Hypoxemia Explained 36 Years Later*

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A 37-year-old man who had an atrial septal defect (ASD) corrected as an infant was found to be hypoxemic with a 22% shunt. An MRI scan revealed that the patient’s inferior vena cava drained into his left rather than his right atrium, a previously undetected complication of his ASD repair 36 years before. (CHEST 2001; 120:1739–1740)

Key words: anomalous venous return; cardiac surgery; congenital heart disease; hypoxemia

Abbreviations: ASD = atrial septal defect; LA = left atrium; IVC = inferior vena cava; RA = right atrium; RV = right ventricle

A left-sided superior vena cava entering the left atrium (LA) is a frequent cause of right-to-left shunt in patients with congenital heart disease, but the presence of an inferior vena cava (IVC) draining into the LA is rare. We present a case of a complication arising from the repair of an atrial septal defect (ASD), a complication undetected for 36 years.

Case Report

A 37-year-old Hispanic man who had been admitted to the hospital with acute hepatitis A was referred for evaluation of hypoxemia. The results of arterial blood gas measurements made while the patient was breathing room air were as follows: pH, 7.53; PO2, 21 mm Hg; PAO2, 55 mm Hg; and alveolar-arterial oxygen pressure difference, 41 mm Hg (at 5,250 feet altitude). The patient denied respiratory complaints, had smoked tobacco for 10 years, and had worked in a uranium mine for 7 years. He denied the presence of other pulmonary diseases and was receiving no medications. He had had open-heart surgery at age 10 months and had not seen a physician in 23 years.

The patient was jaundiced without respiratory distress, and the results of a pulmonary examination were normal. A transverse scar extended under both nipples, but a cardiac examination revealed normal heart sounds without a murmur. No peripheral edema was evident, but there was marked digital clubbing. The patient’s hematocrit was 46% with normal serum electrolytes, and IgM antibodies to hepatitis A were present. A chest radiograph had normal findings.

While breathing 100% oxygen in a closed system, the patient’s PAO2 was 262 mm Hg, indicating a shunt of 22%. The results of pulmonary function tests were normal except for a reduced diffusing capacity (diffusing capacity of the lung for carbon monoxide/alveolar volume ratio, 72% of predicted). A cardiac echocardiogram with bubble study revealed normal heart valves and chambers and no shunt. A CT scan of the chest showed normal lung parenchyma and a markedly enlarged azygous vein.

Medical records from age 6 months noted that the patient had failure to thrive and a loud thrill in the midclavicular line. Fluoroscopy revealed a large right atrium (RA) and right ventricle (RV). Right and left heart catheterization from the femoral vessels revealed RV systolic and diastolic pressures of 55 mm Hg and 3 mm Hg, respectively, arterial saturation of 75%, LA saturation of 80%, and an RV saturation of 80% with pulmonary blood flow 2.5 times the systemic flow (which is consistent with a bidirectional shunt). He underwent hypothermic open-heart surgery in August 1956, at which time an ASD was identified. It was also determined that the left pulmonary veins entered the coronary sinus (the right pulmonary veins entered the LA normally). The ASD was repaired, and the coronary sinus was transplanted into the LA.

A right heart catheterization from the saphenous vein 3 months postsurgery revealed peripheral PAO2 of 95% with an RA PAO2 of 92%. The catheter could not enter the superior vena cava but repeatedly passed into two separate pulmonary veins from the RA. It was concluded that there was a persistent left-to-right shunt due to inadequate closure of the ASD. Since the patient was doing well, repeat surgery was postponed.

At age 13 years, he developed perioral cyanosis during exercise. A repeat catheterization (done from the right brachial vein) revealed normal RA, RV, and pulmonary artery pressures. RA, RV, and pulmonary artery oxygen saturations were all 85%. With leg exercise, the patient’s arterial saturation fell to 85%. Doctors concluded that he had a right-to-left shunt due to the transplanted coronary sinus.

His next medical contact was during the current hospital admission. Because of uncertainty about the etiology of his shunt, a cardiac MRI scan was obtained. In addition to the enlarged azygous vein, it revealed that the IVC drained into the LA (Fig 1, top left, A).

Discussion

This patient’s IVC did not appear to enter the LA at the time of his first catheterization or at surgery. In 1956, a woman’s IVC was inadvertently diverted into the LA when the surgeon incorporated the eustachian valve of the IVC into the repair of her low-lying ASD. Immediately following the surgery, the patient became cyanotic. Catheterization showed that the IVC drained into the LA. A second procedure corrected the defect.

Since then, reports3–5 of similar IVC diversions occurring during ASD repairs have been published. Most patients had a low ASD that was closely associated with the IVC, and most procedures were performed under mild hypothermic conditions, which limits the operating time. Usually, the eustachian valve of the IVC was incorporated into the septal repair, which then isolated it from the RA. Initially, many of these patients had resolution of pulmonary venous congestion and right ventricular strain (as expected with the correction of their left-to-right shunt). Later on, they developed dyspnea with exertion, fatigue, and exercise-induced cyanosis. These patients were thought to have partial diversion of the IVC into the LA initially, but with progressive scarring and retraction of the repair a complete diversion was created. In other patients, the diversion was likely to be associated with stenosis of the IVC with the subsequent development of collaterals, presumably through the azygous veins, which reduced the

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degree of shunting. Most of those eventually identified (8 years postoperatively was the longest duration reported) were taken for surgical correction.3–5

This patient had hypoxemia due to anomalous return of the IVC to the LA, which was an unintended result of his surgery 36 years before. The diversion either was partial or was associated with a significant stenosis, which would explain the absence of a catastrophic postoperative course and the presence of an enlarged azygous vein. It also explains the confusing catheterization results obtained 3 months postsurgery in which the operators encountered well-oxygenated blood, continually entered the pulmonary veins from what they thought was the RA, and incorrectly concluded that the ASD had not been completely repaired. The repeat catheterization performed at age 13 years appeared more normal since it was performed from the brachial rather than the femoral vein.

The patient refused to have the defect corrected despite our concerns about the adverse effects of his chronic hypoxemia as well as the risks of potential CNS embolism or brain abscess due to the direct connection of the venous and arterial circulation.

This difficult and complicated operation appeared, at the end, to have been technically successful, and we will await with interest the outcome for this patient following this degree of disability at the time of operation “(from the original operating surgeon; August 13, 1956).”

REFERENCES