in hemophilia patients, computed tomography (CT) scans are obtained more frequently. However, most CT imaging studies performed on patients with hemophilia for suspected ICH are normal.[12] CT imaging can be expensive, uncomfortable for the patient, and is not always readily available. It also exposes patients to increased radiation with a single head CT averaging 2 mGy/mSV (range 0.9–4) of radiation.[13] Further, the odds of children with bleeding disorders having a CT performed is 42-fold higher if they are less than 2 years old and 23-fold higher if they are older than 2 years.[14] Studies have shown that a cumulative dose of 50–60 mGy/mSv, specifically to the head, has a three-fold increase in brain tumors and bone marrow malignancies.[15] Crawford et al. found that 31.4% of patients with severe hemophilia accumulated high to very high levels of radiation exposure (>20 mSv) compared to 1.4% of controls with mild platelet function defect.[16] In an effort to find an alternate imaging modality, several studies have evaluated the effectiveness of fast magnetic resonance imaging (MRI) for ICH and traumatic brain injury (TBI); however, these studies have varied results and recommendations.[17–19]

The management of head trauma for hemophilia patients varies among hospitals and providers. Witmer et al. conducted a study investigating the clinical management of hemophilia and head trauma through self-reporting by pediatric hematology and oncology physicians. The study concluded that treatment of pediatric hemophilia patients who sustained mild head trauma is diverse, with over half of the providers using factor replacement and CT imaging for mild head trauma without signs or symptoms of ICH compared to the remaining providers who would not intervene or only give factor replacement.[20] Witmer et al. further suggests that the diversity in reported care of hemophilia patients is likely secondary to lack of studies that address the true risk of ICH after mild head trauma in patients with hemophilia.[20] Other investigators in search of guidance regarding management of head trauma have evaluated parent preference regarding CT or observation in their child with a head injury. Karpas et al. in their study concluded that of the 134 parents of children without hemophilia who participated, 40% preferred immediate CT, 57% preferred observation, and 3% did not indicate a preference, which suggests that even among parents there exists a divide on what is the proper clinical management.[21]

The Pediatric Emergency Care Applied Research Network (PECARN), which is a collaboration of pediatric emergency departments (ED) in the United States, has created a validated head trauma clinical protocol for pediatric patients who present within 24 hours of head trauma with a Glasgow Coma Scale/Score (GCS) of 14–15[14,22–24] (Figure 1). The goal of the protocol is to identify children at very low risk of clinically important traumatic brain injury (ciTBI) in whom a CT scan may be unnecessary, thereby decreasing radiation exposure in pediatric patients. Clinically important TBI is defined as death, neurosurgical intervention, intubation for >24 hours, or hospital admission greater than two nights secondary to TBI. The PECARN algorithm for pediatric head trauma assesses the patient's age, GCS, signs for basilar skull fracture, altered mental status (AMS), loss of consciousness (LOC), vomiting, severe headache, and mechanism of injury (MOI) to calculate the risk of the patient having ciTBI. PECARN conducted a further analysis of a subset of the study population with 230 patients with bleeding disorders (hemophilia, von Willebrand disease, congenital or acquired thrombocytopenia, functional platelet disorder, other bleeding disorder, or on anticoagulation therapy) from 25 centers from June 2004 to September 2006 and found that two patients had ICH with a prevalence less than 1%.[8] Although the PECARN protocol is beneficial for non-hemophilic population, it does not provide specific recommendations to identify low-risk hemophilia patients who present with head trauma.

Guidelines currently available from the National Hemophilia Foundation (MASAC #257) and the Children's Hospital of Philadelphia (CHOP) both provide recommendations on ED management of individuals with hemophilia.[25,26] The National Hemophilia Foundation (MASAC #257) details general recommendations for ED management of individuals with hemophilia and other bleeding disorders in respect to triaging patients, diagnostic studies, factor replacement, and treatment. The MASAC #257 recommends that in the case of head trauma or suspected ICH, clotting factor replacement therapy should be given before any diagnostic studies (X-rays, CT scans, etc.). It does not however provide specific guidance on whether or not CT imaging should be performed.[25] Further, the Children's Hospital of Philadelphia (CHOP) ED Clinical Pathway for the Evaluation and Treatment of Children with Hemophilia and Closed Head Injury, revised in December 2021, similarly provides guidance to providers for home and ED treatment based on factor levels, head trauma categorization, medical stability, and presence or absence of inhibitor. It does not delineate if a head CT should or should not be performed, but rather recommends "head CT and hematology consultation" once a stable patient with known presence or absence of inhibitor receives factor replacement. Nagel et al., in an effort to develop a protocol for head trauma and hemophilia, suggests one algorithm for when hemophilia patients present with unexplained headache (Figure S1). This algorithm, based on a systematic review of hemophilia patients 3–18 years old with suspected ICH, suggests getting CT imaging of all patients with unexplained headache except those with mild disease without trauma. Additionally, this article

