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Splenectomy in Biliary Atresia Patients with Recurrent Jaundice Following Partial Splenic Embolization

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Ando, H., Ito, T., Seo, T., Ito, F. and Kaneko, K. *Splenectomy in Biliary Atresia Patients with Recurrent Jaundice Following Partial Splenic Embolization.* Tohoku J. Exp. Med., 1997, **181** (1), 167-174 —— Splenectomy was performed for three patients with biliary atresia because of re-exacerbation of their jaundice following treatment by partial splenic embolization (PSE). The subjects' red blood cell count and hemoglobin, serum level of hepatic enzymes (glutamic oxaloacetic transaminase, glutamic pyruvic transaminase, gamma-glutamyl transpeptidase, alkaline phosphatase, and lactic dehydrogenase), and total bilirubin (TB) were evaluated both before and after splenectomy in order to analyze the effects of splenectomy on these patients. The TB decreased significantly within 3 months after splenectomy in all three patients (13.0 ± 1.6 mg/100 ml to 5.4 ± 0.3 mg/100 ml, $p < 0.05$). The red blood cell count and hemoglobin increased gradually. There was a statistically significant correlation between the TB and the red blood cell count, and/or concentration of hemoglobin. The hepatic enzymes after splenectomy were not significantly different from those before splenectomy. The change in TB following splenectomy was essentially similar to that following PSE. These results suggested that the postoperative improvement in jaundice following splenectomy may not be due to improved hepatic function but merely a reflection of decreased red blood cell turnover. Splenectomy is a useful palliative procedure for jaundice in patients with biliary atresia for whom PSE is no longer effective. —— biliary atresia; splenectomy; partial splenic embolization; jaundice; spleen

Kasai procedure (Kasai and Suzuki 1959) has brought about a marked improvement in both the survival rate and cure rate for patients with biliary atresia (Ohi and Ibrahim 1992). However, many patients continue to have mild to moderate hepatic dysfunction (Toyosaka et al. 1993). These patients with jaundice may not achieve a long-term survival because their liver function will

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continue to worsen and they will eventually die of hepatic decompensation (Otte et al. 1994). These patients should be considered for liver transplantation, but this is not a feasible method with which to treat all patients with this problem (Lin et al. 1992). A complete cure is not possible for these jaundiced patients and quality of life issues become paramount. We previously have reported the effects of partial splenic embolization (PSE) on biliary atresia patients with persistent jaundice (Ando et al. 1996). The purpose of this communication is to report our experience with splenectomy on biliary atresia patients who had a recurrence of jaundice several years after PSE.

Patients and Methods

A splenectomy was performed on three patients with biliary atresia who had had the Kasai procedure and were later treated by PSE for jaundice. The clinical profile of these patients before splenectomy is summarized in Table 1. They underwent hepatic portoenterostomy with a stoma (Suruga II modification) at ages 54, 73, and 66 days. Postoperative serum total bilirubin (TB) decreased completely in two patients (patient No. 2 and 3) but not in one. PSE according to the method of Spigos (Spigos et al. 1979) was performed at ages 2, 5, and 13 years because of progressive jaundice and pancytopenia. PSE was effective but the palliative effect was short lived (Fig. 1).

The pneumococcal vaccine was given prior to splenectomy. Angiographic splenic arterial embolization was performed pre-operatively in the operating room using fluoroscopy in order to reduce the size of the enlarged spleen. Then the operative procedure was begun with a left subcostal incision. The splenic artery was ligated on the upper border of the pancreas and the spleen was then mobilized from its ligamentous attachment. Severe adhesions between the spleen and the lateral abdominal wall were found in all three patients, probably secondary to the localized splenic necrosis caused by the previous PSE. Dilated veins which developed between the portal vein and the systemic veins around the spleen were ligated carefully.

