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Cardiopulmonary manifestations of portovenous shunts from congenital absence of the portal vein: Pulmonary hypertension and pulmonary vascular dilatation

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Abstract

HPS and PPHTN are unusual and challenging pulmonary manifestations of liver disease. We report two pediatric cases in association with heterotaxy polysplenia syndrome and congenital absence of the portal vein. Both patients were symptomatic and hemodynamically compromised and required aggressive medical therapy. One patient with PPHTN alone achieved a successful liver transplant. The second child presented with combined HPS and PPHTN and exhibited a different evolution of pulmonary vascular disease. These cases illustrate associations that must be entertained in the setting of heterotaxy syndrome, cyanosis, or pulmonary hypertension and how strategic medical combined with surgical management can provide a good outcome.

Keywords

portal vein agenesis; portopulmonary hypertension; hepatopulmonary syndrome; pediatric transplantation; congenital heart disease

Diseases involving the portal venous system and pulmonary vascular bed combined are difficult to manage. The major forms are the HPS and PPHTN. In HPS, there is vascular dilatation of the pulmonary bed. The pulmonary precapillary and capillary vessels are dilated and AVMs can sometimes be present leading to hypoxemia from right-to-left shunting. In PPHTN, the pulmonary vasculature is constricted resulting in elevated pulmonary arterial pressure and resistance. Not only are these pathological processes associated with increased morbidity, but they can prevent successful liver transplantation. We report two rare cases of congenital absence of the portal vein in association with severe HPS and PPHTN. They illustrate how strategic medical management can result in successful liver transplant alone and offer some possible insights into the pathogenesis of pulmonary dilatory vasculopathy in congenital heart disease.

Case 1

This patient presented with abdominal situs inversus at five wk of age and received an echocardiogram to survey for congenital heart disease. Echocardiogram findings included levocardia, left aortic arch, and interruption of the IVC with azygous continuation of the

suprarenal IVC to the right SVC. Right- and left-sided hepatic veins drained to the right atrium. There was also a small muscular ventricular septal defect. At age 2½ yr, he developed cyanosis (pulse oximetry 80%) and failure to thrive. Repeat echocardiogram estimated the RVSP to be 2/3rd systemic without pulmonary vein stenosis. Catheterization confirmed the echocardiogram findings and showed a mPAP of 35 mmHg, TPG of 22 mmHg, and PVRI of 1.8 Wood units m^2 (144 dyne $s/cm^5/m^2$). The ventricular septal defect was not seen; instead, a right-to-left shunt from bilateral and diffuse dilated small pulmonary vessels and probable small pulmonary AVMs accounted for the cyanosis (Fig. 1). Selective microbubble injection into the right and left pulmonary arteries showed rapid return of bubbles to the left atrium via the respective pulmonary veins. Angiogram of the azygous vein revealed a venous collateral emanating from below the diaphragm. Subsequent magnetic resonance imaging and abdominal angiograms confirmed that the splenic and superior mesenteric veins converged into a large tortuous venous collateral that drained into the azygous vein (Fig. 2). No portal vein structure was identified.

Over the next 3½ yr, with the aid of a gastrostomy tube, he maintained weight but continued to exhibit PAH, cyanosis, and limitation during physical activities. Consideration was given to creating a mesenteric-left portal vein bypass (REX shunt) (1), which would bridge the superior mesenteric vein to the intrahepatic left portal vein. Repeat direct angiography of the shunt and splenic and mesenteric veins was not able to show a residual portal vein. Furthermore, angiography by priming the hepatic artery with nitroglycerine while balloon occluding the azygous vein also did not demonstrate any patency of the portal vein or its branches (Fig. 3). Thus, a REX shunt was not felt to be feasible. A repeat catheterization showed mPAP increased to 57 mmHg (80% systemic), TPG of 49 mmHg, and PVRI 8.0 Wood units m^2 (640 dyne $s/cm^5/m^2$). There were no improvements in hemodynamics with 100% inspired oxygen and inhaled nitric oxide (40 parts/million). With continuous inhaled epoprostenol (10 mcg/mL), the mPAP decreased to 69% systemic, TPG decreased to 42 mmHg, and PVRI to 6.0 Wood units m^2 (480 dyne $s/cm^5/m^2$). Prior to the catheterization, his pulse oximetry in clinic showed an increase to 90%. Before initiating treatment, at the age of seven yr, he performed 400 m in the six-min walk test.

