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*CKJ Review*

# The curious right-sided predominance of peritoneal dialysis-related hydrothorax

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**Abstract**
A known complication of peritoneal dialysis is the movement of dialysate into the pleural space, termed hydrothorax. Peritoneal dialysis-related hydrothorax is almost uniformly right-sided and represents one of many presentations of the porous diaphragm syndrome. In addition to diaphragm porosity, the inherent intestinal circulation, lower hydrostatic pressure in the right upper quadrant and liver capsule may contribute to this right-sided predominance. Similar right-sided presentations have been described in Meigs syndrome, bilious effusions with gastric or duodenal perforations, hepatic hydrothorax and nephrotic syndrome-related chylothorax. PD-related hydrothorax can be repaired by pleurodesis or video-assisted thoracoscopy followed by resumption of peritoneal dialysis.

**Keywords:** diaphragm; hydrothorax; peritoneal dialysis; pleural effusion

## Introduction

Hydrothorax is a rare complication of peritoneal dialysis (PD) with an estimated prevalence of 1–2% [1, 2]. The clinical presentation is typically acute shortness of breath and the diagnostic chest x-ray reveals a small or massive effusion. The effusion fails to improve or may even worsen after institution of hypertonic dialysate exchanges. The effusion may develop shortly after the initiation of PD suggesting a pre-existing diaphragmatic defect or present months to years after initiation of dialysis, usually after physical exertion, suggesting an acquired defect in the diaphragm from a sharp increase in intraabdominal pressure [3, 4]. The diagnosis of a PD-related hydrothorax is made by thoracentesis revealing a transudative effusion, with a variable glucose concentration in the pleural fluid but higher than the serum concentration, as described by Momenin *et al.* [5]. In patients treated with icodextrin, the effusion can be confirmed as dialysate by addition of iodine which complexes with the starch and results in a bluish-black discoloration of the fluid [6].

A curious finding in PD-related hydrothorax is the striking right-sided predominance of this presentation. Why is the hydrothorax so predominant on the right side? Do other abdominal presentations show this right-side predominance—indicating a wider spectrum of related clinical entities? These questions will be addressed in this review.

## Propensity for right-sided hydrothorax in peritoneal dialysis

Indeed, PD-related hydrothorax has a striking right-sided predominance. In this author's opinion, after reviewing published cases, over 90% of presentations were right-sided. In a review of 50 PD patients presenting with hydrothorax, Nomoto and colleagues noted that 88% were right sided [7]. Mak and colleagues described eight patients who presented with PD-related hydrothorax, all right-sided [8]. Similarly, Tang and colleagues described nine PD patients presenting with hydrothorax between 1998 and 2002. Eight of the nine presented with right-sided hydrothorax [9]. Additional case reports and case series show the most frequent presentation to be on the right side [4, 6, 10–18].

## Anatomy of the normal diaphragm

The normal diaphragm functions as a septum separating the abdominal and pleural cavities. The anatomic components of the diaphragm are a central tendon with radiating muscle fibers that cascade from the central tendon to attach to the ribs, the sternum and lumbar spine. By contraction of the muscle fibers the diaphragm allows for expansion of the chest cavity allowing for inspiration and muscular contraction around the gastroesophageal junction to prevent reflux [19].

The diaphragm is covered by pleura on the superior surface and peritoneum on the inferior surface. Phrenic arteries arising chiefly from the aorta provide the blood supply to the diaphragm and enter from the inferior surface. The diaphragm is innervated chiefly by the left and right phrenic nerves arising from the third to fifth cervical roots [19].

The normal diaphragm has larger openings that allow critical structures to transit through the septum—the aortic hiatus, esophageal hiatus and inferior vena cava. Smaller openings allow for passage of the nerves and veins. These

openings are created by the arrangement of muscle fibers into bands allowing for special openings in the diaphragm. While diaphragmatic openings are present, in the normal diaphragm, none of these anatomic structures allow for transit of intraperitoneal dialysate into the pleural cavity, presumably due to the interface of muscle fibers and projections of serous membranes and connective tissue.

## Porosity of the diaphragm

Dr Paul Kirschner eloquently described the anatomic defects that allow for passage of fluids, cells, tissue and gases through holes in the diaphragm [20]. He termed these medical presentations as 'porous diaphragm syndromes'. While larger diaphragmatic hernias can exist due to failure to form the normal diaphragm during development of the embryo, these defects are more obvious in their clinical presentation (Bochdalek hernia, Morgagni hernia). More insidious are the smaller defects that may present as pinhole defects, slits or webs of a centimeter or more in size. Of all potential anatomic locations within the diaphragm these smaller defects have been identified as occurring within the tendinous portion of the central diaphragm and to be more common in the right hemidiaphragm [20]. The defects may be congenital or acquired due to intraperitoneal pressure spikes or tissue necrosis.

