Iatrogenic cyanosis and clubbing: 25 years of chronic hypoxia after the repair of an atrial septal defect

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M Alanbaei, L Jutras, J Therrien, A Marelli. Iatrogenic cyanosis and clubbing: 25 years of chronic hypoxia after the repair of an atrial septal defect. Can J Cardiol 2007;23(11):901-903.

A case of sinus venosus atrial septal defect repair of the inferior vena caval type with an unintentional diversion of the inferior vena caval blood to the left atrium is reported. Long-standing, anatomical, right-to-left shunting with cyanosis and hypoxia are associated with systemic and cerebrovascular complications. Cardiac risks depend on the presence or absence of pulmonary hypertension, the associated hematological abnormalities and the degree of anatomical, right-to-left shunting. Cardiac magnetic resonance imaging clarified the etiology of the unexplained cyanosis and delineated the surgical anatomy.

Key Words: *Congenital heart disease; Hypoxia; Magnetic resonance imaging; Septal defects; Shunt*

Cyanose iatrogène et hippocratisme : 25 ans d'hypoxie chronique après la correction d'une communication interauriculaire

Il sera question, dans le présent article, d'un cas de correction d'une communication interauriculaire de type sinus veineux de la veine cave inférieure, qui a donné lieu à un détournement involontaire de sang de la veine cave inférieure à l'oreillette gauche. Les shunts anatomiques droite-gauche, de longue date, accompagnés de cyanose et d'hypoxémie sont associés à des complications périphériques et vasculaires cérébrales. Les risques de complications cardiaques dépendent de la présence ou non d'hypertension pulmonaire, d'autres anomalies sanguines et du degré d'importance du shunt droite-gauche par la structure anatomique. L'imagerie par résonance magnétique a permis de découvrir la cause de la cyanose inexpliquée et de préciser la structure anatomique chirurgicale.

Iatrogenic, anatomical, right-to-left shunting is a rare complication of repaired acyanotic congenital heart disease. We report a case of an unintentional connection of the inferior vena cava (IVC) to the left atrium (LA), discovered 25 years after the repair of a sinus venosus atrial septal defect (ASD) of the IVC type.

CASE PRESENTATION

A 30-year-old woman was referred for ovarian cystectomy because of severely symptomatic endometriosis. She was found to be cyanotic and was referred to the McGill Adult Unit for Congenital Heart Disease Excellence at the McGill University Health Centre, Montreal, Quebec, for cardiac preoperative assessment. She denied any chest discomfort, dyspnea or hemoptysis. She denied symptoms of hyperviscosity and was in New York Heart Association functional class I. Her medical history revealed open-heart surgery in another country at the age of five years for a "hole in the heart". Cyanosis and clubbing were long-standing. In 1992, the patient had an episode of transient altered mental status, which was thought to be a small embolic stroke without neurological sequelae, secondary to a hypercoagulable state due to smoking and the use of contraceptive pills. The patient immigrated to North America in 2000. In 2001, she reported another episode of altered mental status for which she underwent a series of investigations, including a transthoracic echocardiogram (TTE), transesophageal echocardiogram (TEE), and a right and left heart catheterization.

On physical examination, the patient had no dysmorphic features. She had moderate central and peripheral cyanosis. The physical examination showed stage 3 clubbing with a drumstick appearance of the fingers and toes. Her arterial oxygen saturation was 84% on room air, her pulse was 82 beats/min and her blood pressure was 120/80 mmHg in the right and left arms at rest. A cardiac examination revealed a normal apical impulse, no right ventricular heaves or thrills, and normal first and second heart sounds. A chest examination was unremarkable and no lower limb edema was noted.

The initial laboratory data included serum hemoglobin of 177 g/L and hematocrit of 51%. Serum iron and electrolyte levels and renal function were normal. The electrocardiogram and chest x-ray were within normal limits. A review of the results of the previous cardiac catheterization performed elsewhere in 2001 revealed measured systemic right ventricular pressures with no significant left-to-right shunting.