Table 1. *Clinical characteristics of the patients before splenectomy*

	Patient No.		
	1	2	3
Gender	Female	Male	Female
Age at the Kasai procedure (days)	54	73	66
Age at PSE (year)	2	5	13
Age at splenectomy (year)	5	9	20
Red blood cell count ($\times 10^4/mm^3$)	236	375	240
Concentration of hemoglobin (g/100 ml)	9.0	10.5	9.0
Serum bilirubin concentration (mg/100 ml)	9.9	15.2	13.8

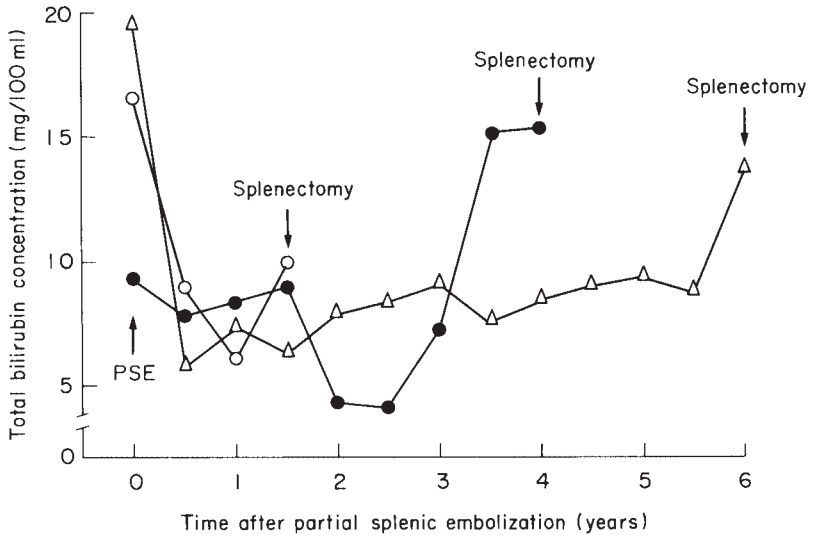

Fig. 1. Changes in the total bilirubin concentration after partial splenic embolization (PSE). ○ patient 1; ● patient 2; △ patient 3.

White blood cell (WBC), platelet, and red blood cell (RBC) counts, concentration of hemoglobin (Hb), serum glutamic oxaloacetic transaminase (GOT), serum glutamic pyruvic transaminase (GPT), gamma-glutamyl transpeptidase (g-GTP), alkaline phosphatase (ALP), lactic dehydrogenase (LDH), TB, direct bilirubin (DB), and indirect bilirubin (IB) were evaluated 1, 2, 3, 4, 5, and 6 months prior to splenectomy and 1, 2, 3, 4, 5, 6, 7, and 8 months following splenectomy.

Statistical analysis was performed for TB, RBC count, and Hb using the one tailed *t*-test for the comparison of values before 1 month and after splenectomy. An unpaired t test was used for the other parameters for the comparisons of values before and after splenectomy. Spearman's rank correlation was used for the comparison of TB and RBC count or Hb. Significance was defined as a p value <0.05. Results are expressed as the mean $\pm$ S.E.

Results

The clinical profile of the patients after splenectomy is summarized in Table 2. The spleens were grayish and bunchy, and weighed 457 g on average (Fig. 2). Congestion of the spleen was the most significant abnormal finding. Postoperative complications encountered were massive ascites in patient No. 3, which was controlled with diuretics, and hemorrhage from esophageal varices in patients Nos. 1 and 2 after 3 months, which was controlled by endoscopic injection sclerotherapy. One patient (No. 1) fell into a shock-like state with hypotension and unconsciousness secondary to middle otitis 7 months after splenectomy. She recovered with supportive treatment. All three patients regained their appetites and returned to active lifestyles.

