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Increasing Observation Rates in Low Risk Pediatric Immune Thrombocytopenia Using a Standardized Clinical Assessment and Management Plan (SCAMP®)

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Abstract

An observational approach is recommended in newly diagnosed children with ITP at low risk of bleeding; however, there is no standard definition of risk. A SCAMP®, a modifiable practice guideline, was implemented and revised (SCAMP-1 and SCAMP-2) and applied to 71 newly diagnosed patients with ITP. The Buchanan and Adix bleeding score guided treatment and was modified by stratifying by low and high risk grade 3 bleeding in SCAMP-2. Observation rates increased from 40% to 74% from SCAMP-1 to SCAMP-2 ($p<0.05$) with no bleeding complications. We propose a modified bleeding score that increased observation in low risk patients with ITP.

Keywords

immune thrombocytopenia; pediatrics; guideline; treatment; bleeding

Introduction

Immune thrombocytopenia (ITP) is usually an acute, self-resolving illness in children. While severe bleeding, specifically intracranial hemorrhage (ICH), is the most feared complication, its incidence is rare. In observational cohorts, the reported incidence of severe bleeding in children is approximately 1/800 (1–3). It is not clear that pharmacologic therapy prevents ICH or other severe bleeding (4, 5). Both the American Society of Hematology (ASH) and an International Working Group (IWG) recommend pharmacologic treatment based on bleeding and observation without drug therapy in newly diagnosed patients who do not have severe bleeding (6,7). There are several bleeding severity instruments for ITP but no consensus regarding their use.

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The 2011 ASH ITP guidelines define “mild bleeding” as bruising or petechiae and recommend observation in patients with these symptoms (6). The IWG also recommends observation in such patients and subsequently proposed the ITP Bleeding Assessment Tool to define severe bleeding requiring treatment (7, 8). Other bleeding scales have been less clear about specific thresholds for treatment (9, 10).

The Buchanan and Adix bleeding scale provides an overall grade from 0–5 which incorporates skin, oral, and mucosal bleeding (11). Grade 0–2 includes bleeding of the skin only, grade 3 includes mucosal bleeding, and grade 4–5 is any bleeding that requires immediate medical attention or is life threatening (Table 1). Because it is quick, easy to apply, and has high inter-rater reliability for non-cutaneous bleeding, patients with ITP seen at our institution are assigned an overall Buchanan and Adix score by their primary hematologist (11).

A standardized clinical assessment and management plan (SCAMP®) is a modifiable practice guideline. An ITP specific SCAMP was developed by local expert consensus for newly diagnosed children with ITP and implemented at our institution (12). The SCAMP was designed to decrease practice variation, identify and learn from deviations in decision making, and decrease resource utilization by increasing observation rates in low risk patients. This report describes the outcomes of utilizing an ITP SCAMP at a single pediatric institution and proposes a modified Buchanan and Adix score that may facilitate employing an observation approach in patients with low risk of serious bleeding.

Methods

A SCAMP was developed by consensus of local ITP experts at Boston Children’s Hospital in 2012. Eligibility criteria for the SCAMP included: clinical diagnosis of ITP, platelet count $\leq 30 \times 10^9$ cells/uL, duration of thrombocytopenia < 4 weeks, no prior treatment, and age 1–16 years. All children seen in the Emergency Department or hematology clinic who met these eligibility criteria were followed with implementation of the SCAMP. Bleeding scores were assigned by a hematologist using the Buchanan and Adix scoring system. Scores were not confirmed by another provider, but scores were recorded for some patients by both an attending physician and a fellow or nurse practitioner. Patients exited the SCAMP after 6 months.

SCAMP-1 was in use from July 2012–April 2014 and recommended treatment with either prednisone or intravenous immunoglobulin (IVIG) in patients with a Buchanan and Adix score ≥grade 3. After data analysis, the SCAMP was revised in April 2014. SCAMP-2 differed by 1) dividing grade 3 bleeding into high and low risk categories, 2) recommending observation for low risk grade 3 bleeding, and 3) recommending steroid monotherapy as first line for ≥ high risk grade 3 bleeding. Examples of high risk grade 3 bleeding are shown in Table 1. SCAMP-2 was in use from April 2014–July 2015. While recommendations were provided, treatment was at the discretion of the provider, so deviations from the SCAMP were anticipated.

Categorical data were summarized with percentages while continuous data were summarized as median and range. Groups were compared using a Fisher's exact test or Wilcoxon rank sum test. A P value <0.05 was considered significant. Analysis was performed using SAS v9.4 (SAS Institute, Cary, NC).

Results

SCAMP-1 was applied to 40 patients and SCAMP-2 to 31 patients. Median age, platelet count, and bleeding scores at presentation were not significantly different among the two groups (Table 2). Initial observation rate in SCAMP-1 was 40% (16/40), while in SCAMP-2, observation increased significantly to 74% (23/31, $p<0.05$).

In patients with bleeding scores ≤ grade 2, the recommendation of observation without drug therapy was followed, 16/17 (94%) in SCAMP-1 and 19/20 (95%) in SCAMP-2. In SCAMP-1, 56% (23/41) of patients had grade 3 bleeding, all of whom received pharmacologic treatment with prednisone (22/23) or IVIG (1/23) as per SCAMP-1 recommendation. The SCAMP was then revised. Based on the lower than expected rate of observation, the modified grading system was created for grade 3 bleeding, and steroids were recommended rather than IVIG or steroids, given that our institution did not have equipoise about these initial treatments in SCAMP-1.

