



Partial Splenectomy as a Definitive or Interim Solution for Young Children with Transfusion-dependent Hemolytic Anemia: Long-term Results from a 15-Year Cohort

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Abstract

Objectives To evaluate the long-term efficacy and safety of partial splenectomy (PS) as a definitive or interim spleen-preserving strategy in young children with transfusion-dependent congenital hemolytic anemia (HA).

Methods Patients with congenital HA who required frequent blood transfusions and underwent PS at the present center between 2010 and 2025 were included. The hemoglobin (Hb) level, reticulocyte count, and the need for transfusions (if any) during the follow-up period were noted. Ultrasonography of the splenic remnant was obtained at baseline, 1 mo, and 18 mo following the PS.

Results Fourteen children (median age: 23 mo; range: 7–40) underwent PS by laparotomy ($n=12$) or laparoscopy ($n=2$). Median follow-up after PS was 9 y (IQR: 4.8–10.5). Following PS, 8/14 (57%) patients became transfusion-free, with a median Hb of 10.3 g/dL at last follow-up. Five (36%) patients continued to require occasional transfusions and subsequently underwent total splenectomy after a median of 3 y; All of them became transfusion-free thereafter. No episode of mortality, sepsis, or thrombosis occurred.

Conclusions PS may serve as a definitive procedure in selected young children with congenital hemolytic anemia and as a bridge to later total splenectomy in others. This approach may be particularly relevant in resource-limited settings, where blood availability, transfusion safety, and the risk of transfusion-transmitted infections are concerning.

Keywords Congenital dyserythropoietic anemia · Glucose phosphate isomerase deficiency · Hereditary pyropoikilocytosis · Laparoscopic · Overwhelming post-splenectomy infection

Introduction

Splenectomy reduces hemolysis, increases hemoglobin (Hb) levels, and decreases transfusion requirements in selected patients with congenital hemolytic anemias (HA), including red-cell membranopathies, enzymopathies, and hemoglobinopathies. However, total splenectomy (TS) results in lifelong susceptibility to overwhelming post-splenectomy infection (OPSI) and is also associated with thromboembolic complications and pulmonary hypertension. Due to the perceived long-term risks of TS, there is increasing interest in partial splenectomy (PS). In children <5 y, where TS is avoided, PS may reduce or eliminate transfusion needs, thereby decreasing iron overload, transfusion-transmitted infections, and the logistical/financial challenges of repeated transfusions, without increasing the

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risk of post-splenectomy sepsis. The experience, however, is limited worldwide [1–3]. There is a lack of consensus on the volume of spleen to be left behind during surgery; the reported range is 5–25% [3, 4]. When a PS is performed, a subsequent TS may be indicated due to regrowth of the splenic remnant, resulting in worsening anemia. In this retrospective study, the authors report the outcome of PS in children with HA from a single center.

Material and Methods

The study was conducted in the Postgraduate Institute of Medical Education and Research, Chandigarh, India. Patients with congenital HA requiring frequent blood transfusions who underwent PS during 2010–2025 were included. HA was suspected from a suggestive clinical and laboratory profile. A tier-wise diagnostic approach was used to determine the etiology of HA. Tier 1 included a complete blood count with red cell indices, reticulocyte count, and a peripheral blood smear. Tier 2 included high-performance liquid chromatography, bone marrow examination, osmotic fragility testing, and the eosin-5'-maleimide binding test, selected according to the clinical and laboratory phenotype. Tier 3 comprised clinical exome sequencing when the diagnosis remained unresolved [5].

Hereditary xerocytosis was excluded in all patients, as splenectomy is contraindicated in this condition because of the risk of thromboembolic complications. The parents of patients were counseled regarding the two options: (a) regular blood transfusions until age 5, when a TS could be performed safely, or (b) a PS below 3 y as a definitive or an interim procedure to a TS later. All patients received prophylactic vaccination against pneumococcal, meningococcal, *Haemophilus influenzae* type b, typhoid, and seasonal influenza infections, in accordance with recommendations for TS. The duration of penicillin prophylaxis following PS was determined at the treating physician's discretion and was typically for five years. In the absence of definitive guidelines, post-splenectomy thromboprophylaxis with aspirin was initiated at the treating physician's discretion, generally at a platelet count exceeding 700 to $1,000 \times 10^9/\text{L}$. The Hb level, reticulocyte count, and the need for transfusions (if any) during the follow-up period were noted. Ultrasonography (USG) of the splenic remnant was obtained at baseline, 1 mo, and 18 mo following the PS. Spleen volume (mL) was calculated as for a prolate ellipsoid using the formula: length X width X height in cms X 0.523. Institutional ethics committee approval was obtained (INT/IEC/2018/000393), and a waiver of consent was granted, as the study involved retrospective data collection. A fixed volume of splenic remnant (10–20 mL) was preserved, irrespective of the initial