The TB decreased significantly within 3 months after surgery. The time

TABLE 2. *Clinical characteristics of the patients after splenectomy*

	Patient No.		
	1	2	3
Operative time (hr)	5.42	4.25	6.42
Blood loss during surgery (g)	435	812	4050
Weight of the resected spleen (g)	430	420	520
Red blood cell count ($\times 10^4/mm^3$)	376	475	323
Concentration of hemoglobin (g/100 ml)	10.8	10.6	11.4
Serum bilirubin concentration (mg/100 ml)	4.8	5.7	5.5
Postoperative complications	Variceal bleeding, Serious infection	Variceal bleeding	Massive ascites

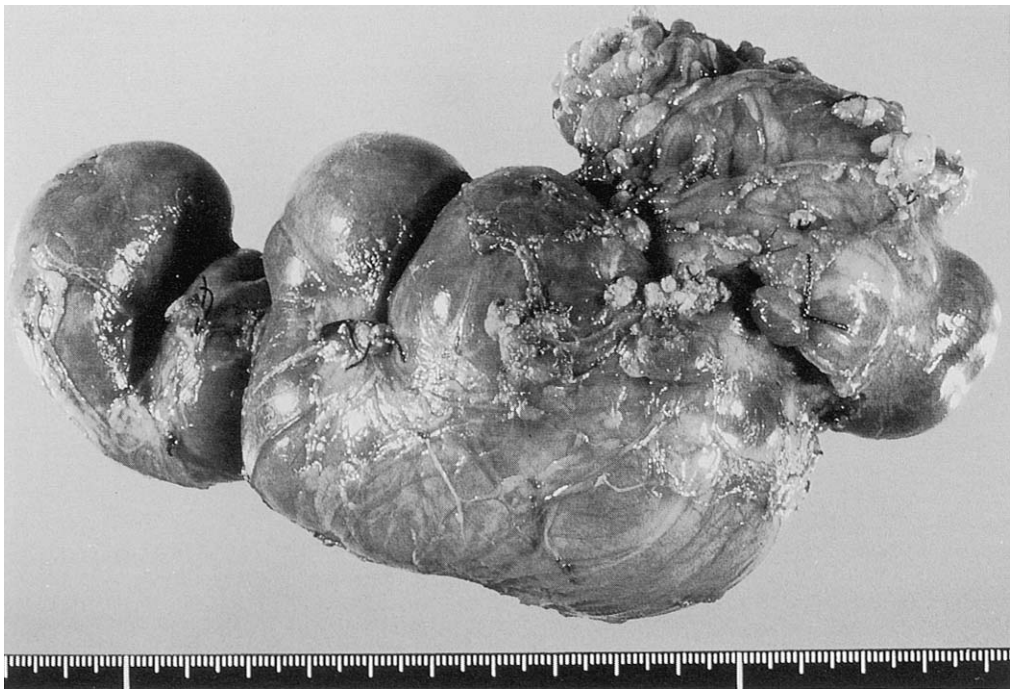

Fig. 2. Resected spleen in patient 2.

course of the decrease in TB following splenectomy was similar to that observed after PSE (Fig. 3). Both DB and IB concentrations similarly decreased following PSE. The mean platelet count increased significantly ($p<0.01$); $21.0\pm2.3\times10^4/mm^3$ before and $64.8\pm4.7\times10^4/mm^3$ after splenectomy. The WBC count increased as well; $4.8\pm0.4\times10^3/mm^3$ before and $13.0\pm0.8\times10^3/mm^3$ after splenectomy ($p<0.01$). The RBC count increased and correlated well with the TB in patient No. 2 ($r=0.791$, $p<0.01$) (Fig. 4). The Hb increased and correlated well with the TB in patient No. 1 ($r=0.619$, $p<0.05$) and No. 3 ($r=0.759$, $p<0.01$) (Fig. 5). On the other hand, splenectomy did not influence the values of GOT, GPT, γ-GTP, LDH, or ALP.