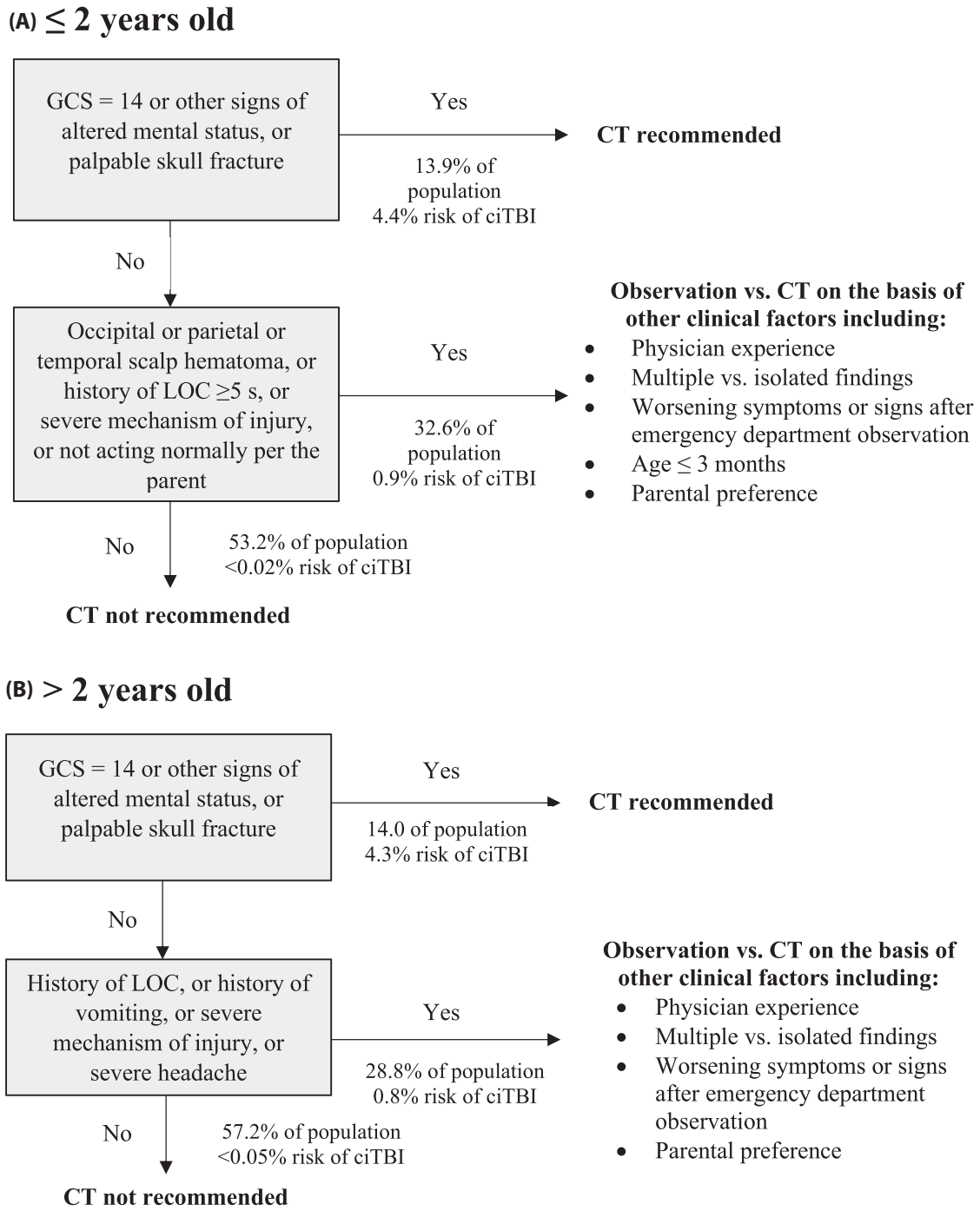

FIGURE 1 Pediatric Emergency Care Applied Research Network (PECARN)-validated algorithm to rule out the presence of clinically important traumatic brain injuries (ciTBI) and determine which patients do not need computed tomography (CT) imaging for children younger than 2 years (A) and for those 2 years and older (B) with Glasgow Coma Scale (GCS) 14 or 15 after head injury[13,21–23] (image from Kuppermann et al.[14])

highlights an important step in management of these patients, which is to treat with factor prior to CT imaging.