He was treated with sildenafil reaching a maximum dose of 20 mg TID. Over the next two yr, when his echocardiogram and symptoms did not improve and with the development of headaches and erections, sildenafil was discontinued, and iloprost was started and advanced to 5 μ g five times a day. After one yr with no improvement in symptoms, saturations, or RVSP, plus the unwillingness of the family to use parenteral prostanoid, bosentan was added and uptitrated to 62.5 mg BID without adverse effects. A cardiac catheterization seven months later showed improvement in hemodynamics. The mPAP was 36 mmHg, TPG 28 mmHg, and PVRI 4.8 Wood units m^2 (384 dyne $s/cm^5/m^2$). There was minimal reactivity to oxygen and nitric oxide. Angiography continued to show diffuse and dilated small vessels in both lungs. There were no gross AVMs. The latest follow-up occurred at the age of 11 yr. He remains physically limited with poor growth and is hypoxemic with an oxygen saturation of 81%. The family continues to decline parenteral prostanoid. Sildenafil was restarted at a dose of 5 mg TID. His six-min walk test distance was unchanged at 395 m. The latest catheterization showed an mPAP 38 mmHg, TPG 32 mmHg, cardiac index 5.3 L/min/ m^2 , PVRI 7.3 Wood units m^2 (584 dyne $s/cm^5/m^2$). There was a decrease in the TPG and PVRI to 24 mmHg and 4.4 Wood units m^2 (352 dyne $s/cm^5/m^2$), respectively, under nitric oxide, but this was secondary to an increase in the pulmonary capillary wedge pressure. The cardiac index was normal and maintained under all conditions. Through his course, his symptoms, sub-maximal exercise capacity, and oxygen saturation have not improved, but his right heart hemodynamics have improved on triple PAH therapy of sildenafil, iloprost, and bosentan plus warfarin.

Case 2

This patient presented with abdominal situs inversus at two wk of age and received an echocardiogram to survey for congenital heart disease. She was found to have levocardia, azygous continuation of an interrupted IVC to the SVC, and a small atrial septal defect. There was also suggestion of delayed fall of pulmonary artery pressures because of right ventricular hypertrophy, dilated right ventricle and main pulmonary artery, and flattening of the interventricular septum. She exhibited respiratory distress with minor viral infections but was otherwise asymptomatic. To further define the magnitude of the atrial septal defect and PAH, she was catheterized at 14 months of age. A left-to-right shunt was not present, and her PVRI was 6.3 Wood units m^2 (504 dyne $\text{s}/\text{cm}^5/\text{m}^2$). She was referred for further investigation at a tertiary center at 30 months of age when she became less active. A complete pulmonary hypertension hemodynamics study showed her mPAP to be 37 mmHg (88% systemic), TPG 29 mmHg, and PVRI of 8.7 Wood units m^2 (696 dyne $\text{s}/\text{cm}^5/\text{m}^2$) without a left-to-right shunt. With administration of oxygen and 40 ppm of nitric oxide, her mPAP was unchanged, TPG increased to 38 mmHg, and PVRI to 7.9 Woods units m^2 (632 dyne $\text{s}/\text{cm}^5/\text{m}^2$). Her pulse oximetry was above 96%, and there was no evidence of dilated pulmonary vessels or AVMs. At four yr of age, a CT angiogram revealed polysplenia and absence of the main portal vein as well as the left and right intrahepatic portal vein branches, with the superior mesenteric and splenic veins draining via the azygous vein. Further confirmation of the abnormal venous anatomy was obtained by ultrasound and a transjugular retrograde hepatic venogram. She was treated at home with oxygen and sildenafil. She did not show clinical or echocardiographic improvement of RVSP on increased doses of sildenafil. At this time, a repeat catheterization showed her right heart hemodynamics to have worsened: mPAP 86% systemic, TPG 40 mmHg, and PVRI 12–14 Wood units m^2 with minimal reactivity to oxygen and nitric oxide. A liver biopsy showed pericentral sinusoidal fibrosis secondary to passive congestion, diminutive intrahepatic portal venules, minimal portal fibrosis, and microvesicular steatosis. She was started on intravenous treprostinil.