spleen size. Splenic volume exceeding 500 mL on USG was defined as massive splenomegaly. Concomitant cholecystectomy was performed in patients with cholelithiasis.

Final data analysis was done using SPSS v23 (IBM Corp.). Normality was assessed using the Kolmogorov–Smirnov and Shapiro–Wilk tests. Median (and interquartile range) were determined for numerical variables. The Mann–Whitney U test was used to assess the association between two groups of non-parametric numerical data.

Surgical Procedure: Abdominal access, whether by laparoscopic or open method, was similar to that of TS. After exposing the splenic vessels, either the upper or lower pole was selected for preservation based on vascular anatomy, and the short gastrics or the inferior polar vessels were left in continuity with the main vessel, respectively. The blood supply to all other parts was ligated near the spleen with sutures or harmonic shears. Following the demarcation of the devascularised segment, a 2–3 cm splenic remnant (approximately 10–20 mL) was preserved with maximal capsular conservation. Hemostasis of the cut surface was achieved using spray cautery and topical hemostatic agents. Lower-pole remnants were wrapped in omentum and fixed to the lateral abdominal wall with Vicryl to prevent kinking of the delicate blood supply. Upper-pole remnants remained supported by their gastric attachments.

Results

Fourteen patients underwent PS; 12 by laparotomy and 2 by laparoscopy. The median age was 23 mo (interquartile range [IQR]: 11.7–32.2). Diagnoses included pyruvate kinase deficiency (PKD) ($n=6$), hereditary spherocytosis (HS) ($n=3$), congenital dyserythropoietic anemia type 2 ($n=1$), hereditary pyropoikilocytosis ($n=1$), glucose phosphate isomerase deficiency ($n=1$), and uncertain etiology ($n=2$). Clinical exome sequencing was used to establish diagnoses in 9 of 14 patients, including PKD in 5 and hereditary pyropoikilocytosis, HS, congenital dyserythropoietic anemia type II, and glucose phosphate isomerase deficiency in 1 patient each.

The profile of the 14 patients is illustrated in Table 1. Indications for blood transfusion varied, with pre-transfusion Hb targets determined by the physician at the local healthcare facility and by the patient's signs and symptoms.

Before PS, the disease burden was substantial. The median interval between transfusions was 47 d (range: 2 wk to 4 mo), and the median number of transfusions before surgery was 11 (range: 2–30). The median follow-up duration after the PS was 9 y (IQR: 4.8–10.5). Following PS, the transfusion requirement resolved completely in 8 (57%) patients. Transfusion frequency decreased by more than 50% in five (36%)