A TTE was performed and showed normal left and right ventricular function and dimension. No tricuspid insufficiency jet was identified from which a systolic pulmonary arterial pressure could be estimated; however, estimations of the mean and diastolic pulmonary arterial pressures using the acceleration time and pulmonary regurgitation, respectively, were normal. There were no secondary signs of pulmonary hypertension (ie, right ventricular hypertrophy or dilation). No residual shunt or persistent left superior vena cava was seen. The IVC was not observed draining to the right atrium.

At this point, causes of right-to-left shunting without evidence of pulmonary hypertension were sought. These included anatomical right-to-left shunts that are cardiac or extracardiac.

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Received for publication August 18, 2005. Accepted February 16, 2006

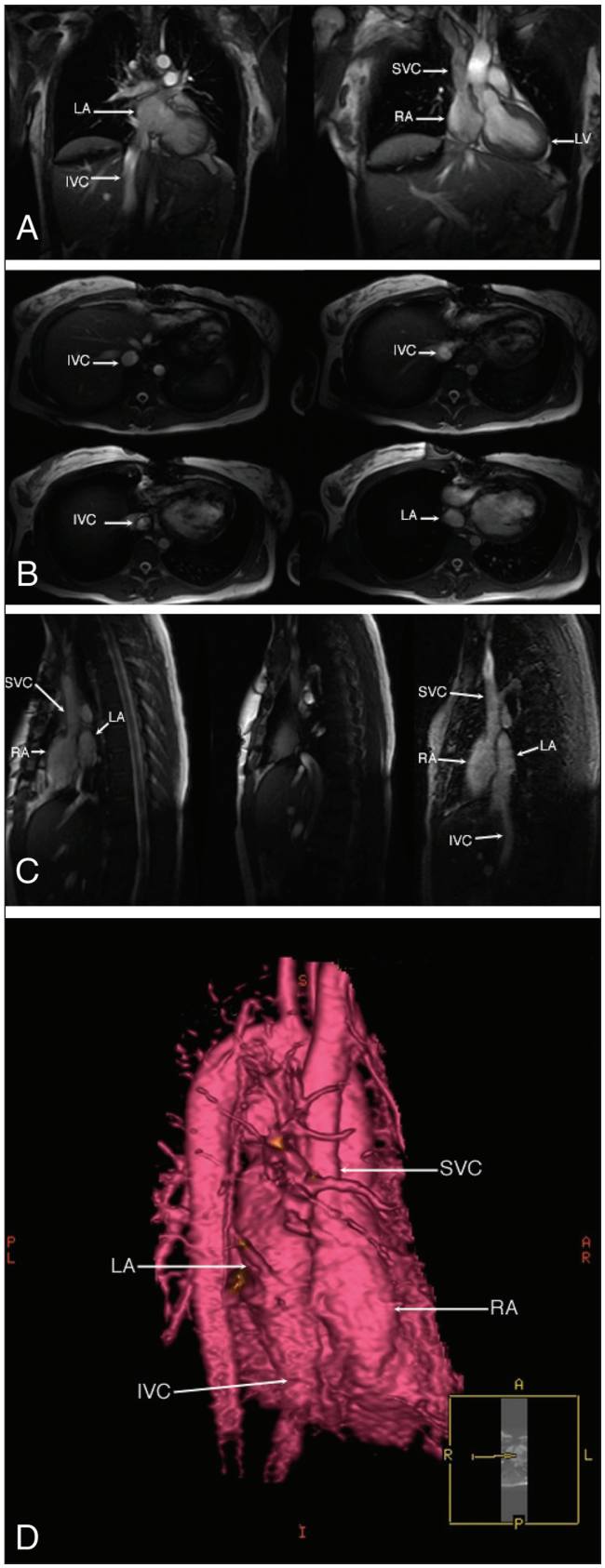

Figure 1) *Magnetic resonance imaging of a patient with iatrogenic cyanosis and clubbing.* **A** *Coronal section;* **B** *Horizontal section;* **C** *Sagittal section;* **D** *Three-dimensional reconstruction. A Anterior; IVC Inferior vena cava; L Left; LA Left atrium; LV Left ventricle; P Posterior; R Right; RA Right atrium; SVC Superior vena cava*

Cardiac magnetic resonance imaging angiography was performed to visualize the anatomy and revealed IVC drainage to the LA with unobstructed flow. There were no large pulmonary arteriovenous malformations. A small leak across an atrial septal patch repair was the cause of an insignificant left-to-right shunt (Figure 1).

