Successful Peritoneal Dialysis in 2 Siblings with Alport's Disease and Gastric Pull-Up

Previous abdominal surgery is traditionally considered in many centers as an absolute contraindication for peritoneal dialysis (PD) (1). Possible concerns include increased risk of hernia, gastric reflux, cough, respiratory distress, mechanical issues related to dialysate drainage and inadequacy secondary to intra-abdominal adhesions (2,3). These issues are readily addressed with individualization of the PD prescription with or without the use of an automatedycler (3). It has been suggested that poor candidates can be identified pre-initiation by use of Technetium 99 scintigraphy to detect abdominal adhesions (4). However, preoperative scintigraphy has not come into wide use, and many patients are being denied PD simply because of previous surgical procedures.

Leiomyomas of the esophagus are a rare cause of dysphagia and are usually associated with syndromes such as Alport's syndrome (AS) (5). Alport's syndrome is associated with diffuse leiomyomatosis in 2 – 5% of patients. A variant of AS with diffuse leiomyomatosis (ASDL) is defined by the association of AS with esophageal, gastric, vulvovaginal, and bronchial leiomyomas along with clitoral hypertrophy (6). It is proposed that leiomyomas in ASDL are a result of defective differentiation of smooth muscle in the viscera secondary to partial deletions of the type IV collagen genes COL4A5 and COL4A6 on chromosome Xq22 (5,6).

We present 2 siblings with renal failure secondary to AS who have successfully conducted PD having undergone esophagectomy and gastric pull-up for esophageal leiomyomatosis. This is the first report, to our knowledge, of PD after this procedure.

PATIENT 1

A 52-year-old female, body mass index 16.3, diagnosed at 16 years of age with AS with esophageal leiomyomatosis, who underwent extensive surgery involving esophagectomy with gastric pull-up (Figure 1) through a midline laparotomy, thoracotomy and neck incision. She also underwent a radical hysterectomy along with a vulvectomy for extensive leiomyomatosis through a lower midline Pfannenstiel incision. She subsequently also had an open appendectomy. The patient underwent live-related kidney transplantation that lasted for 17 years, needing PD initiation eventually. A diagnostic laparoscopy in 2011 revealed adhesions of the transverse colon with omentum limited to the upper abdomen. A Tenckhoff catheter was inserted laparoscopically and tidal continuous cycling PD started. Initially she could not tolerate 2-liter exchanges. However she is now comfortable with 1.5-liter exchanges and is adequately dialysed. She has had no gastrointestinal, pulmonary, mechanical, or peritonitis episodes related to PD for the 36 months that she has been on the therapy.

PATIENT 2

This is the sibling of patient 1. He underwent the same esophagectomy and gastric pull-up procedure as his sister at 17 years of age. He received a preemptive live-related kidney transplant that failed after 5 years. Prior to PD initiation he had a major gastric bleed from a Dieulafoy lesion needing surgical approach by midline

Figure 1 — Small bowel barium series — arrow points to direct origin of small bowel intra-abdominally in the absence of stomach.

laparotomy and posterolateral thoracotomy to arrest the bleeding. He also had a pericardiectomy in 2001 for minoxidil-induced pericarditis. He started PD at the age of 33 and his course was complicated by only 1 episode of peritonitis that responded well to standard treatment. He was on continuous cycling PD for 6 years with no mechanical issues and subsequently received a deceased donor kidney transplant and is doing well.

CONCLUSION

Prior abdominal and thoracic surgery including esophagectomy and gastric pull-up should not be considered a contraindication for PD.

DISCLOSURES

The authors have no financial conflicts of interest to declare.

Venkat S. Vellanki¹
Joanne M. Bargman^{2*}

Clinical Fellow, Department of Nephrology¹
Toronto General Hospital, University of Toronto
Toronto, Ontario, Canada
Professor of Medicine, University of Toronto²
Staff Nephrologist, University Health Network
Toronto, Ontario, Canada

*email: joanne.bargman@uhn.ca

REFERENCES

1. Palomar R, Gonzalez-Martin V, Martin L, Morales P, Martin de Francisco AL, Arias M. Is abdominal surgery still a contraindication for peritoneal dialysis? *Nephrol Dial Transplant* 2007; 22:2360–1.
2. Wanke T, Auinger M, Lahrmann H, Merkle M, Formanek D, Irsigler K, *et al.* Diaphragmatic function in patients on continuous ambulatory peritoneal dialysis. *Lung* 1994; 172:231–40.
3. Tarlo SM. Peritoneal dialysis and cough. *Perit Dial Int* 2003; 23:424–6.
4. Guditi S, Sudhakar P, Ram R, Prasad N, Prabhakar VVS, Dakshinamurthy KV. Peritoneal Scintigraphy in the diagnosis of adhesions. *Perit Dial Int* 2010; 30:112–21.
5. Rabushka LS, Fishman EK, Kuhlman JE, Hruban RH. Diffuse esophageal leiomyomatosis in a patient with Alport syndrome: CT demonstration. *Radiology* 1991; 179(1):176–8.
6. Garcia-Torres R, Orozco L. Alport-leiomyomatosis syndrome: an update. *Am J Kidney Dis* 1993; 22:641–8.
doi: 10.3747/pdi.2013.00127