2 | METHODS

This study is a retrospective chart review of existing medical records dated between January 1, 2007 and June 1, 2019, at Phoenix Children's Hospital for patients diagnosed with hemophilia who present to the ED with head trauma. The objective of the study is to determine prevalence of ICH, to describe clinical characteristics of these patients, and to describe CT scan use. Patients with hemophilia who had an ED visit within this timeframe were identified, reviewed to ensure inclusion criteria were met, and specific data elements were extracted from the electronic medical record (Table S1). Inclusion criteria included male or female age 0–25 years, a diagnosis of hemophilia A or B, and at least one ED visit for head trauma or injury. Data elements extracted

TABLE 1 Clinical characteristics of emergency department (ED) visits: age, hemophilia type, prophylaxis status, and disease severity

Variable	Overall ($n = 89$)	ICH present ($n = 4$)	No ICH ($n = 85$)	p-Value
Age (months): mean [range]	48.66 [2–206]	8.25 [2–11]	50.56 [5–206]	<.0001
Hemophilia A	73 (82%)	3 (75%)	70 (82%)	.9999
Hemophilia B	16 (18%)	1 (25%)	15 (18%)	.9999
On prophylaxis	28 (32%)	1 (25%)	27 (32%)	.9999
Severity of hemophilia				
Mild	28 (31%)	2 (50%)	26 (31%)	.5940
Moderate	12 (13%)	0 (0%)	12 (14%)	
Severe	49 (55%)	2 (50%)	47 (55%)	

Note: Demonstrates distribution of clinical characteristics between overall population ($n = 89$), those with intracranial hemorrhage (ICH) present ($n = 4$), and those without an ICH ($n = 85$).

included severity of hemophilia and head trauma, chief complaint, MOI, PECARN criteria, pertinent history and physical exam (PE) findings, diagnostic results, prophylaxis and factor details, and clinical outcome.

All variables were summarized descriptively with either count (%) or mean (standard deviation), as appropriate, unless otherwise noted. Comparisons between ICH groups were analyzed using a Fisher's exact test for categorical variables as sample size in the ICH group was small, and for continuous variables with a two-sample independent *t*-test, when normality was achieved. The unit of analysis used was visit, as correlation between visits within a single person was expected to be minimal. Statistical analysis was done using R version 3.6.2 and statistical significance was set at .05 level.

3 | RESULTS

A total of 89 ED visits and 43 patients met inclusion criteria and had data elements extracted. Of the 89 ED visits, 73 (82%) had hemophilia A and 16 (18%) had hemophilia B. Regarding disease severity, 28 (31%) patients had mild hemophilia, 12 (13%) had moderate hemophilia, and 49 (55%) had severe hemophilia (Table 1).

Further clinical characteristics described included pertinent review of systems (ROS) related to head injury, patient vitals, and PE findings (Table 2). Nausea was the most common symptom in those without ICH. Of the four visits with an ICH present, symptoms reported were AMS, lethargy, and seizures. Clinically significant differences in weight and heart rate were attributed to the difference in age between those with and without ICH. A total of 53 ED visits (59%) reported a GCS and of those reported all were 15. Abnormalities in the PE were noted in categories of general, ears, eyes, nose, and throat (EENT), cardiac, and abdominal exam. General exam abnormalities included a child in distress and EENT abnormalities were bulging tympanic membrane, cerumen impaction, periorbital edema and ecchymosis, photophobia, rhinorrhea, mouth lacerations, and a chipped tooth. Cardiac exam abnormality was I/VI systolic ejection murmur, and abdominal exam abnormalities were stool palpated in bilateral lower left and right quadrants.

The three most common MOI included a collision with an object (23%), a fall from furniture (20%), and slipping/falling while in motion (16%) (Figure S2). Of the patients determined to have an ICH, two visits reported a fall from furniture, one reported a stumble or fall from the child's own height, and one reported a blow to head by an object.

Of the total 89 visits, 69 (78%) visits had a head CT performed, whereas 20 (22%) visits did not have a CT performed. Of the 43 patients, 22 (51%) patients were imaged at least once. It is notable that one patient was imaged seven times leading to an estimated radiation exposure of 14 mGy/mSv (Table S2).

