

Intrahepatic and periduodenal embolization: New technique for Protein-Losing Enteropathy treatment in congenital heart disease patients

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Background

Protein-losing enteropathy (PLE) treatment is primarily symptomatic management to reduce fluid overload and electrolyte disturbances. Liver lymphatics are a mediator of PLE pathogenesis and their embolization has shown varying success with some patients having minimal or no improvement.

Objectives

This study sought to describe extrahepatic causes of PLE using MR lymphangiography and evaluate the outcomes of a combined intrahepatic and periduodenal lymphatic embolization strategy for PLE treatment.

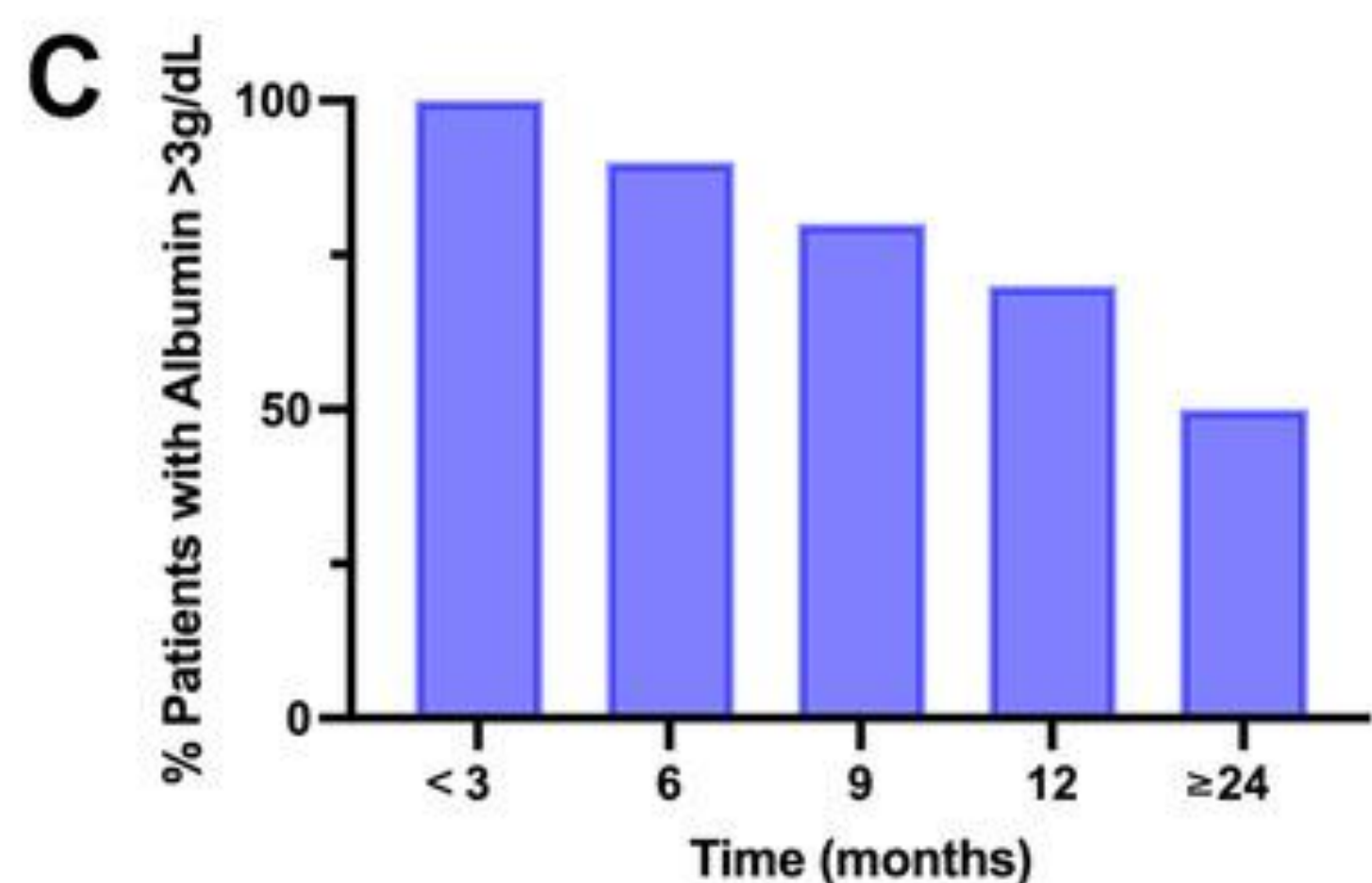
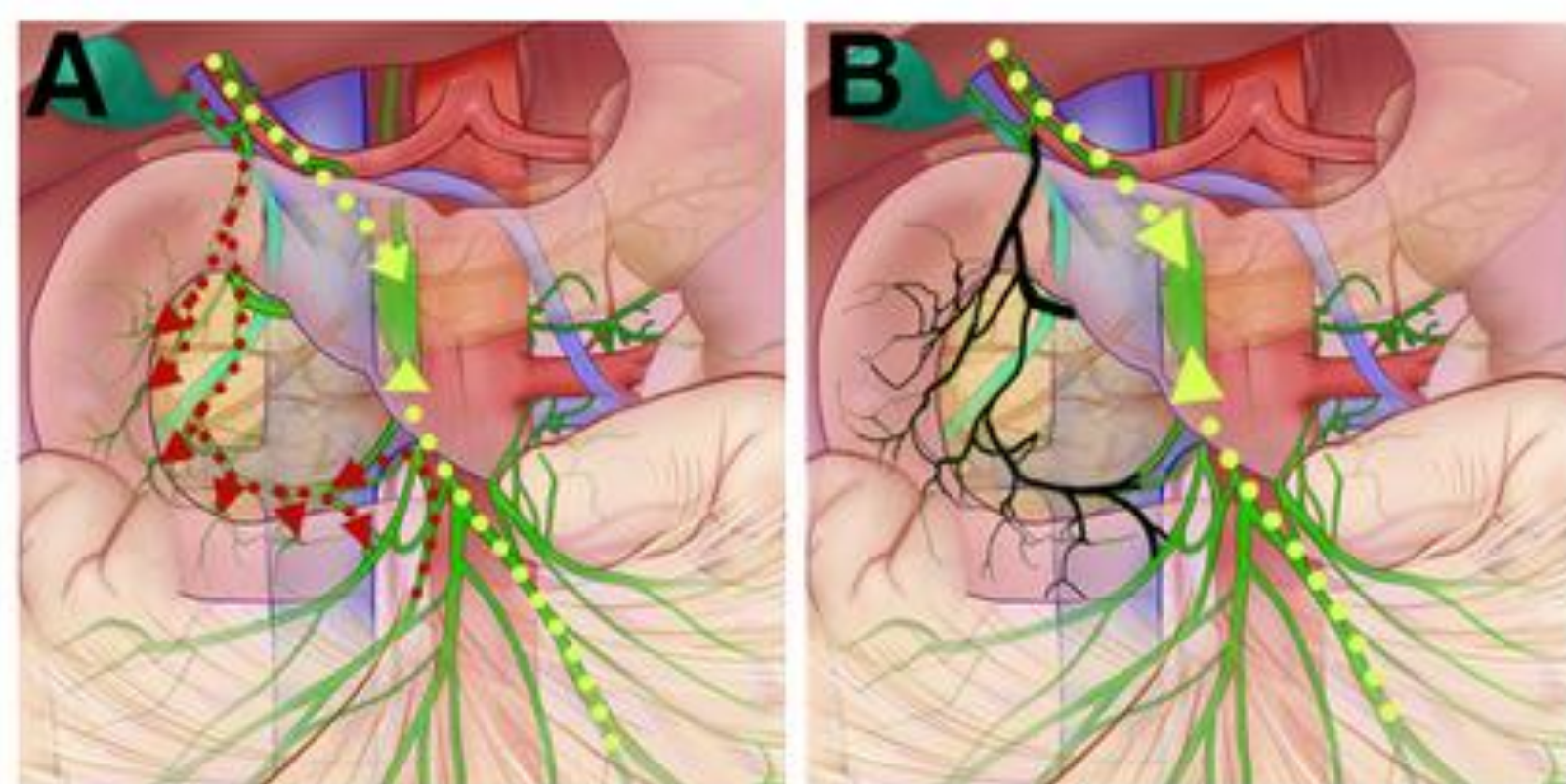
Methods