Table 1 Profile of patients with hemolytic anemia who had a partial splenectomy

S. No.	Age (months) at PS, Sex	Liver and spleen below the costal margin (cms)	Reticulocyte count (%) (Measured, corrected)	Etiology of hemolytic anemia	Transfusion requirement prior to PS	Hb at the last follow-up (g/dL)	Follow-up duration (months) after PS	Response after surgery
1.	8, F	5, 6	11.9, 2.8	PKD	Two transfusions until 8 mo of age	8.2	156	Transfusion frequency reduced after PS. TS performed 4 ½ y after PS. Transfusion-free after TS.
2.	26, M	2, 3	3.2, 1.2	PKD	Transfusion q 1 mo, starting at 6 mo of age	9.8	126	Transfusion frequency reduced after PS. TS was performed 3 y after PS. Transfusion-free after TS.
3.	11, F	3, 1	2.4, 1.1	PKD	Transfusion q 1.5 mo, starting at 2.5 mo of age	8.6	102	Transfusion-free after PS
4.	22, M	4, 2	18, 2.4	PKD	Transfusion q 1.5 mo, starting at 2 mo of age. Had an exchange transfusion at birth for jaundice.	8.5	84	Transfusion-free after PS
5.	22, M	4, 2	12, 2.5	HS	Transfusion q 4 mo, starting at 7 mo of age	12.3	168	Transfusion-free after PS
6.	8, M	5, 4	17, 2.9	HS	Transfusion q 1 mo, starting at 1 mo of age. Had an exchange transfusion at birth for jaundice.	10.3	156	Transfusion-free after PS
7.	40, F	4, 5	4.9, 2.7	HS	Transfusion q 1.5 mo, starting at 1.5 mo of age	13	58	Transfusion-free after PS
8.	30, M	3, 2	3.3, 0.68	CDA type II	Transfusion q 2 mo, starting at 4 mo of age	8.4	108	Transfusion frequency reduced after PS. TS was performed 1 y after PS. Transfusion-free after TS.
9.	24, F	6, 10	1.5, 0.3	Undiagnosed	Transfusion q 1 mo, starting at 8 mo of age	12.2	112	Transfusion-free after PS
10.	36, M	3, 2	0.1, 0.02	Hereditary pyropoikilocytosis	Transfusion q 1 mo, starting at 1 mo of age. Had an exchange transfusion at birth for jaundice.	11.6	108	Failure of PS; continued to be transfusion dependent. TS was performed 6 y after PS. Transfusion-free after TS.
11.	18, F	2, nil	5.4, 2.16	PKD	Transfusion q 1.5 mo, starting at 2 mo of age	8.7	3	Transfusion-free after PS
12.	36, F	Nil, 1	20, 4	PKD	Transfusion q 1.5 mo, starting at 2 mo of age. Had an exchange transfusion at birth for jaundice.	10.2	108	Transfusion-free after PS

Table 1 (continued)

S. No.	Age (months) at PS, Sex	Liver and spleen below the costal margin (cms)	Reticulocyte count (%) (Measured, corrected)	Etiology of hemolytic anemia	Transfusion requirement prior to PS	Hb at the last follow-up (g/dL)	Follow-up duration (months) after PS	Response after surgery
13.	12, M	3, 1	0.72, 0.33	GPI deficiency	Transfusion q 1 mo, starting at 2 mo of age	9.2	5	Transfusion dependent after PS
14.	31, M	4, 3	2.1, 0.98	Undiagnosed	Transfusion q 15 d	7.8	38	Transfusion frequency reduced after PS. TS was performed 3 y after PS. Transfusion-free after TS.

CDA Congenital dyserythropoietic anemia; *F* Female; *GPI* Glucose phosphate isomerase; *Hb* Hemoglobin; *HS* Hereditary spherocytosis; *M* Male; *PKD* Pyruvate kinase deficiency; *PS* Partial splenectomy; *q* Every; *TS* Total splenectomy

Note: Patients No. 2 and 3 are siblings

patients, while 1 (7%) remained transfusion-dependent. Serial hematologic assessment demonstrated an early postoperative rise in Hb, with the median Hb increasing from 6.25 g/dL preoperatively to 10.15 g/dL at 1 mo. Thereafter, the median Hb in transfusion-free patients was 8.85 g/dL at 3 mo, 9.0 g/dL at 6 mo, 8.8 g/dL at 12 mo, 9.2 g/dL at 2 y, and 9.4 g/dL at 5 y, suggesting a durable reduction in hemolytic burden as illustrated in Fig. 1A. The median increase in hemoglobin from baseline is shown in Fig. 1B. The median Hb at the last follow-up visit of the 8 patients who were rendered transfusion-free after PS was 10.3 g/dL (IQR: 8.7–12.3). The median reticulocyte count decreased from 9.65% at baseline to 5.7% at 1 y following PS. It remained unchanged in PKD (10.7% preoperatively vs. 10.6% one year post-PS) but decreased in HS (12.0% vs. 5.3%). A subsequent TS was performed in 5 (36%) patients with a suboptimal response after age 5. TS successfully improved Hb and rendered all 5 patients transfusion-free. A cholecystectomy for cholelithiasis was performed in 2 patients; along with PS in one patient, and 1 y following PS in another. No mortality, episodes of sepsis, or thrombosis were recorded during the follow-up period. No procedure-related intraoperative or postoperative complications were observed following PS. However, the subsequent TS was technically challenging due to adhesions from the previous surgery.