Of the head CTs performed, five (6.25%) were read as abnormal, with four positives for an ICH (4.49%) and one positive for a nasal fracture. Clinical characteristics, medication management, and CT use for the four ED visits with ICH are described in Table 3. The mean age of those with ICH present was 8.25 months (range 2–11), with all patients being under the age of 1 year. Seventy-five percent of those with an ICH had hemophilia A, and the disease severity was split between mild and severe. At the time of the visit, only one patient was on scheduled daily prophylaxis, one had factor replacement prescribed as needed, and the other two patients had no prescribed medications. Three of the four patients had a known hemophilia status, whereas one patient was diagnosed following the specified ED visit. On ROS, few commonalities were found; only two patients had a GCS documented, and a common PE finding for those with an ICH included a scalp hematoma (50%). If PECARN were used and the patients were not diagnosed with hemophilia, it would have been recommended to CT one patient, observe versus CT for two patients, and no CT scan for the remaining patient. All patients with an ICH did have a CT performed, and three (75%) were given factor prior to CT. The patient who was not given factor medication, did not have their hemophilia diagnosis determined until later in their hospital stay.

4 | DISCUSSION

In this study, we determined the prevalence of ICH of patients with hemophilia following head trauma to be 4%, which is within the range of 2%–16%.[7,8] Clinical characteristics varied among the patient

TABLE 2 Clinical characteristics of emergency department (ED) visits: Review of systems (ROS), vitals and physical exam (PE)

Review of systems	ICH present (*N* = 4)	No ICH (*N* = 85)	*p*-Value
Nausea	0 (0%)	7 (8%)	.9999
Vomiting	0 (0%)	2 (2%)	.9999
Lethargy	1 (25%)	2 (2%)	.3006
Double vision	0 (0%)	2 (2%)	.9999
Seizures	1 (25%)	1 (1%)	.1569
Muscle weakness	0 (0%)	1 (1%)	.9999
Vitals			
Weight (kg)	7.97 (1.3)	19.89 (13.8)	<.0001
Heart rate	131 (4.2)	110 (22)	.0144
Respiratory rate	26 (8.5)	26 (7.9)	.9549
Blood pressure systolic	101 (18.4)	108 (14)	.7012
Blood pressure diastolic	69 (26)	67 (15)	.9291
Physical exam			
Glasgow Coma Scale (yes)	2 (50%)	50 (59%)	.9999
Head (abnormal)	2 (50%)	60 (71%)	.7498
Neuro (abnormal)	0 (0%)	2 (2%)	.9999
Musculoskeletal (abnormal)	1 (25%)	6 (7%)	.7247
Skin (abnormal)	1 (25%)	13 (15%)	.9999
Extra (abnormal)	1 (25%)	11 (13%)	.9999

Note: Demonstrates distribution of clinical characteristics between those with intracranial hemorrhage (ICH) present ($n = 4$) and those without an ICH ($n = 85$).

population with few commonalities between MOI, ROS, and PE findings, which further contributes to the uncertainty of the history and physical to help physicians clinically rule in or out the presence of an ICH. On PE, 62 (70%) of the 89 visits had an abnormal head exam, with 37 (60%) of those indicating the presence of a scalp hematoma. The most important clinical characteristic of the patients with an ICH was that all were under the age of 1 year. Younger children may be diagnosed more often with ICH due to an unknown hemophilia diagnosis, their new onset of mobility, not yet being on prophylaxis, and having less experienced caregivers who may not recognize the need for immediate medical attention. Of the total 89 visits, 20 (23%) did not have a head CT performed and 69 (78%) did. Twenty-two (51%) of our patients were scanned at least once. Risk is ultimately small with one CT performed, but the problem arises when children undergo multiple scans over a short span of developmentally critical time. For example, one patient was scanned seven times with all scans performed between the ages of 2–6 years, with an estimated accumulated radiation exposure of 14 mGy/mSv, which is 30% less than very high levels of radiation leading to malignancies.[16]

The PECARN protocol can be a potentially useful tool that could help clinicians discern those with hemophilia at low risk of ciTBI or ICH where CT imaging may be unnecessary, thereby decreasing radiation exposure. If the PECARN protocol recommendations were followed for the 89 ED visits/43 patients, one patient with ICH would have been missed. One alternative in patients with hemophilia and head trauma is to utilize PECARN recommendations in children older than 2 years and to obtain a CT scan in all patients younger than 2 years. By following this recommendation, all four patients with ICH would have been discovered (Figure 2).