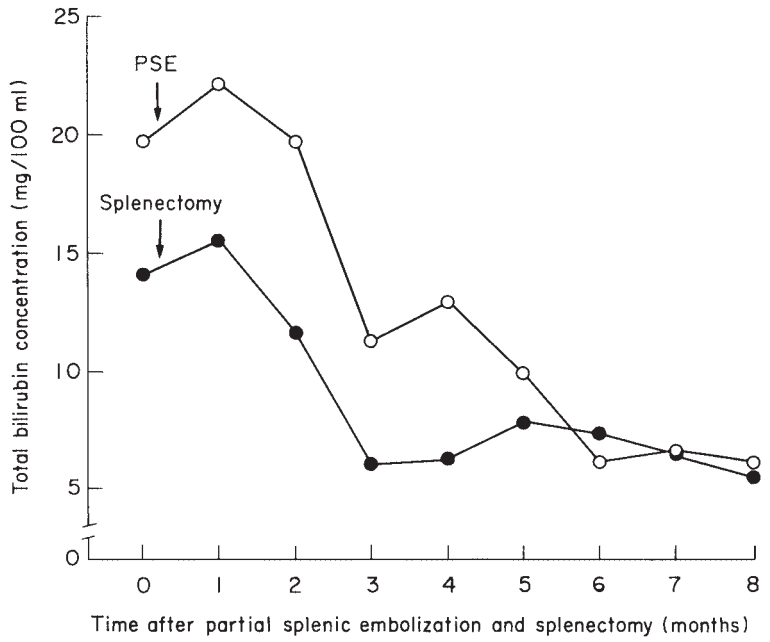

Fig. 3. Changes in the total bilirubin after splenectomy in patient 3.
The time course of the decrease in the total bilirubin after splenectomy was similar to that observed following partial splenic embolization (PSE).
○ PSE; ● splenectomy.

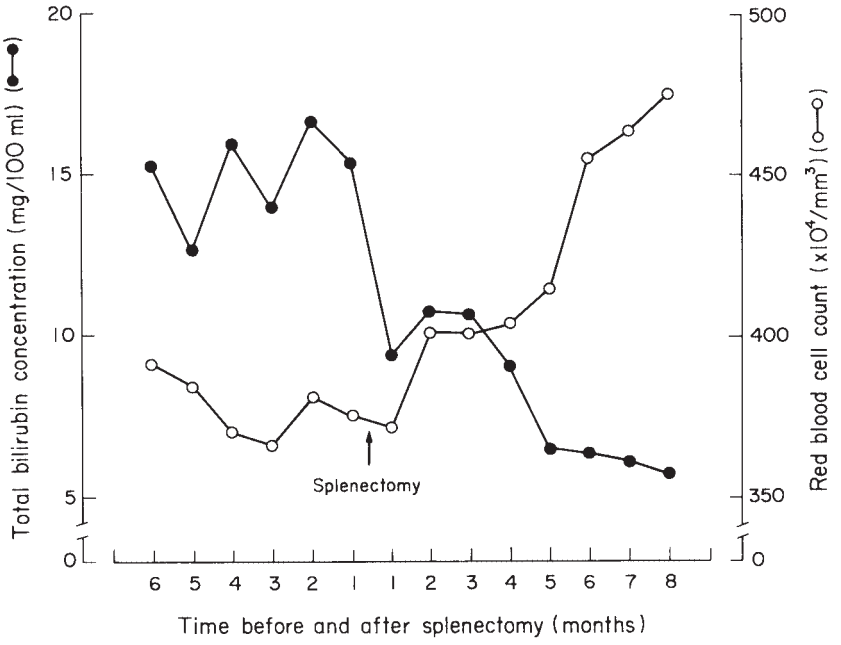

Fig. 4. Changes in the total bilirubin (●—●) and the red blood cell (○—○) count before and after splenectomy in patient 2.
There is a statistically significant correlation between them ($r=0.791$, $p<0.01$).