Six months later, repeat catheterization showed significant improvement in hemodynamics: mPAP 33 mmHg (75% systemic), TPG 25 mmHg, and PVRI 5.4 Wood units m^2 (432 dyne $\text{s}/\text{cm}^5/\text{m}^2$). Interestingly, she continued to not be reactive to nitric oxide. Shortly thereafter, at the age of five yr, she received a split left lateral segment liver transplant. The explanted liver confirmed no identifiable portal vein or main branches and diminutive portal venules on histology. During transplantation, a pulmonary artery catheter was placed to measure the pulmonary artery pressures. At the time of native liver explantation, the mPAPs were suprasystemic, and nitric oxide was added to treprostinil and sildenafil. The donor hepatic vein was anastomosed end-to-end with the vena cava. The donor portal vein was then anastomosed to the recipient dilated supramesenteric vein, followed by end-to-end anastomoses of the hepatic arteries. She was extubated on postoperative day two with mPAPs between 35 and 50 mmHg. Without hemodynamic compromise, she was discharged on prostanoil and silde-nafil. Repeat catheterization one month post-transplant did not show improvement in right heart hemodynamics. She subsequently developed intrahepatic biliary strictures, presumably secondary to ischemia related to her severe pulmonary hypertension at the time of transplantation (when PAPs exceeded systemic pressures for a prolonged period of time). As a prelude to re-transplantation, a catheterization was performed five months from the original transplant. At a treprostinil dose of 37 ngm/kg/min, her mPAP was 42 mmHg (60% systemic), TPG 32 mmHg, and PVRI 5.7 decreasing to 3.1 Wood units m^2 (248 dyne $\text{s}/\text{cm}^5/\text{m}^2$) with oxygen or nitric oxide for the first time. A successful whole liver transplant was performed one month later utilizing an aortoiliac graft for an inadequately small recipient hepatic artery found at transplant. The patient was discharged on postoperative day 13 without hemodynamic compromise during her post-operative course. Follow-up catheterization five months later showed stable mPAPs. By one yr post-

retransplant, she had weaned off of treprostinil. At that time, an echocardiogram showed a mildly flat septum with normal right atrial and ventricular size. Repeat hemodynamics showed the mPAP to be 30 mmHg (52% systemic), TPG 22, and PVRI 5.1 Wood units m^2 (408 dyne $s/cm^5/m^2$). Her liver allograft function was excellent in the first 1½ yr post-transplant. Unfortunately by two yr post-transplant, she had developed chronic allograft rejection and died from liver failure while awaiting retransplant. She had remained on sildenafil throughout her post-transplant course. An echocardiogram performed within a week of her death showed moderate septal flattening, mild right ventricle enlargement, and good bi-ventricular function.

Discussion

Congenital absence of the portal vein, also known as Abernethy syndrome, is a rare disorder with 19 cases reported since its original description in 1793 (2–4). Its association with pulmonary AVMs was described in three cases (5–7) in the more recent era but none were associated with PAH until a recent report of three additional cases (8). In the historical cases, additional features, such as a predominantly female disposition, polysplenia, biliary atresia, and interrupted IVC, suggest it may be associated with left isomerism heterotaxy syndrome. In fact, one has to wonder if other cases cited (9, 10) of dilated vessels and intrapulmonary shunting seen in patients with heterotaxy and polysplenia, may in fact have had absent portal veins that had gone unnoticed.

With broader identification and longer experience with the use of pulmonary arterial-specific vasodilators, medical management of PPHTN may now allow the safe performance of liver transplantation. Both patients in this report did not have serious adverse effects from pulmonary vasodilators. The headaches and erections were reversible and did not return when a smaller dose was used. Clearly, no strong conclusion can be made but neither patient seemed to have had a robust response to sildenafil or oxygen and nitric oxide in the cathlab. However, one may have responded to bosentan and the other to intravenous prostanoil. Interestingly, after transplantation, case 2 reacted for the first time to nitric oxide. In contrast, a case series of pediatric patients with PPHTN from Condino et al. (11) reported 4/7 pediatric patients succumbing to PAH in the setting of advanced liver disease or portal vein thrombosis. However, these subjects were older, did not present with vascular disease until years later, and had significantly longer follow-up. Krowka et al. (12, 13) provided hemodynamic guidelines to risk stratify cirrhotic patients with PPHTN for liver transplantation, and Arguedas et al. (14) and Krowka et al. (13) also described acceptable transplant outcome data for HPS. Hence, early diagnosis in conjunction with close monitoring for cyanosis and PAH (low threshold for catheterization) without delay in medical intervention may be the reason for the short-term success of liver transplantation, particularly in alleviating PAH, as seen in case 2. To this end, the work up of left isomerism heterotaxy syndrome should include identification of the portal vein. Alternatively, in the differential diagnosis of unexplained cyanosis or pulmonary hypertension, absent portal vein should be considered.