Therefore, the proposed revised algorithm for patients with hemophilia who present to the ED after head trauma is described below (Figure 3). Essentially, if a patient with hemophilia presents to the ED with head trauma, they should immediately receive factor replacement. If the patient is under the age of 2 years, despite the presence or absence of history and PE findings, it is recommended that they undergo a CT scan to rule out an ICH. Conversely, if a patient with hemophilia and head trauma is between the ages of 2 and 18 years, then after factor replacement, it is recommended to follow PECARN. Patients with GCS less than or equal to 14, AMS, or signs of a basilar skull fracture require CT scans. If the patient experienced LOC, vomiting, severe MOI, or severe headache, then CT versus observation should be performed, depending on physician/parental preference. Other factors that may influence this preference for CT over observation would include if the patient were nonverbal, the trauma was unwitnessed, and the additional known risk factors for ICH in hemophilia patients: HIV, hepatitis C, presence of inhibitor, use of prophylaxis, and disease severity. In patients without any of these signs or symptoms, CT would not be required. This proposed algorithm is consistent with National Hemophilia Foundation (MASAC #257) and the CHOP clinical pathway for hemophilia and closed head

TABLE 3 Clinical details of emergency department (ED) visits with intracranial hemorrhage (ICH) after head trauma

	Patient identification number			
	9	**17**	**18**	**64**
Age (months)	11	2	9	11
Hemophilia A or B	B	A	A	A
Severity of hemophilia	MILD	SEV	SEV	MILD
Prophylaxis	No	No	Yes; daily	No
On PRN medication	No	Yes	No	No
Hemophilia status known	Yes	Yes	Yes	No
Inhibitor	No	No	No	No
Mechanism of injury (MOI)	Fall from height	Blow by object	Fall from furniture	Fall from furniture
PECARN recommendation if patient did not have a bleeding disorder	Yes	Observe vs. CT	No	Observe vs. CT
Review of systems (ROS)	+Altered mental status (AMS), lethargy, seizures	None	None	None
Glasgow Coma Scale (GCS)	15	–	15	–
Abnormal physical exam (PE)	None	L occiput hematoma	None	L parietal hematoma
CT performed	Yes	Yes	Yes	Yes
Factor administration	Yes, after CT	Yes, before CT	Yes, before CT	No
Factor medication specifics	Benefix	Advate	Advate	No
CT impression	Subdural hematoma	Subarachnoid hemorrhage	Small contusion parietal lobe	Epidural hematoma

Note: Illustrates clinical details of ED visits determined to have an ICH ($n = 4$) regarding age, hemophilia type, disease severity and status, prophylaxis, mechanism of injury (MOI), PECARN recommendation, review of system (ROS), Glasgow Coma Scale (GCS), abnormal physical exam (PE), if CT was performed with respective results, as well as timing and type of factor administered.