A review of the records of the right and left heart catheterization originally performed in another hospital revealed that the catheter introduced into the right femoral vein had entered the LA from the IVC. As a result, the left-sided measurement was falsely designated systemic right ventricular pressure. The patient refused to preoperatively undergo a cardiac catheterization to document the absence of pulmonary hypertension.

Because the patient was very symptomatic with endometriosis and there was no evidence of pulmonary hypertension by clinical or echocardiographic assessments, she was cleared to undergo a mini-laparotomy with a right oophorectomy under general anesthesia. Intraoperatively, a Swan-Ganz catheter documented normal pulmonary arterial pressure, and a TEE with a modified transgastric view showed the IVC connection to the LA. Agitated saline injected into the right arm showed no evidence of premature appearance into the LA to suggest a pulmonary atrioventricular fistula.

The postoperative course was complicated by significant intra-abdominal bleeding, which was controlled with the transfusion of blood products. The patient was stable and was discharged three days later. Although elective admission for intracardiac repair was recommended and planned, the patient decided to return to her native country.

DISCUSSION

Similar earlier cases have been reported with iatrogenic diversion of IVC flow to the LA after a surgical incorporation of the Eustachian valve of the IVC in the repair of a low-lying ASD (1-4). This complication was more frequent before the use of cardiopulmonary bypass because time limitations were imposed by only hypothermia and no inflow occlusion (5). These cases are usually discovered soon after surgery when the patient becomes cyanotic and hypoxic; however, some factors, such as the following, may mask the discovery of such complications: the relief of pulmonary venous congestion and right ventricular strain by the correction of the left-to-right shunt; the occurrence of only partial diversion of the IVC flow to the LA; and the occurrence of stenosis of the IVC, with collaterals draining to the superior vena cava through an azygos vein.

To our knowledge, there are two similar cases of ASD discovered late in a patient's adulthood (6,7). The present case was initially misdiagnosed in another institution as Eisenmenger's syndrome after a series of tests, including a TTE, a TEE and a right and left heart catheterization, showing the rarity of this condition. But with the advances in cardiac imaging and more awareness of this complication, these lesions are now being discovered earlier.

The perioperative care of the patient presently reported for a noncardiac surgery requires special attention to several factors: cardiac risk, which depends on the presence or absence of heart failure, pulmonary hypertension, valvular dysfunction or uncontrolled arrhythmias; hematological risk, which should be assessed preoperatively to avoid excessive bleeding or thrombosis with paradoxical emboli; and risk caused by the anatomical right-to-left shunt, which includes thrombotic, air or septic paradoxical emboli.

Prophylactic preoperative phlebotomy for cyanotic patients is recommended if hematocrit is more than 65%. Early mobilization and the use of elastic stockings to prevent deep venous thrombosis

with paradoxical thrombotic embolism are recommended. Intravenous lines should be used with air filters to reduce the risk of a paradoxical air embolism in patients with right-to-left shunts. Corrective cardiac surgery was strongly recommended to this patient to avoid long-term complications of chronic cyanosis. Meanwhile, we advised the patient against pregnancy or the use of estrogen-containing oral contraceptives.

CONCLUSION

Cardiologists and cardiac surgeons should be aware of this rare but potentially important complication of what should be a simple ASD repair surgery. Cardiac magnetic resonance imagery is an excellent tool to identify anatomical abnormalities that may otherwise be missed.

Cyanotic patients with long-standing, anatomical, cardiac right-to-left shunting preparing for a noncardiac surgery require special perioperative care to avoid major complications.

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