It may seem unusual for the portal vein to be absent in left isomerism heterotaxy syndrome because the spleen, a “left”-sided structure, and its vein are present (unlike right isomerism). The splenic vein still joins other intestinal veins that would otherwise be labeled as the portal vein if it were not for the connection to the azygous vein. Hence, the portal vein is clearly not absent or aplastic. Instead, in the setting of left isomerism, the contralateral side’s “right”-sided structures are absent so that the portal vein cannot home in to the portahepaticus and further divide into venules that are part of the portal triad. Perhaps, a better description of this vascular malformation in heterotaxy syndrome would be

“anomalous portal venous return,” conforming to the classification of other congenital vascular malformations such as “anomalous pulmonary venous return.”

In patients with portal-systemic venous shunts, the pulmonary manifestations are generally vascular dilatation or PAH in isolation. The rare presence of one entity followed by the other has also been described (15, 16). Their coexistence in liver disease, as described in case 1, is rare and described in only one other case series (8). In this report of three children with Abernethy syndrome, PAH coexisted with vascular dilatation and hypoxemia although the degree of each was not quantified. Other than supplemental oxygen, pulmonary vasodilators were not administered, and it is unclear how symptomatic these children were. Interestingly, a hypoplastic portal vein was identified in each of the patients by direct angiography, and all underwent portovenous shunt occlusion percutaneously with short-term improvement of hypoxemia and pulmonary hypertension in two obviating the need for transplantation or pulmonary artery-specific vasodilators. It is difficult to compare the severity of these two groups of patients, but the fact that they were treated very differently illustrates some important facets of the syndrome. First, it is difficult to image the mesenteric venous anatomy but a portal venous connection to the liver was identified by meticulous imaging, allowing the occlusion of the alternative venous pathway (portovenous shunt). Similar cross-sectional body imaging and direct angiography in case 1 plus direct visualization at liver transplant in case 2 did not reveal a portal venous connection to the liver. Either because the portal venous structure was never present or it became too small to recognize over time as the children in our case series were older at the time of diagnosing Abernethy syndrome compared to Newman et al. Second, a clinical response is seen when the shunt is eliminated, and the portal venous flow reestablished either by shunt occlusion or by liver transplantation, indicating the disease process is reversible. In fact, because there was no adverse outcome of shunt occlusion in the setting of a patent portal vein, this intervention should be attempted first with liver transplantation as backup. In the cases of congenital absence of the portal vein and pulmonary hypertension, liver transplantation should be considered as a primary option early in the disease course, so as to prevent the scenario of severe pulmonary hypertension (mean PAP greater than 2/3rd mean systemic arterial pressure) at the time of transplant. There remains a role for pulmonary arterial vasodilators. Because pulmonary hypertension is likely to be progressive in this disease, minimization of reversible and irreversible pulmonary vascular remodeling with medical therapy is indicated. As shown in this case series, medical therapy allowed the second case to receive a liver transplant alone with steady improvements in PAH before and after sequential liver transplants.