The median baseline splenic volume was 318 mL (IQR: 226–835). Baseline splenic volume did not differ significantly between patients who subsequently required TS and those who did not ($p=0.61$). Follow-up splenic volumes measured with USG were available for 10 patients. Median splenic volumes at 1 and 18 mo post-PS were 16.5 mL (IQR: 5.5–27) and 41 mL (IQR: 35–113), respectively. The median splenic volume at 18 mo post-PS did not differ significantly between patients who subsequently required TS and those who did not ($p=0.52$). The median platelet count was $228.5 \times 10^9/L$ at baseline, $567 \times 10^9/L$ at 1 y after PS, and $978 \times 10^9/L$ at 1 y after TS.

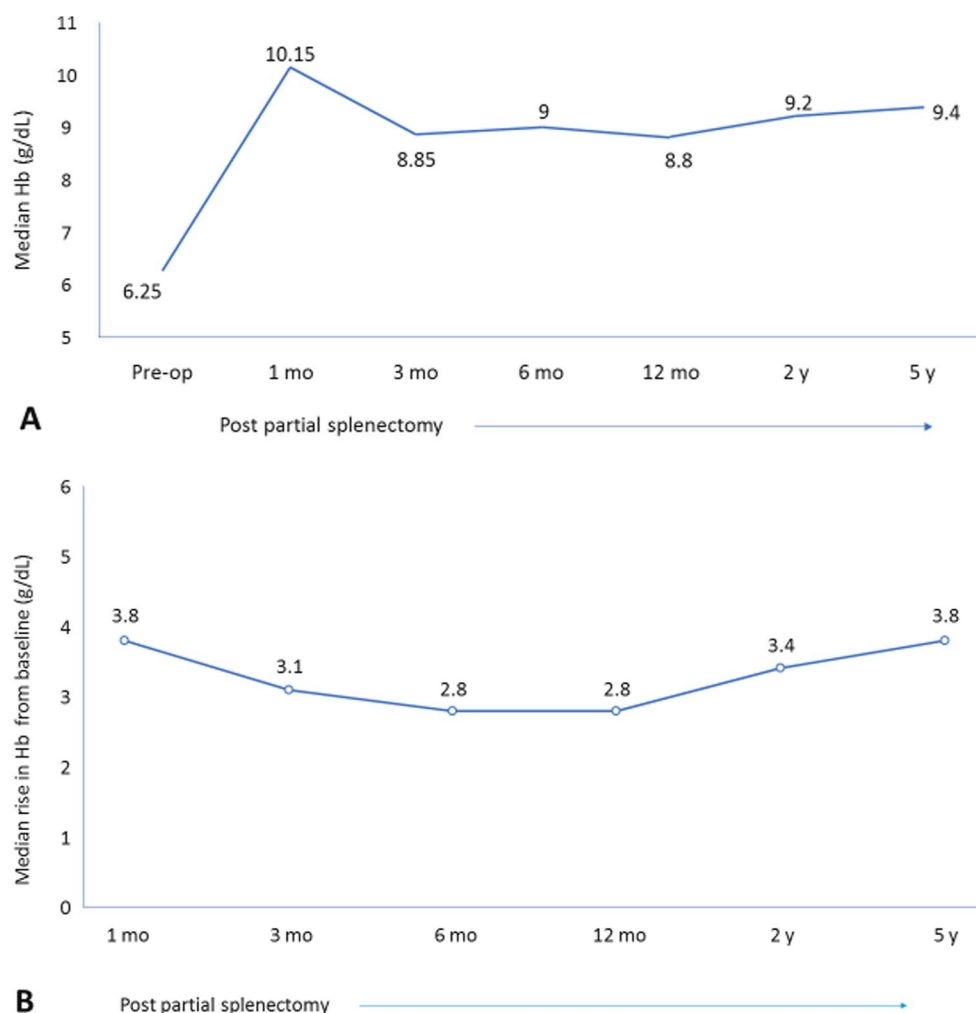
Discussion

TS has a small but definitive lifelong risk of OPSI despite immunizations [1, 2]. The risk doubles in children with comorbid conditions, including hematologic disorders, who are less than 5 y old [1, 2, 4]. In addition, TS is associated with thromboembolic events, enhanced arteriosclerosis, and late coronary artery disease. Given the late complications, PS has been considered, although it has not gained popularity. The potential benefits of PS include avoiding OPSI and late thromboembolic and vascular adverse effects while achieving the desired rise in Hb [6–11]. Guizzetti, in a meta-analysis, noted that PS increased Hb concentration by an average of 2.2 g/dL; the rise persisted at 5–6 y with no report of OPSI [6]. In another study across 16 sites in North America, none of the children undergoing PS for congenital HA experienced sepsis at 1-y follow-up, compared with 3 (4.2%) after TS [7].

The safety, effectiveness, and technical feasibility of PS across various HAs have been repeatedly demonstrated, even in the setting of massive splenomegaly [2, 12]. In the present study, three patients had massive splenomegaly. Although there was no difficulty performing the procedure, two patients later required TS.

The volume of splenic tissue to be left behind during surgery in PS is not well defined. Leaving behind a particular percentage of the original spleen would be inappropriate, as some spleens are very large, and a large remnant negates the purpose of the procedure [12]. The authors' experience has shown that leaving a fixed volume (10–20 mL) of the splenic remnant, irrespective of the original spleen size, gives optimal results. An intraoperative image of the enlarged spleen is illustrated in Fig. 2. Stoehr et al. also advised leaving a splenic core measuring 10 cm³ containing all three layers of the spleen for optimal results [13]. It is important to maintain a good blood supply to support phagocytic activity and prevent atrophy.