This was a single-center, retrospective review of imaging and medical records of the first 10 consecutive patients with congenital heart disease previously surgically palliated and complicated by PLE who underwent isolated lymphatic intervention using both intrahepatic and periduodenal lymphatic embolization at our institution.

Results

- Twelve lymphatic interventions were performed in 10 patients.
- Intrahepatic and intranodal MR lymphangiography was performed in all procedures and intramesenteric in five.
- Duodenal leaks were identified from intrahepatic and/or intramesenteric access in all patients.
- Endoscopy with injection of isosulfan blue in hepatic or periduodenal lymphatics confirmed lymphatic leak in 11/12 procedures.
- Glue embolization of hepatic periportal lymphatics was performed followed by targeted embolization of periduodenal lymphatics.
- All patients responded to a single intervention with symptom improvement and albumin $\geq 3\text{g/dL}$ (median 15d, IQR 6-20) (Central Figure).
- Most common post-procedure complication was transient pancreatitis in 75% (9/12) of patients, followed by hyperbilirubinemia in 33% (4/12); both either improved or resolved by the time of discharge or transfer.
- Longer-term follow-up (median 668d, range 111-1059d) identified two populations, complete responders (n=5) and incomplete responders (n=5) with either a slow or more acute decline (<5 months) in albumin (Figure 1).

Central Figure



Intrahepatic and Periduodenal embolization strategy and outcomes in congenital heart disease patients with PLE.