It may also seem paradoxical or at least pathologically divergent to have exuberant vascular dilatation in the same organ as vasoconstrictive disease. However, coexistence of at least AVMs and PAH is observed in patients with hereditary hemorrhagic telangiectasia (17). Although the exact mechanism is unclear, it may reside in the emerging role of the superfamily of TGF- β receptors in disease. *ALK1* and endoglin mutations are thought to be the molecular genetic basis behind hereditary hemorrhagic telangiectasia. Mutations in another TGF- β receptor superfamily member, *BMPR2*, have been identified in familial PAH. The TGF- β receptor gene itself is mutated in some patients with a syndrome of dilated vasculopathy that is now labeled as Loeys-Dietz (18). Similarly, there is suggestion that the fibrillin mutation can also lead to abnormal signaling in the TGF- β receptor in Marfan syndrome, in which the hallmark is a dilated vasculopathy of the ascending aorta (19). Therefore, one may speculate whether it is the regulation of TGF- β receptor stimulation/inhibition by mediators from the liver, dictated by whether the liver has the opportunity to perform first pass metabolism of portal venous blood that is important in the development of pulmonary vasculopathy, as opposed to an isolated imbalance of circulating vasodilators and vasoconstrictors. Although the description of the vascular malformation in patients with

Abernethy or left isomerism syndrome includes AVMs and HPS predominantly involves capillaries, when the disease process is diffuse, it is likely that precapillary/capillary dilatation and AVMs coexist. In case 1, for example, although small vessel involvement was clearly identified and no gross AVMs were seen, it is difficult to differentiate tiny or miliary AVMs from precapillary and capillary malformations alone. As a result, the cases of small diffuse AVMs in some reports and capillary dilatations in others may possibly involve both in the same patients.

It is also of interest that patients with congenital heart disease whose sole pulmonary blood supply is from a surgically created SVC anastomosis to the pulmonary artery (Glenn or cavopulmonary shunt) also have a disposition to the formation of dilated pulmonary vessels and/or AVMs (20). These “AVMs” tend to be very small and diffuse and not amenable to coil embolization. These patients have normal portal venous return to the liver. Addition of hepatic venous blood directly or unfiltered (not second pass such as after traversing another capillary bed) via a systemic arterial shunt to their pulmonary circulation will prevent these vascular malformations. Thus, it appears that portal blood that has undergone hepatic passage delivered to the lungs without traversing another tissue bed may be the circulatory prerequisite to mitigate the development of dilated vasculopathy and intravascular shunting or possibly even constrictive arteriopathy in the lungs.

Conclusion

The clinical manifestations of congenital absence of the portal vein, or Abernethy syndrome, have similarities to what is seen in cirrhotic liver disease: hypoxemia (HPS) and pulmonary arterial hypertension (PPHTN). The unexplained presence of these manifestations or anatomic anomalies suggestive of left isomerism heterotaxy syndrome requires an investigation into the portal venous anatomy. Meticulous identification of the mesentericportal venous anatomy will dictate treatment options, which may include liver transplantation alone. If PPHTN is present, it can be stabilized with pulmonary vasodilators, and this disease modification is likely to be critical to the success of liver transplantation.

Abbreviations

AVMs	arteriovenous malformations
BID	twice a day
HPS	hepatopulmonary syndrome
IVC	inferior vena cava
mPAP	mean pulmonary artery pressure
PAH	pulmonary arterial hypertension
PPHTN	portopulmonary hypertension
PVRI	pulmonary vascular resistance index
RVSP	right ventricular systolic pressure
SVC	superior vena cava
TID	three times a day
TPG	transpulmonary gradient

