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The Intraoperative Finding of a Large Atrial Septum Defect Pre-Liver Transplantation: A Case Report

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Abstract: The case report discusses a significant discrepancy between the pre-transplant cardiac evaluation and post-induction transesophageal echocardiography, leading to aborting the surgical procedure. The transthoracic echocardiography [TTE] pre-liver transplant indicated only a minor intracardiac shunt, while the post-induction transesophageal echocardiography [TEE] showed a large atrial septal defect with a more significant left to right shunt. The attempt to close the defect with a transcatheter closure device failed and the perioperative care team decided not to proceed with the liver transplantation. Later cardiac evaluation with cardiac magnetic resonance imaging demonstrated a secundum atrial septum defect in the inferior border of the interatrial septum with 43 ml flow difference between systemic and pulmonary circulation. The failure to identify a large atrial septal defect/inferior venosus defect in the preoperative screening process for liver transplantation resulted in failing to optimize the patient pre transplant. Although complications related to large atrial septal defects are rare during liver transplant (paradoxical embolisms, malignant arrhythmias, myocardial infarctions or cerebrovascular complications), they can be potentially fatal. The preoperative cardiac assessment should have included optimization of this congenital defect. The case report discusses implications of atrial septal defects for liver transplant and why the screening process may have missed the defect. The findings of a positive bubble study during the transthoracic echocardiography should not be trivialized and may require further workup.

Keywords: Liver Transplant, Atrial Septal Defect, Transthoracic Echocardiography, Transesophageal Echocardiography, Pre-liver Transplant Screening, Preoperative Optimization

1. Introduction

Although liver transplantation remains a high-risk procedure, the presence of an atrial septal defect (ASD) or a patent foramen ovale (PFO) in a patient who is to undergo this procedure is often seen as a trivial finding and one without relevance for perioperative mortality. However, not every ASD is as insignificant as a PFO, and preoperative transthoracic echocardiography (TTE) may lead one to underestimate the size and shunt fraction of such defect. The following presented case involves a postinduction transesophageal echocardiogram (TEE) finding with significant impact on the related surgical plan [3, 6, 22].

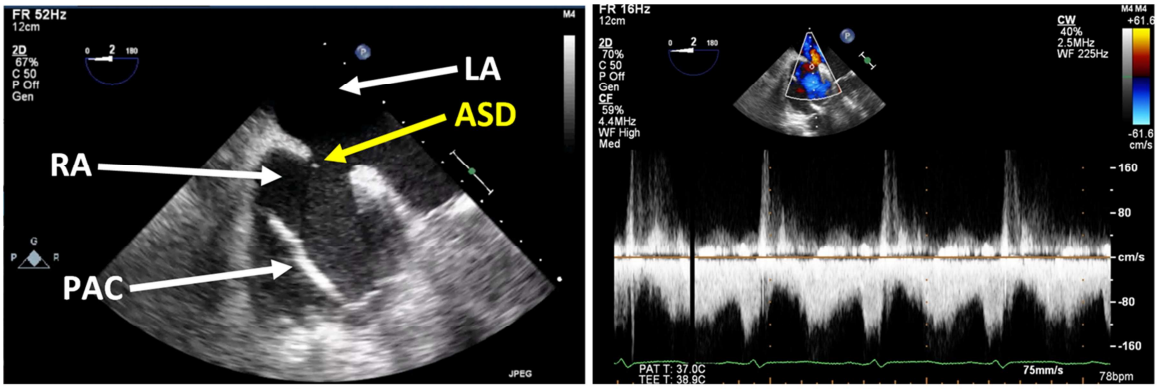
2. Case Report

In December 2018, a fifty-nine-year-old male patient presented to our institution with progressive shortness of breath due to ascites. Liver dysfunction due to polycystic liver disease and nonalcoholic steatohepatitis (NASH) in the patient had been known for several years. However, formal diagnosis and treatment were only recently initiated. At presentation, the patient's Model for End-stage Liver Disease (MELD) score was 17 (based on an International Normalized Ratio of 1.6, creatinine levels of 1.45 mg/dL, bilirubin levels of 1.3 mg/dL and sodium levels of 145 mEq/L). The medical history included the diagnosis of coronary artery disease with myocardial infarction and Percutaneous Coronary

Intervention with a drug-diluting stent in 2013, atrial fibrillation and hypertension. Anticoagulation was held because of recurrent upper gastrointestinal bleeding episodes. Dyspnea, due to the progressive ascites development, improved with weekly large-volume paracentesis. Liver transplantation eligibility assessment was initiated, and the patient was compliant with monthly liver transplant clinic visits and pre-transplantation screening procedures.