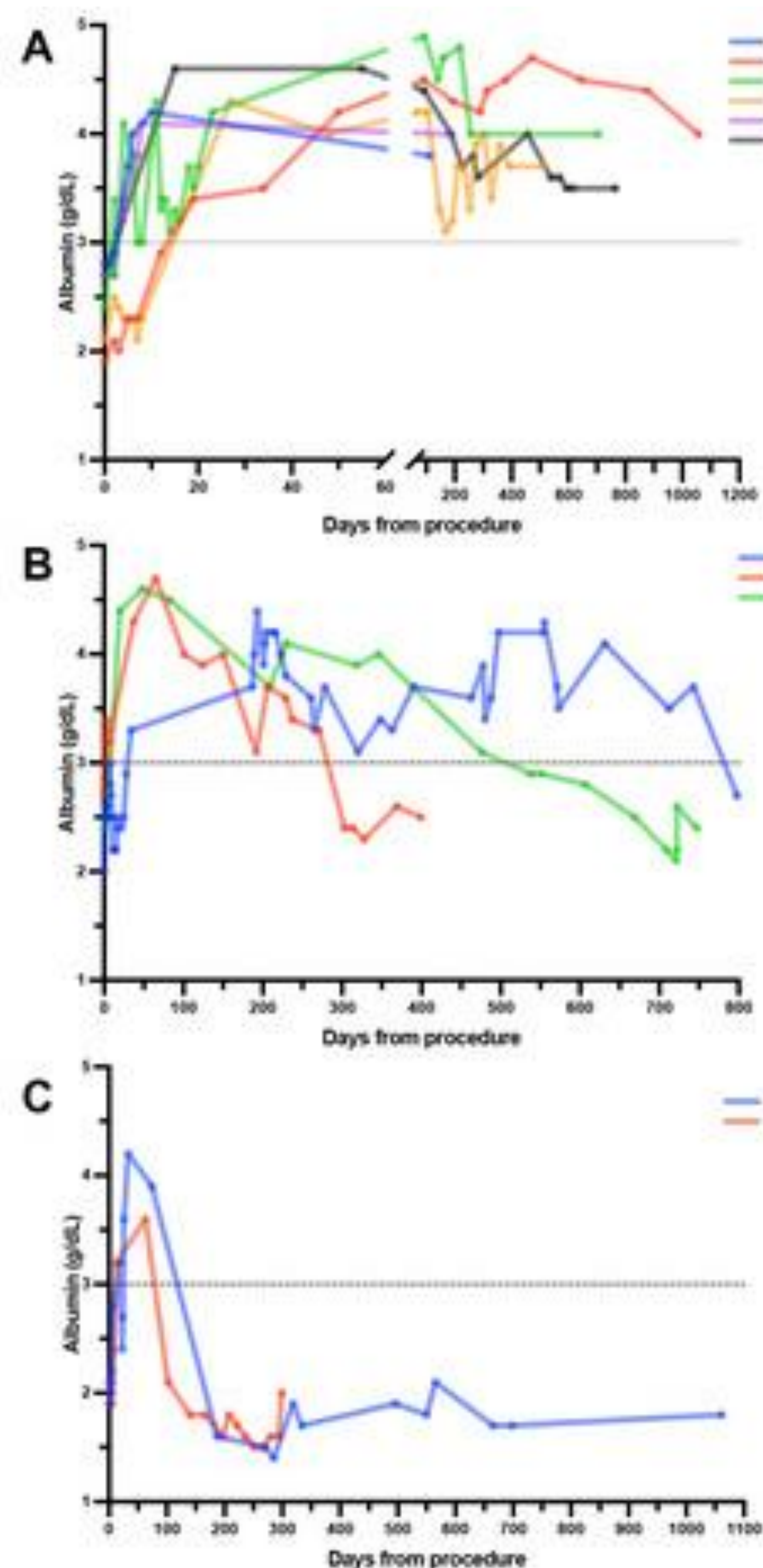
(A) Lymphatic perfusion sources in PLE (intrahepatic and mesenteric, yellow arrows) with connections that converge on the duodenum (red arrows) in addition to normal drainage (yellow arrows).

(B) Embolization of the intrahepatic and periduodenal lymphatics while keeping normal drainage intact (yellow arrows).

(C) Albumin levels in patients over time after the procedure.

Median time to recurrence (albumin <3 g/dL) was 303 days (range 102 – 798).

Figure 1



Albumin levels after combination of hepatic and periduodenal embolization. There are 3 response patterns: (A) Complete responders with normalization of albumin levels and no recurrence at time of last follow up. (B) Incomplete responders with initial normalization of albumin, but with a variable decline before recurrence of PLE. (C) Initial responders but with more acute decline and PLE recurrence (within 6 months).

Conclusions

The authors demonstrate additional sources of PLE and describe a new technique targeting both the hepatic lymphatic source and periduodenal destination to treat PLE.

This comprehensive strategy led to an initial improvement of albumin levels in all patients and sustained albumin levels in 50% of patients with a single intervention.

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Acknowledgements

Supported in part by: The Cardiac Center at The Children's Hospital of Philadelphia, Jill and Mark Fishman Center for Lymphatic Disorders at the Children's Hospital of Philadelphia

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