Fig. 1 (A) Trend in median hemoglobin levels before and after partial splenectomy among patients who remained transfusion-free following the procedure **(B)** Median rise in hemoglobin from baseline following partial splenectomy among patients who remained transfusion-free



The outcome of PS may depend on the etiology of HA. Although Rice et al. did not observe a difference, others have reported superior long-term outcomes in patients with HS compared with other congenital HAs [2, 7]. None of the patients with HS in the present cohort required subsequent TS after PS. Guizzetti et al. analyzed 14 observational studies of children under 18 y with HS, in which subsequent TS was performed in only 5% [6]. Buesing et al., in another multicenter study from North America between 1990 and 2008, noted that only 4.8% of patients with HS required subsequent TS over a long follow-up period [14]. On the other hand, Pincez et al. reported 21/79 (27%) patients with HS required subsequent TS at a mean duration of 8.4 y after PS [3]. They noted that with longer follow-up, the requirement of TS might increase. This was not observed in the present study or in that of Englum et al. [7]. Faster re-growth may be associated with hematologic relapse and transfusion dependency, and follow-up with USG is useful [9]. This may occur in the first 2 y following surgery, particularly in children under 2 y [3, 12, 13, 15]. Following PS, TS was required in 11/26 patients (42.3%)

with HS, with a mean interval of 7 y, in a French national, multicenter, retrospective study [16]. In a recent single-institution study of 31 patients undergoing PS for HS in the USA, 6 (19%) subsequently required TS after a median follow-up of 10.5 y [17].

In a meta-analysis of 5 studies involving children with HS, hematological outcomes in the early postoperative period were more robust with TS than with PS, whereas in late follow-up, there was no difference between the two [18]. The chief benefit of PS in very young children predisposed to OPSI is potentially avoiding transfusions, iron overload, and evading or delaying TS. In addition, PS is particularly relevant to low- and middle-income countries, where blood availability and transfusion safety remain significant concerns. The lack of universal nucleic acid testing of blood increases the risk of transfusion-transmitted infections. Parents who opted for PS did so mainly as regular transfusions were a logistical challenge, besides the accompanying adverse effects. In the present cohort, PS helped to avoid the need for transfusions in very young children who were not candidates for TS.

