Acta Medica et Biologica
Vol. 42, No. 1, 13-17, 1994

Endoscopic Injection Sclerotherapy for Variceal Hemorrhage in Children with Congenital Biliary Atresia

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Received December 22, 1993

Summary. **Endoscopic injection sclerotherapy (EIS) was performed in seven children for the treatment of esophageal varices. Six of the patients had congenital biliary atresia (CBA) and had undergone a previous Kasai procedure, and biliary hypoplasia was the cause of varices in the remaining case. EIS was employed therapeutically for variceal rupture in six instances and prophylactically in one patient. From the initiation of EIS, no bleeding occurred in any case. There were no serious complications. Endoscopic grade and variceal size improved after treatment. Ultimately, five patients died of hepatic failure, including two patients who died of hepatic failure resulting from bleeding of esophagogastric varices at 4 days and 28 days after EIS. EIS of large varices in children with CBA is a safe and useful modality. Prophylactic EIS should be considered in patients with varices with high risk of rupture.**

INTRODUCTION

More than a decade has passed since the introduction of endoscopic injection sclerotherapy (EIS) in the field of pediatric surgery.[1-4] Due in part to improved operative results on congenital biliary atresia (CBA) with Kasai's portoenterostomy[5,6] the number of children developing esophageal varices has increased.[7-9] The therapy for portal hypertension in children is controversial. In some children esophageal varices disappear spontaneously. EIS is recommended for bleeding esophageal varices after kasai's correction in patients with CBA. We describe below our methods and the results of EIS in children since 1980.

PATIENTS

During the period 1980 through 1991, EIS was performed on 93 patients with esophageal varices.[10] Seven children with CBA or biliary hypoplasia underwent EIS. Six patients with CBA had undergone a previous Kasai procedure. The above seven patients consisted of four boys and three girls, with ages at the beginning of EIS ranging from 1 to 13 years (Table 1). Two patients with acute hemorrhage underwent emergency EIS and four patients with a prior history of variceal bleeding underwent elective EIS. EIS was performed prophylactically in one patient with varices at high

Table 1. Clinical features.

Patients	Age	Sex	Diagnosis	Timing	Child's category	TB mg/dL	Albumin g/dL	Ascites
1. M H	1y 8m	F	CBA	elective	C	35.0	4.5	−
2. N H	2y 1m	M	CBA	elective	C	4.6	3.2	+
3. S H	1y 5m	F	CBA	elective	C	36.1	4.2	+
4. R H	8y 8m	M	CBA	elective	C	14.3	3.0	+
5. H K	13y 7m	M	CBA	emergency	C	31.1	3.2	++
6. K O	5y 3m	F	CBA	prophylactic	A	1.4	3.7	−
7. G R	1y	M	biliary hypoplasia	emergency	C	41.4	2.3	++

TB: serum total bilirubin concentration, CBA: congenital biliary atresia,
Ascites +: controllable ascites, Ascites ++: uncontrollable ascites

Table 2. Changes of endoscopic findings, complications and prognosis.

Patients	Endoscopic findings* (before EIS)	(after EIS)	Complications	Variceal bleeding	Prognosis	
1. M H	F3 Cb RC (+)	F1 Cw RC (−)	(−)	(−)	Dead of liver failure	(7m)
2. N H	F2 Cb RC (−)	F1 Cw RC (−)	(−)	(−)	Dead of liver failure	(4m)
3. S H	F3 Cb RC (+)	F1 Cw RC (−)	fever	(−)	Dead of liver failure	(1y 2m)
4. R H	F2 Cb RC (+)	F1 Cw RC (−)	(−)	(−)	Alive	(1y 2m)
5. H K	F3 Cb RC (+)	/	(−)	(−)	Dead of liver failure	(28d)
6. K O	F2 Cb RC (+)	F1 Cw RC (−)	fever	(−)	Alive	(1y 4m)
7. G R	F2 Cw RC (+)	/	(−)	(−)	Dead of liver failure	(4d)

*: Adapted from the general rules for recording endoscopic findings on esophageal varices, edited by Japanese Research Society for Portal Hypertension.
F: form, Cb: variceal fundamental coclor is blue, Cw: variceal fundamental color is white, RC: red color sign.