2.1. Preoperative Optimization

The screening TTE in January 2019 indicated a normal right and left ventricular size and function. The left atrium was mildly dilated and the results of a bubble study were immediately positive, suggesting an intracardiac shunt without a significant dynamic component. Additionally, the right ventricular systolic pressure was found to be elevated (35-40 mmHg). After a cardiology consultation, the patient underwent a right and left heart catheterization in March 2019, to assess right heart pressures and coronary circulation. The right heart catheterization indicated normal right heart and pulmonary artery pressures (right ventricle pressure at 28/1 mmHg, pulmonary artery pressure at 26/8 mmHg, and pulmonary artery occlusion pressure at 11 mmHg), and normal cardiac output (4.9 l/min/m2). However, systemic vascular resistance (630 dyn·s/cm^5) and pulmonary vascular resistance (53 dyn·s/cm^5) were representative for hemodynamics of end-stage liver disease. In addition, a coronary angiography indicated moderate nonobstructive coronary artery disease with patent stents in the left anterior descending coronary artery and right anterior descending coronary artery. It was deemed acceptable to proceed with liver transplantation due to the progression of liver disease and declining renal function in the patient. Their MELD score in July 2019 was 25 (based on an International Normalized Ratio of 1.5, creatinine levels of 2.3 mg/dL, bilirubin levels of 4.5 mg/dL and sodium levels of 138 mEq/L). Then, on August 30, 2019, a suitable donor organ became available and the patient presented for liver transplantation.

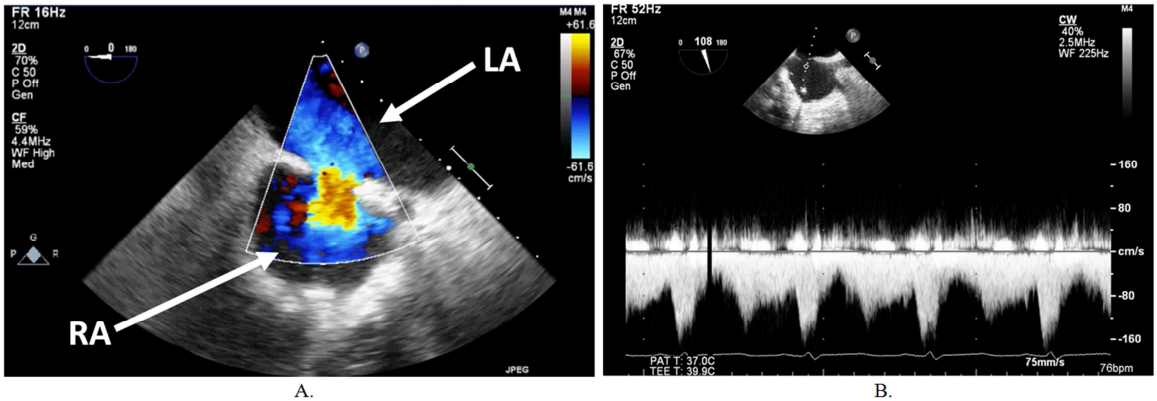


A. B.

Figure 1. *Transesophageal Echocardiography Images Obtained After Anesthesia Induction.*

A. This image is a midesophageal four-chamber view that is focused on the right atrium and ventricle. The arrow labeled "RA" the right atrium, the arrow labeled "PAC" points to the artifact created by the pulmonary artery catheter, the arrow labeled "LA" points to the left atrium, and the yellow arrow points to an atrial septal defect (with its size in systole).

B. This image is of continuous wave Doppler that indicates almost constant left-to-right flow across an atrial septal defect. The image involves a similar probe position as that which is involved in A., and it provides a view that is similar to that which is in A.



A. B.

Figure 2. *Transesophageal Echocardiography Images Obtained After Induction.*

A. This image is a midesophageal bicaval view. The arrow labeled "RA" points to the right atrium, the arrow labeled "LA" points to the left atrium, and the included color Doppler imaging indicates left-to-right shunt through an atrial septal defect.

B. This image is of continuous wave Doppler that has been applied through an atrial septal defect, and it demonstrates continuous left-to-right flow.