## Intestinal circulation and piston action of the liver

The right-sided predominance of porous diaphragm syndromes may not be due to increased numbers of diaphragmatic defects on the right portion of the central tendon, in isolation, but exacerbated also by the movement of substances from the pelvis to the right upper quadrant during intestinal peristalsis. The ascending peristalsis of the right colon, followed by movement laterally and downward by the transverse and descending colon create an 'intestinal circulation' that sweeps pelvic fluids or substances up into the right upper quadrant. Additionally, hydrostatic pressure is highest in the pelvis and lowest in the suprahepatic region during inspiration, presumably due to the outward movement of the ribs which enlarges the space in the right upper quadrant [21]. This circulation and collection of fluid in the right upper abdomen was confirmed by peritoneography and occurs in patients that are lying as well as standing [22]. As the right diaphragm contracts it is met by the relatively firm liver capsule which acts as a piston, driving fluid thru the porous defects on the right diaphragm. The left upper quadrant is believed to be more soft and compliant and does not create this piston-like force [20]. As concluded by Kirschner, this combination of tendinous defects on the right, the intestinal circulation drawing fluid or substances into the right upper abdomen and piston-like action of the liver capsule explain the right-sided predominance to these syndromes (Table 1).

## Select porous diaphragm syndromes

An astonishing variety of right-sided porous diaphragm syndromes exist in clinical medicine, impacting many medical specialties (Table 2).

Bilious right-sided effusions have been described in duodenal ulcer and gastric perforations [23, 24]. Portal hypertension from liver cirrhosis can present with ascites and pleural effusion, typically right-sided, and has been termed 'hepatic hydrothorax' [25–27]. Right-sided chylothorax has been described in situations of abdominal chyle such as abdominal lymphoma and nephrotic syndrome [28, 29].

Many fascinating porous diaphragm syndromes are gynecological presentations. Meigs syndrome is characterized by a benign ovarian fibroma presenting with ascites, and right-sided effusions [30]. Pseudo-Meigs syndrome is the presentation of a right-sided effusion associated with other non-fibromatous lesions such as uterine leiomyomas [31]. Recurrent right-sided hemothorax and pneumothorax (catamenial pneumothorax) are due to migration of endometrial tissue up the fallopian tubes into the abdominal cavity across the porous diaphragm into the pleural cavity where it implants and erodes into lung tissue [32, 33]. A similar pathway has been attributed to the findings of hemothorax with ectopic pregnancies.

**Table 1.** Pathophysiology of right-sided PD-related hydrothorax

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| 1. Abdominal cavity containing dialysate |
| 2. Congenital or acquired porous diaphragm |
| 3. Intestinal circulation sweeps fluid preferentially to abdominal right upper quadrant |
| 4. Reduced hydrostatic pressure in abdominal right upper quadrant collects dialysate |
| 5. Piston-like motion of liver capsule shunts fluid across diaphragmatic vent |

**Table 2.** Abdominal presentations with concurrent right-sided pleural effusions

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| 1. Peritoneal dialysis-related hydrothorax |
| 2. Bilious effusion with perforated gastric or duodenal ulcer |
| 3. Hepatic hydrothorax |
| 4. Chylothorax secondary to nephrotic syndrome or lymphoma |
| 5. Benign ovarian adenoma with pleural effusion (Meigs syndrome) |
| 6. Pseudo-Meigs syndrome |
| 7. Hemothorax due to pleural endometriosis with/without right catamenial pneumothorax |

## Discussion

In patients presenting with PD-related hydrothorax, it should be appreciated that this presentation is part of a larger group of syndromes that share a common pathophysiology—the preferential development of diaphragmatic defects on the right side. While PD-related hydrothorax has been rarely described on the left side, practitioners should be especially vigilant to rule out other causes of effusions in left-sided presentations, such as those secondary to pancreatitis, congestive heart failure or infection. PD-related hydrothorax has been managed by minimally invasive techniques such as pleurodesis or more invasive approaches using video-assisted thoracoscopic surgery (VATS) [1, 2, 4]. The treatment goal is to seal the porous diaphragmatic vent to allow full separation of the peritoneal cavity from the pleural cavity. Successful resumption of PD after this repair is possible.

## Conclusions

PD-related hydrothorax is among a spectrum of porous diaphragm syndromes, the hallmark of which is the clinical presentation of a right-sided pleural effusion.

*Conflict of interest statement.* Dr Guest is an employee of Baxter Healthcare Corporation, Deerfield, IL, USA.

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