References

1. De Ville De Goyet J, Alberti D, Clapuyt P, et al. Direct bypassing of extrahepatic portal venous obstruction in children: A new technique for combined hepatic portal revascularization and treatment of extrahepatic portal hypertension. *J Pediatr Surg.* 1998; 33:597–607. [PubMed: 9574759]
2. Abernethy J. Account of two instances of uncommon formation in the viscera of the human body. *Philos Trans R Soc.* 1793; 83:59–66.
3. Howard E, Davenport M. Congenital extrahepatic portocaval shunts – The Abernethy malformation. *J Pediatr Surg.* 1997; 32:494–497. [PubMed: 9094026]
4. Wolfe JD, Tashkin DP, Holly FE, Brachman MB, Genovesi MG. Hypoxemia of cirrhosis: Detection of abnormal small pulmonary vascular channels by a quantitative radionuclide method. *Am J Med.* 1977; 63:746–754. [PubMed: 930949]
5. Alvarez AE, Ribeiro AF, Hessel G, Baracat J, Ribeiro JD. Abernethy malformation: One of the etiologies of hepatopulmonary syndrome. *Pediatr Pulmonol.* 2002; 34:391–394. [PubMed: 12357487]
6. Kinane TB, Westra SJ. Case 31-2004: A four-year-old boy with hypoxemia. *N Engl J Med.* 2004; 351:1667–1675. [PubMed: 15483286]
7. Cheung KM, Lee C, Wong CT, Chan AKH. Congenital absence of portal vein presenting as hepatopulmonary syndrome. *J Paediatr Child Health.* 2005; 41:72–75. [PubMed: 15670231]
8. Newman B, Feinstein JA, Cohen RA, et al. Congenital extrahepatic portosystemic shunt associated with heterotaxy and polysplenia. *Pediatr Radiol.* 2010 January 13. EPub.
9. Amodeo A, Di Donato R, Carotti A, Marino B, Marcelletti C. Pulmonary arteriovenous fistulas and polysplenia syndrome. *J Thorac Cardiovasc Surg.* 1994; 107:1378–1379. [PubMed: 8176990]
10. Kawata H, Kishimoto H, Ikawa S, et al. Pulmonary and systemic arteriovenous fistulas in patients with left isomerism. *Cardiol Young.* 1998; 8:290–294. [PubMed: 9731642]
11. Condino AA, Ivy D, O'Connor JA, et al. Portopulmonary hypertension in pediatric patients. *J Pediatr.* 2005; 147:20–26. [PubMed: 16027687]
12. Krowka MJ, Plevak DJ, Findlay JY, Rosen CB, Wiesner RH, Krom RA. Pulmonary hemodynamics and perioperative cardiopulmonary-related mortality in patients with portopulmonary hypertension undergoing liver transplantation. *Liver Transpl.* 2000; 6:443–450. [PubMed: 10915166]
13. Krowka MJ, Mandell MS, Ramsay MA, et al. Hepatopulmonary syndrome and portopulmonary hypertension: A report of the multicenter liver transplant database. *Liver Transpl.* 2004; 10:174–182. [PubMed: 14762853]
14. Arguedas MR, Abrams GA, Krowka MJ, Fallon MB. Prospective evaluation of outcomes and predictors of mortality in patients with hepatopulmonary syndrome undergoing liver transplantation. *Hepatology.* 2003; 37:192–197. [PubMed: 12500204]
15. Mal H, Burgiere O, Durand F, Fartoukh M, Cohen-Solal A, Fournier M. Pulmonary hypertension following hepatopulmonary syndrome in a patient with cirrhosis. *Hepatology.* 1999; 31:360–364.
16. Martinez-Palli G, Barbera JA, Taura P, Cirera I, Visa J, Rodriguez-Roisin R. Severe portopulmonary hypertension after liver transplantation in a patient with preexisting hepatopulmonary syndrome. *J Hepatol.* 1999; 31:1075–1079. [PubMed: 10604582]
17. Trembath RC, Thomson JR, Machado RD, et al. Clinical and molecular genetic features of pulmonary hypertension in patients with hereditary hemorrhagic telangiectasia. *N Engl J Med.* 2001; 345:325–334. [PubMed: 11484689]
18. Loeys BL, Chen J, Neptune ER, et al. A syndrome of altered cardiovascular, craniofacial, neurocognitive and skeletal development caused by mutations in TGFBR1 or TGFBR2. *Nat Genet.* 2005; 37:275–281. [PubMed: 15731757]
19. Neptune ER, Frischmeyer PA, Arking DE, et al. Dysregulation of TGF-beta activation contributes to pathogenesis in Marfan syndrome. *Nat Genet.* 2003; 33:407–411. [PubMed: 12598898]
20. Srivastava D, Preminger TJ, Lock JE, et al. Hepatic venous blood and the development of pulmonary arteriovenous malformations in congenital heart disease. *Circulation.* 1995; 92:1217–1222. [PubMed: 7648668]

Fig. 1. Selective right pulmonary arteriogram in case 1 shows diffuse dilated intraparenchymal pulmonary vessels with rapid return of contrast to left atrium, in less than two beats. The left pulmonary arteriogram had identical findings.

Fig. 2. Selective injection in the hepatic artery (upper) in case 1 with venous return demonstrating the large collateral draining to the azygous vein below the diaphragm (lower). The portal vein is not seen.

Fig. 3. Venous phase in Case 1 from injection into the hepatic artery with simultaneous balloon occlusion of the venous collateral. A portal venous stump was not seen on the liver side.