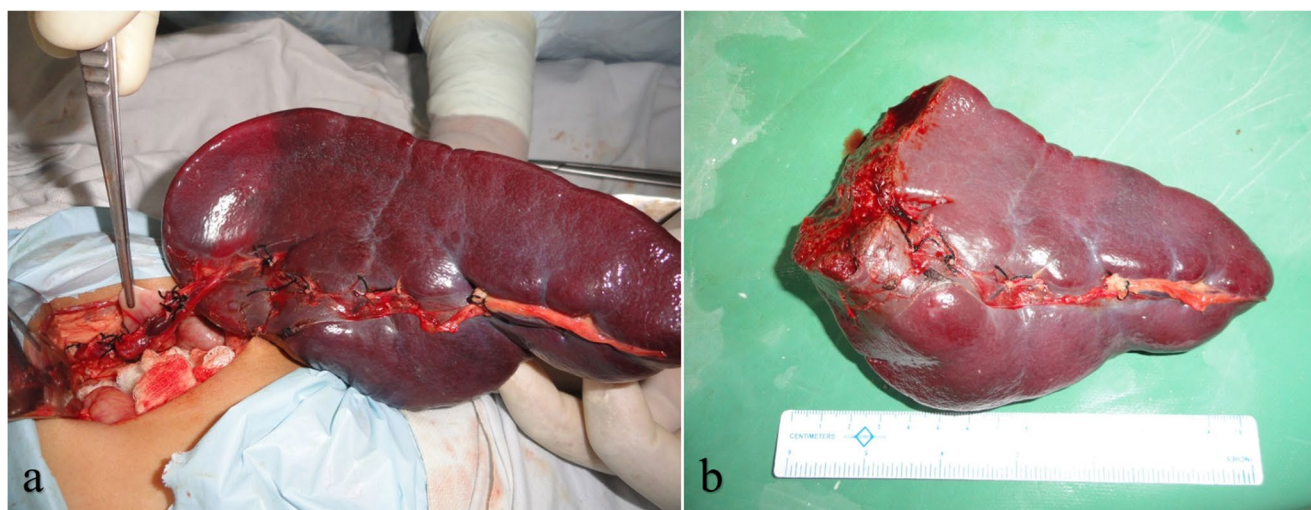


Fig. 2 (a) Intraoperative image obtained during laparotomy showing a congested, enlarged spleen in a 2-y-old child with pyruvate kinase deficiency undergoing partial splenectomy (b) Postoperative specimen

The notable feature of the present cohort is the young age of the patients. The median age of the cohort is 23 mo (all less than 3 – 1/2 y old), as compared with 5.3 y [16] and 9.7 y [17] in the previously published larger series. Infants and toddlers with severe transfusion-dependent hemolysis occupy a difficult therapeutic space in which continued transfusion is undesirable, but definitive TS is typically avoided or considered contraindicated due to the risk of OPSI. In this series, PS functioned as a definitive surgery in roughly two-thirds of children and as a bridge to later TS in the remainder, thereby shifting the timing of total splenic removal out of the period of greatest infectious concern. In the present study, PS was found to be safe in young (< 3-y) patients even in the presence of massive splenomegaly.

Another notable finding is the heterogeneity of diagnoses in this study. Published PS experience is dominated by HS-focused cohorts [3, 6, 7, 9, 13, 14, 16–18]. The current cohort had six patients with PKD, of whom four were rendered transfusion-free post-PS. Transfusion burden was reduced in other rarer diagnoses, including glucose-6-phosphate isomerase deficiency and congenital dyserythropoietic anemia II, as well, though TS was required later.

Limitations of the present study include the retrospective design, limited cohort size, and single-center experience. The hematological response following TS is reported to be more robust than that following PS [6, 7, 18]. However, the potential benefits of PS include retaining functional splenic tissue to avoid OPSI and late vascular complications. A second surgical procedure for TS will be required in a proportion of patients (36% in the present study) in whom PS is unable to maintain Hb levels. As most of patients in the present study were under 3 y old, their young age may have

showing the resected portion of the spleen; the upper pole has been preserved in situ

contributed to greater-than-expected growth of the splenic remnant compared with older children.

Conclusions

PS represents a valuable therapeutic option in selected young children with congenital HA requiring splenectomy, serving as a definitive procedure in most patients and as a bridge to later TS in the remainder. This approach may be particularly relevant in resource-limited settings, where blood availability, transfusion safety, and the risk of transfusion-transmitted infections remain important concerns.

Authors' Contributions PM and DB: Conceptualized the study and collected the data; MJ, SP, RJ, RD, and AT: Collected the data. NAM: Performed the analysis and drafted the manuscript; DB: Critically revised the manuscript. DB will be the guarantor for this manuscript.

Declarations

Conflict of Interest None.

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