Fig. 1. Sclerosant (EOI) is injected into the esophageal varices.

risk of rupture. Endoscopic findings are demonstrated in Table 1, based on the Japanese Rasearch Society for portal hypertension classification.[11] According to Child's category,[12] the seven children were divided into three stages. Six patients were classified as Child's C and the other (case 6) as Child's A (Table 2). Six patients were jaundiced with serum bilirubin levels ranging from 4.6 mg/dL to 41.1 mg/dL (Teble 2). Hypoalbuminemia below 3.5 g/dL was present in four patients, and uncontrollable ascites in two.

METHODS OF EIS

EIS was performed under general anesthesia in orally intubated patients. A flexible fiberscope, XK10 (Olympus Ltd., Tokyo, Japan), with an attached balloon around the fiberscope was used in all cases. A 23 or 25 gauge needle was used for the injection of a sclerosant. The sclerosant was a 5% solution of ethanolamine oleate containing radiopaque medium Iopamidol^{R} (EOI).

In two cases with acute hemorrhage (cases 5 and 7), a Sengstaken-Blakemore tube remained in place with the stomach balloon inflated during the early phase of the procedure. At the conclusion of EIS a nasogastric tube was passed into the stomach to remove air inflated by the fiberscope. The nasogastric tube remained overnight. EOI was injected into the varix (Fig. 1) during the initial treatment by the free hand technique with the balloon inflated with 10-15 mL of air. Approximately 2-3 mL of EOI was injected per puncture and the total amount of sclerosant was limited to 2-3 mL per kilogram body weight. When extravasation of EOI was visualized fluoroscopically, the infusion of EOI was stopped. After infusion of EOI the external cylinder was left in place to maintain compression. If bleeding continued after compression, the bleeding site was occluded by the balloon attached to the fiberscope. The fiberscope tip must be passed more than 5 cm distal to the injection site.

Large varices were treated by combined sclerotherapy consisting of intra- and extra-variceal injection.[13] EOI (1 ml) was initially injected on both sides follow-

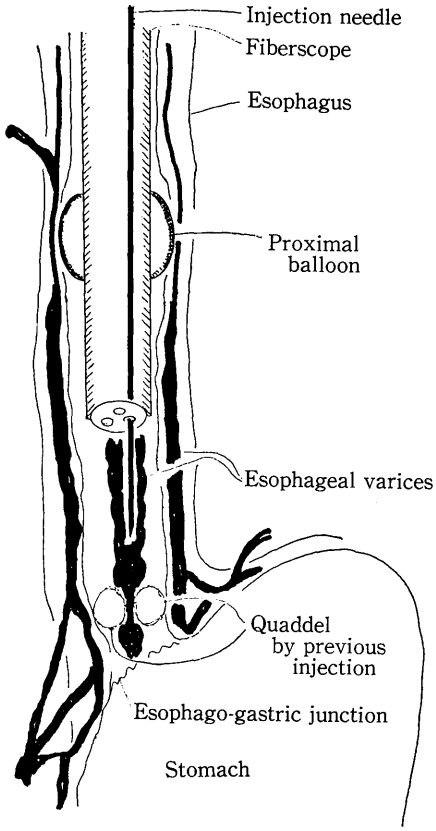

Fig. 2. Carbon schema of the combined sclerosis procedure consisting of para-esophageal injections and an intravariceal injection with proximal balloon compression.

ed by injection of 2-3 ml of sclerosant into the varix proximal to the initial paravariceal injection sites (Fig. 2).

RESULTS

EIS was performed on 17 occasions (average 3, range 1 to 6) in seven children. The treatment periods varied from 2 weeks to 2 months with an average of 3 weeks, excluding two in-hospital deaths. Successful hemostasis was obtained in two emergency cases (cases 5 and 7). However, both patients died of hepatic failure secondary to the initial hemorrhage.

After only one series, the form of the varices (F) was diminished to grade 1 and the red color sign (RC sign) had disappeared in all cases (Table 2). Complete disapperance of the varices was not observed after only one series. Pipe line varices[14] as occurred in case 5 were controlled by combined sclerosis according to the modified Suzuki method.[12] A thrombosed vein was seen at the second treatment (Fig. 3).