Discussion

Laurent et al. (1990) believed that jaundice in patients with biliary atresia is a result of liver cirrhosis and liver-cell failure. The present study, however,

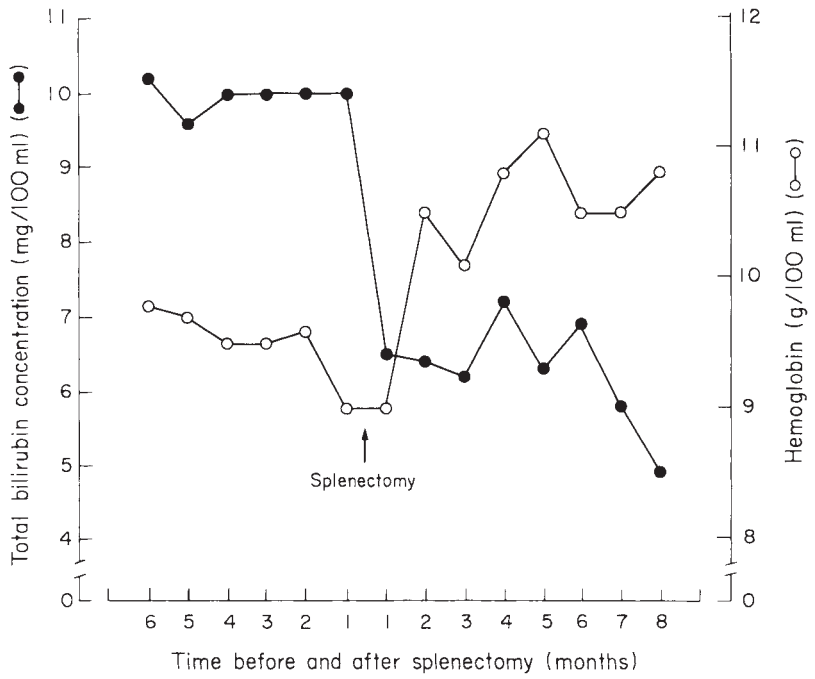

Fig. 5. Changes in the total bilirubin (●—●) and the concentration of hemoglobin (○—○) before and after splenectomy in patient 1.
There is a statistically significant correlation between them ($r=0.619$, $p<0.05$).

demonstrated a marked improvement in jaundice following splenectomy in spite of no improvement in hepatic function. It suggests that the progression of jaundice, which is often found on long-term, post-operative follow-up in some patients with biliary atresia, may not be a consequence of progressive liver cirrhosis or liver-cell failure. The decrease in jaundice which was observed after splenectomy may reflect a decreased turnover in bilirubin. Chiba (1985) has suggested that the life span of the RBC is shorter in patients with biliary atresia after Kasai procedure. Cytopenia and an elevation of serum bilirubin can be explained by increased sequestration and subsequent destruction of RBCs in the enlarged spleen (Ando et al. 1996). This hypothesis suggests why splenectomy may be an effective palliative treatment in patients with biliary atresia.

Splenectomy is, however, by no means free of complications. The spleen is the main site of human IgM production and is important in its anti-bacterial actions (Brown 1981; Baesl and Filler 1985). Immunoglobulin levels, particularly IgM, decrease significantly after splenectomy (Schumacher 1970; Koren et al. 1984) and results in an increased risk for severe infections illness that required hospitalization (Singer 1975; Shaw and Print 1989; Jahn et al. 1993). Rather than the prophylactic use of antibiotic therapy after splenectomy, patients should be advised to bring the child to the hospital anytime an illness or fever develops for immediate loading dose of an appropriate antibiotic (Posey and Marks 1983). Splenectomy may sacrifice established collateral veins and have a negative effect on portal circulation resulting in variceal bleeding. Fortunately, endoscopic

injection sclerotherapy (EIS) is an effective method of treatment of acute hemorrhage from esophageal varices (Donovan et al. 1986; Thapa and Mehta 1990). Splenectomy may also produce a hyper-coagulable state with its attendant thromboembolic complications when the platelet count increases more than $100 \times 10^4/mm^3$ (Goldstone 1978; O'Sullivan et al. 1994).

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