In SCAMP-2, 35% (11/31) of patients had grade 3 bleeding, 7 of whom had symptoms deemed low risk, for which the SCAMP recommended observation. Although 4 were observed, in the 3 others, the treating physician deviated from the guideline and treated with steroids or IVIG. All 4 patients with high risk grade 3 bleeding received steroids. More patients in SCAMP-1 had grade 3 bleeding than those in SCAMP-2. To explore whether the increased observation rate in SCAMP-2 was due only to overall decreased bleeding, the modified grading system of low and high risk grade 3 bleeding was retrospectively applied to the cohort of patients enrolled in SCAMP-1. Based on recorded signs and symptoms of bleeding, 74% (17/23) of patients were classified as low risk grade 3 bleeding and 26% (6/23) high risk. If the modified bleeding scale had been applied during SCAMP-1 and all low risk grade 3 patients were initially observed, the recommended observation rate would have substantially increased from 43% (17/40) to 85% (34/40).

Of the 39 patients initially observed in SCAMP-1 and SCAMP-2, 37/39 (95%) ultimately received no pharmacologic therapy and were observed at least until 6 months from initial diagnosis. One patient in SCAMP-1 was started on prednisone for low risk grade 3 bleeding, and one in SCAMP-2 received 6-mercaptopurine after developing autoimmune hemolytic anemia. There were no high risk grade 3 or grade 4 bleeding events in any patients.

Discussion

An ITP SCAMP, based on local expert consensus, was successfully implemented within a hematology program at a large academic center. SCAMP-1 did not increase the observation rate of newly diagnosed pediatric patients with ITP, which has historically been approximately 40% at our institution. Following the ASH guidelines with regard to treatment based on bleeding manifestations, there is agreement that grade 0–2 bleeding

warrants observation whereas pharmacologic treatment is indicated for grade 4–5 bleeding. However, appropriate management for grade 3 bleeding is controversial; thus, SCAMP-2 implemented a modified grading system which discriminated between relatively higher and lower risk grade 3 bleeding (13). This more precise characterization of grade 3 bleeding led to a significant and safe increase in the rate of observation.

Since SCAMP-2 was associated with less grade 3 bleeding than SCAMP-1, the modified Buchanan and Adix score was retrospectively applied to the cohort on SCAMP-1. By doing so, observation would have increased from 43% to 85% with 17 additional patients observed. Given this finding, it is unlikely that the increased observation rate using the modified Buchanan and Adix score was solely related to less grade 3 bleeding in SCAMP-2.

This study has several limitations. Since the SCAMP was implemented at a single institution, each version was applied to only a small cohort of patients. Furthermore, the impact of the modified Buchanan and Adix score is difficult to assess, because providers were not required to follow guidelines within the SCAMP concept.

The ITP SCAMP increased consistency and observation rates in the management of newly diagnosed ITP. A similar model could be applied at other institutions. We propose clinical use of a modified Buchanan and Adix scoring instrument, which stratifies low and high risk bleeding to different treatment approaches and might help to increase observation rates and diminish overall disease burden.

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Abbreviation Key

ITP	Immune Thrombocytopenia
SCAMP	Standardized Clinical Assessment and Management Plan
SCAMP-1	Standardized Clinical Assessment and Management Plan Version 1
SCAMP-2	Standardized Clinical Assessment and Management Plan Version 2
ICH	Intracranial hemorrhage
ASH	American Society of Hematology
IWG	International Working Group
IVIG	Intravenous Immunoglobulin

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TABLE 1

Modified Buchanan and Adix bleeding score, overall bleeding severity

Grade		
0	None	No new hemorrhage of any kind
1	Minor	Few petechiae (≤100 total) and/or ≤ 5 small bruises (≤ 3 cm diameter), no mucosal bleeding
2	Mild	Many petechiae (>100 total) and/or > 5 large bruises (>3 cm diameter)
3	***Low Risk* Moderate***	***Blood crusting in nares, painless oral purpura, oral/palatal petechiae, buccal purpura along molars only, mild epistaxis ≤ 5 minutes***
	High Risk* Moderate	***Epistaxis > 5 minutes, hematuria, hematochezia, painful oral purpura, significant menorrhagia***
4	Severe	Mucosal bleeding or suspected internal hemorrhage (brain, lung, muscle, joint, etc) that requires immediate medical attention or intervention
5	Life threatening/ Fatal	Documented intracranial hemorrhage or life threatening or fatal hemorrhage at any site

* Modification: Original Buchanan and Adix Grade 3/Moderate- overt mucosal bleeding (epistaxis, gum bleeding, oropharyngeal blood blisters, menorrhagia, gastrointestinal bleeding, etc) that does not require immediate medical attention or intervention

TABLE 2

Demographics, presentation, and outcome of patients

	SCAMP-1 N=40	SCAMP-2 N=31	p-value
Median Age in years (range)	3.8 (1.3–13.8)	7.8 (1.3–14.3)	0.06
Female (%)	18 (45%)	16 (52%)	0.63
Initial platelet count (10^9 cells/uL) (median, IQR)	7 (5– 10.5)	6 (4–12)	0.86
Initial Bleeding Score n (%)			0.093
≤Grade 2	17 (43%)	20 (65%)	
Grade 3	23 (56%)	11 (35%)	
SCAMP Recommended Observation pathway	17 (43%)	27 (81%)	0.0002
Observation Rate	16 (40%)	23 (74%)	0.007
Patients hospitalized within 6 months of diagnosis *	4/35 (11%)	2/25 (8%)	1.0
Reasons for Admission	Medication administration (2) Parental anxiety (1) Distance from the hospital (1)	Medication administration (1) Observation for bleeding (1)	

* Some patients exited the SCAMP earlier or were lost to follow up.