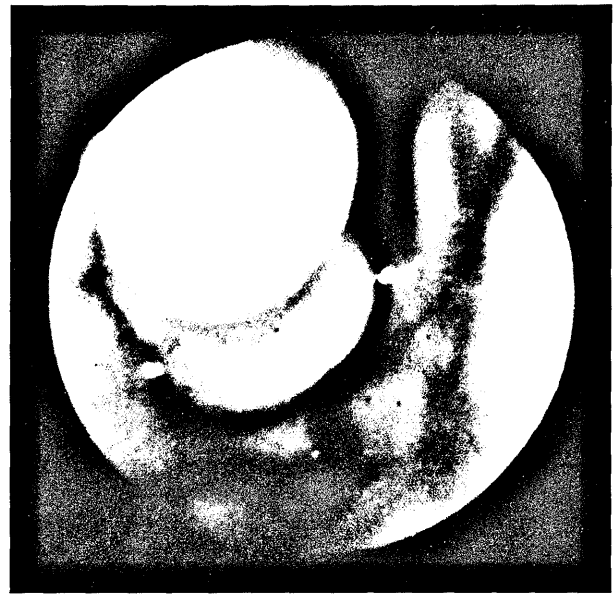

Fig. 3. A thrombosed varix as visualized 1 week after the initial sclerosis.

There were no complications due to EIS, including upper gastrointestinal bleeding, esophageal ulceration or stenosis, pleural effusion, increased ascites, or a worsening of jaundice. Fever was noted immediately after EIS in two children (cases 3 and 6).

DISCUSSION

Bleeding from the esophago-gastric varices is a frequently fatal complication of portal hypertension. EIS has recently been introduced for the treatment of the esophageal varices and offers the following advantages in comparison with surgical treatment: EIS can be safely performed even if the hepatic function is poor. Repeated procedures are possible and little to no deterioration of hepatic function follows EIS.

Portal hypertension may develop following a successful Kasai procedure in some children with CBA.[8,9] Esophageal varices frequently develop in jaundiced patients whose hepatic function has deteriorated because of insufficient biliary drainage.[8,9] It is reason-

able to perform EIS for the management of these end-stage patients with varices. However, reports of EIS in children are lacking. Special considerations include repeated anesthesia, difficulty of injection into varices and side effects of the sclerosant.

Although our series is small and the follow-up period has been short, there has been no recurrent hemorrhage, and endoscopic findings have improved in all cases after the treatment. Furthermore, complications were minimal. These results reflect the necessity of using a smaller amount of sclerosant and accurate puncture of the varix (Fig. 1). Eliminating all large risk varices and the red color sign is essential for a good outcome.

Complete irradication[15] of varices requires a large amount of sclerosant and repeated injections. According to report, esophageal stenosis frequently occurred, requiring dilatation with an esophageal bougie. We took care during extra-variceal injections to use less than 1 ml of radiopaque sclerosant. This may have prevented esophageal ulceration which leads to esophageal perforation or stenosis.

Endotracheal intubation and general anesthesia during EIS prevent unexpected movements of the patients and the aspiration of blood. Deterioration of the hepatic function did not occur due to repeated exposure to general anesthetics.

It is difficult to treat the pipe line varices[13] which are characterized by a large diameter and high blood flow. However, we eliminated large varices as described above. The combination of paravariceal injection and inflation of the balloon attached to the fiberscope slows the flow through the varix. This damming up procedure may augment the coagulability of the sclerosant injected into the varix because of the prolonged contact with stagnant blood.

Long-term survivals did not occur in this series. EIS for end-stage patients with liver failure is predominantly palliative. Shunt surgery is poorly tolerated by these patients. When liver transplantation[16] is considered, EIS is useful to control bleeding without causing any injury to the abdominal organs.

Despite ongoing controversies,[17] we currently recommend prophylactic EIS for CBA patients with high risk varices. In our series, two patients who underwent emergeny EIS died of hepatic failure secondary to the initial variceal bleeding. Neither patient experienced recurrent hemorrhage after EIS. In patients with poor hepatic reserve, bleeding is often fatal. For this reason we recommend prophylactic EIS under general anesthesia in children with high risk varices.

In conclusion, EIS is a safe and effective modality in the management of esophageal varices in children with CBA. EIS should be considered when treating esophageal varices of any origin in children.

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