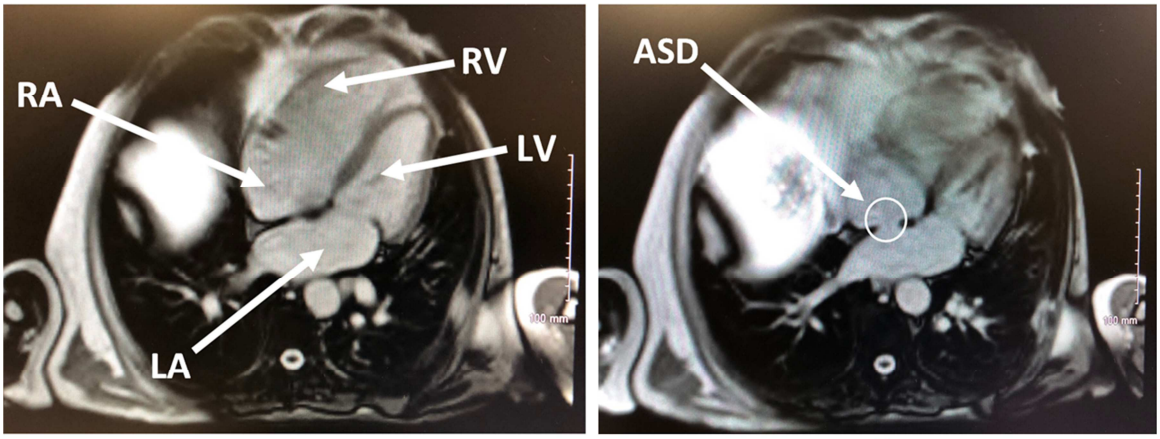
2.2. Intraoperative Course

After anesthesia induction and invasive line placement, the anesthesia team on this case placed a TEE probe for pre-transplantation function assessment and perioperative monitoring, according to the institutional standard for liver transplantation. The TEE imaging showed a large ASD with a significant left-to-right shunt. (See Figure 1 and Figure 2.) Cardiology specialists were consulted intraoperatively to discuss the significance of this finding, as well as treatment options. Based on multidisciplinary discussions between the surgical, anesthesia and cardiology teams, and with the cold ischemia time of the donor organ in consideration, it was decided to immediately transfer the patient to the interventional cardiology suite—to close the ASD. The patient remained intubated and arrived in the catheterization laboratory two hours after anesthesia induction.

ASD closure was attempted using femoral venous access for Amplatzer device placement. Using intracardiac echocardiography for guidance, the ASD in the patient was measured to be approximately 32 mm in diameter, and it was noted that this defect extended inferiorly and posteriorly into a region that was poorly visualized by TTE and transesophageal echocardiography. Due to the lack of sufficient inferior and posterior margins, suggesting an inferior sinus defect, this ASD was not amendable to transcatheter closure and this interventional approach was aborted. Cardiothoracic surgery specialists were consulted to review the advisability of surgical closure, and it was decided to defer surgical closure. Furthermore, the surgical team and anesthesia team decided to abort the liver transplantation for the patient; the patient was brought to the recovery room for anesthesia emergence and the available donor organ was offered to the backup recipient.

2.3. Postoperative Work up

In September 2019, the patient in this case underwent cardiac magnetic resonance imaging for further assessment and planning of possible treatment options. The use of this diagnostic test confirmed the presence of a secundum ASD in the inferior border of the interatrial septum with left-to-right flow and severe biatrial enlargement. (See Figure 3) The shunt fraction (Q_p/Q_s) calculation was 1.4, representing a 43 ml flow difference between systemic and pulmonary circulation.



A. B.

Figure 3. *Images from Cardiac Magnetic Resonance Imaging.*

A. This CINE_4CH image depicts the four chambers as marked with arrows (an arrow labeled "RA" points to the right atrium, an arrow labeled "LA" points to the left atrium, an arrow labeled "RV" points to the right ventricle, and an arrow labeled "LV" points to the left ventricle), showing a dilated right atrium and ventricle.