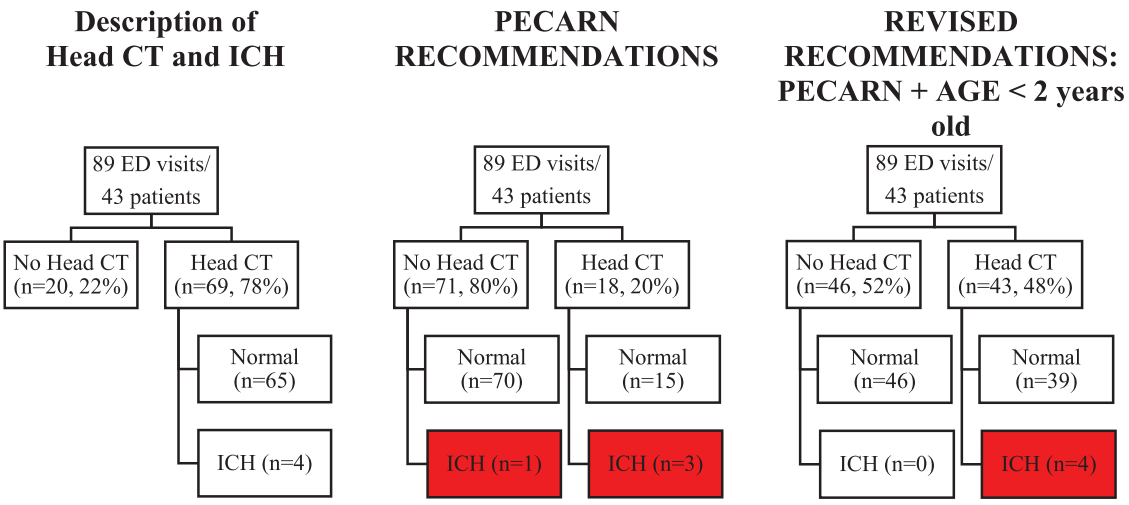

FIGURE 2 Comparison of data collected, Pediatric Emergency Care Applied Research Network (PECARN) recommendation, and revised recommendations for head computed tomography (CT) after head trauma for patients with hemophilia. Demonstrates number of head CT performed and intracranial hemorrhage (ICH) discovered if PECARN protocol recommendations were applied and under revised recommendations. Revised recommendations include imaging hemophilia patients if younger than 2 years old and following PECARN protocol if hemophilia patient is older than 2 years old. Under revised recommendations, all ED visits with ICH would be discovered, whereas under PECARN recommendations, one ED visit with ICH would have been missed

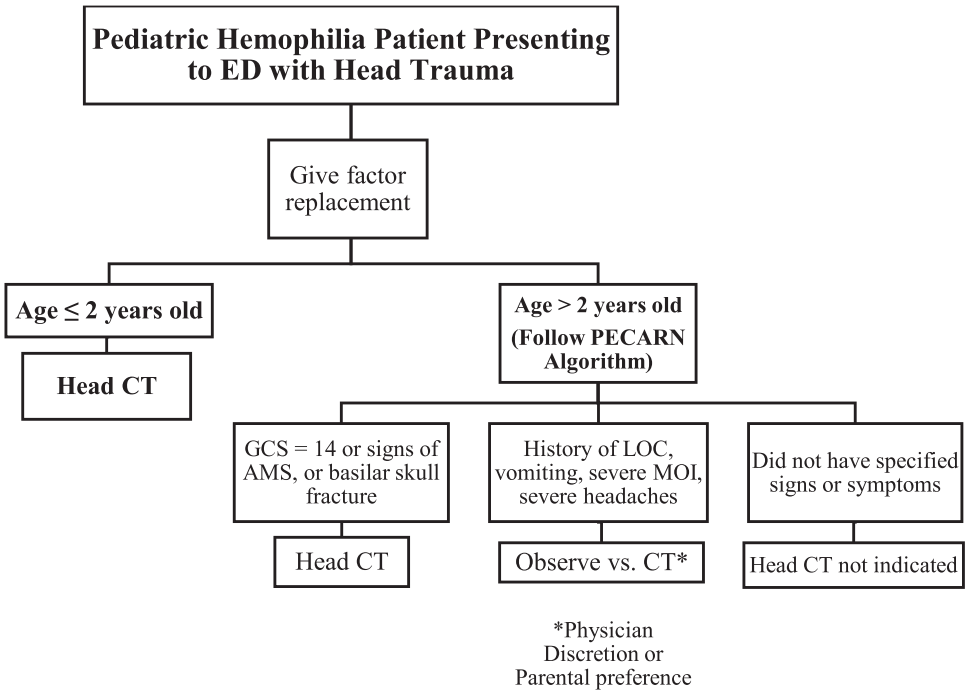

FIGURE 3 Proposed algorithm for when a patient with hemophilia presents to the emergency department (ED) following head trauma or injury. Illustrates revised algorithm for providers in the ED, where age is the first discerning factor after factor replacement. If a patient with hemophilia is less than 2 years old, then a head CT is performed, whereas if the patient is older than 2 years, the PECARN algorithm is followed

injury, where all patients with hemophilia who present to the ED with head trauma should receive factor replacement prior to diagnostic studies. This proposed algorithm would decrease the number of unnecessary CT scans for pediatric patients with hemophilia, and would provide more clarity and confidence for clinicians and parents alike when assessing the risk of an ICH after head trauma or injury.

In conclusion, the prevalence of ICH in patients with hemophilia who present to the ED with head trauma was determined to be 4%. Based on our findings, we propose an adaptation to the PECARN protocol for children less than 2 years of age, due to the higher risk of ICH in these young hemophilia patients. This new algorithm will help physicians decrease unnecessary radiation exposure to patients with hemophilia and head trauma, while still capturing life-threatening ICH.

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CONFLICT OF INTEREST

The authors have no conflicts of interest to declare. All co-authors have seen and agree with the contents of the manuscript and there is no financial interest to report.

DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available from the corresponding author upon reasonable request.

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SUPPORTING INFORMATION

Additional supporting information can be found online in the Supporting Information section at the end of this article.

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