B. This image is a modified CINE_3CH view, showing an atrial septal defect.

3. Discussion

This case report describes the intraoperative finding of a large ASD, which altered the perioperative risk involved in a scheduled orthotopic liver transplantation. Therefore, this procedure was cancelled. The first point of discussion is whether the liver transplantation could have been performed with acceptable perioperative risk. The second point of discussion leads to an exploration of why pre-transplantation screening procedures did not identify the aforementioned congenital intracardiac abnormality and thus why it was not considered when conducting an initial risk assessment. The third point of discussion concerns systemic modifications that could optimize the transplantation screening process, to prevent similar situations.

The perioperative team deemed the perioperative risk for this patient unacceptable considering the significant left-to-right shunt and the potential for catastrophic complications, should the shunting reverse direction. The risk assessment of ASDs in the liver transplantation procedure is difficult because serious complications are rare but potentially fatal; case reports have described paradoxical embolisms, malignant arrhythmias, myocardial infarction and

cerebrovascular complications [8, 11, 13-15, 17, 23]. However, several recent publications have documented that the presence of an ASD does not negatively impact the outcomes after liver transplantation [3, 22]. For example, increased perioperative mortality was not seen in pediatric patients with complex congenital heart disease and ASDs [6]. The controversy arising from these findings may be related to the fact that embolization risk may vary depending on the size and location of the ASD [19], and grading the impact of ASDs on perioperative mortality only by the presence and severity of a positive bubble study on TTE may not be sufficient [3, 22]. The current American Heart Association guidelines for perioperative management of patients with congenital heart disease separate small, isolated ASDs (simple CHD) from moderate or large ASDs, as well as from sinus venosus defects (moderate complexity CHD) [10]. While sinus venosus defects and secundum ASDs are much less common than PFOs, with PFOs being reported as prevalent in approximately 26-30% of the general population, the TTE finding of a positive bubble study should be neither automatically attributed to hyperdynamic circulation nor an intrapulmonary shunt that is due to end-stage liver disease or that is related to a PFO [10].

This leads one to the following question: why did the pre-transplantation screening not result in a detailed and accurate description of the intracardiac abnormality in this case? A TTE found an intracardiac shunt, but based on the amount of crossover bubble, its size was underestimated and its location was not identified. While cardiology specialists performed left and right heart catheterization for right heart function, pulmonary vascular resistance and coronary circulation, no further investigation concerning this intracardiac shunt was deemed necessary because of the trivial shunt amount. Additionally, the aforementioned intraoperative TEE did not lead to full consideration of the posterior location of the intracardiac shunt. Though using transesophageal echocardiography (TEE) is seen as a superior method for identification, the diagnosis of defects like this is difficult through its use [5, 9, 16, 20]. Furthermore, ASDs that are posterior, relatively complex, or both may call for more advanced imaging options than TEE (such as contrast-enhanced computed tomography or cardiac magnetic resonance imaging) [1, 7]. In this case, TEE was not sufficient; it did not enable one to adequately describe the location and extent of the defect that was present. Under the assumption that a secundum ASD was present, an attempt was made to close the defect with a transcatheter device. However, inferior sinus venosus defects are not amenable for transcatheter device closure due to the absence of inferior margins, so it was ultimately determined that the remaining treatment option for this patient was open surgical repair prior to liver transplantation [2, 4, 12].

The cardiovascular screening for risk assessment regarding liver transplantation candidates includes only TTE to evaluate for structural heart disease and function [18, 21]. This test is sufficient in identifying a PFO, but it is not reliable when used to characterize more complex ASDs, and it is not dependable when used to quantify the severity of a shunt—as demonstrated through this case report. Werlang et al suggest the use of patient symptomology to determine further need for diagnostic testing, which is a suggestion that is based on the findings of their retrospective analysis on this topic [22]. The origin of dyspnea or cerebral hypoperfusion may be difficult to define in advanced liver disease. Since the surgical team usually initiates multiple imaging procedures (involving abdominal computed tomography scanning, magnetic resonance imaging, or both) and uses the produced images to plan the surgical approach, it may be reasonable to broaden the utilization of computed tomography or magnetic resonance imaging to screen for congenital cardiac abnormalities.

4. Conclusion

In summary, this case report documented the failure to identify a large ASD/inferior venosus defect during the preoperative screening process for liver transplantation. The perioperative care team decided not to proceed with the procedure since the significance of this finding and possible implications on postoperative outcome could not be discussed with the patient after anesthesia induction. The findings of a positive bubble study during a transthoracic echocardiogram should not be trivialized and may require further diagnostic examination of the patient.

Note

The patient provided written consent to use the incorporated data and images for publication.

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