Bol Med Hosp Infant Mex 2013;70(3):244-252

CLINICOPATHOLOGICAL CASE

Tricuspid regurgitation in a child after closure of a ventricular septal defect

Sergio Ruiz González,[1] Alexis Arévalo Salas,[2] Jorge Luis Villatoro Hernández,[2] Ma de Lourdes Marroquín Yáñez,[3] Begoña Segura Stanford,[2] Julio Erdmenger Orellana,[4] Marco Antonio Sarmiento Abril,[5] Mario Pérezpeña-Díazconti[6]

SUMMARY OF THE CLINICAL HISTORY (A-10-09)

We present the case of a 4-year-old female admitted for surgical correction of a tricuspid valve defect.

Perinatal and pathological history. The patient was the product of G IV with a hospital birth at 37 weeks gestation. Birth weight was 3350 g, length 51 cm, Apgar unknown. Delivery was without complications. The patient had chicken pox at 1 year of age without complications. In 2005, the patient was diagnosed with persistence of patent ductus arteriosus (PDA) and interventricular communication (IVC) at 2 years of age at a third-level hospital. Catheterization detected pulmonary vascular disease and biventricular dysfunction. The patient was not considered to be a surgical candidate and was managed with sildenafil (0.5 mg/kg/dose) every 8 h and continuous oxygen.

September 6, 2007 (outpatient cardiology consultation). The patient presented for the first time to the Hospital Infantil de México Federico Gomez (HIMFG) for congenital cyanogenic heart disease, mild exercise-related dyspnea, and NYHA functional class II. An echocardiogram revealed situs solitus, levocardia, normal venous and systemic return, A-V connection and concordant arterial ventricle. Pulmonary ring was 20 mm, right branch of the pulmonary artery was 13 mm, left branch of the pulmonary artery 12.4 mm, right ventricular systolic pressure 75 mmHg with oxygen decreases to 56 mmHg, IVC due to 14 x 10 mm misalignment, IVC gradient of 12 mmHg with bidirectional flow, 6-mm PDA pulmonary opening, aortic gallop of 30%, left aortic branch without obstruction, and tricuspid prolapse. The patient was administered furosemide, spironolactone and captopril and during October 2007 had a community-acquired pneumonia treated with amoxicillin at 150 mg/kg/day without complications.

October 21–November 13, 2007 (cardiovascular surgery). The patient is aged 4 years, weight 11.5 kg and height 95 cm. Surgical closure of the IVC, ligation of the arterial duct and tricuspid surgery with plication of the tendinous cord of the anterior valve were carried out. An infundibular IVC of 15 mm was found, PDA of 1 cm, and severe tricuspid insufficiency due to elongation of the tendinous cords. Aortic clamping was carried out for 1 h. Perfusion time was 1 h 40 min. Echocardiogram demonstrated residual IVC of 3.3 x 4.4 mm with bidirectional flow, moderate to severe tricuspid insufficiency, and systolic pressure of the right ventricle 48 mmHg. She was discharged due to good progression, with spirinolactone,

[1] Sevicio de Cirugía Cardiovascular
[2] Servicio de Hemodinamia
[3] Servicio de Terapia Intensiva
[4] Servicio de Cardiología
[5] Departamento de Radiología
[6] Departamento de Patología

Hospital Infantil de México Federico Gómez, México D.F., México

Received for publication: 3-21-13
Accepted for publication: 3-26-13

furosemide and captopril. In July 2008, at 5 years of age, she presented with hepatitis A. She was treated conservatively and there were no complications.

June 11, 2009 (cardiology). The patient was presented to the Joint Session because she continued with NYHA class II functional capacity and lack of weight gain due to residual IVC, severe tricuspid insufficiency, and obstruction of the right pulmonary veins by the interatrial septum. It was decided to perform a tricuspid repair (or replacement) and patch in the interatrial septum (rigid). Postoperative risk was high due to pulmonary hypertension.

Current illness. The patient was admitted February 7, 2010 for scheduled surgical correction. On physical examination she weighed 14.9 kg, height 95 cm, cardiac frequency (CF) 89/min, respiratory frequency (RF) 20/min, blood pressure (BP) 90/50 mmHg, temperature 36.5 °C, and Glasgow score 15.

The female patient is intact and appears to be her chronological age without characteristic facies: skull without step-off or exostosis, eyes symmetrical, isochoric 3-mm pupils, normal reflexes, reactive to light. Permeable nares, well-set ear lobes, permeable ear canals, and tympanic membranes intact without alterations are observed. The oral cavity demonstrated moist mucous membranes, hyperemic pharynx, and without postnasal discharge. Neck was without changes. Chest was normal with adequate respiratory movements, good intake and exit of air, and without pleural pulmonary syndrome. Heart sounds were without systolic murmur, grade III/VI in the fourth left intercostal space. Abdomen with liver edge at 2-2-2 cm of the right costal margin and normal peristalsis. Female genitalia, Tanner I, without changes. Pulses palpable in four extremities, tone and strength preserved. Cranial nerves without changes. O2 saturation between 88 and 92%.

Laboratory and imaging studies. Laboratory studies were as follows: hemoglobin (Hb) 14.4 g/dl, hematocrit (Hct) 43%, platelets (Plaq) 214,000, leukocytes (Leu) 6,600 U/L, neutrophils (Neu) 45%, lymphocytes (Lymph) 26%, monocytes (Mon) 24%, bands (Ban) 3%, prothrombin time (PT) 14.4 sec, partial thromboplastin time (PTT) 30.3 sec.

Echocardiogram. Sinus rhythm, FC 100/min, P axis +70, left bundle branch block of the bundle of His.

Chest X-ray. *Situs solitus*, levocardia, cardiothoracic index 0.78 at the expense of the right atrium and ventricle. Management continued. Aspirin was suspended. The patient remained in stable condition, hospitalized, and awaiting surgery.

February 9, 2010 (admission). The patient was admitted to surgery. A tricuspid valve replacement was done with an Edwards biological valve and closure of the residual IVC. At the start of surgery she presented hypotension 42/32 mmHg (36 mmHg), CF 25/min, saturation O_2 0% and cardiorespiratory arrest of 3 min duration. She was managed with 300 mg of adrenaline, bicarbonate 10 mEq and a gram of calcium. Blood loss was 1400 ml and urinary output (UO) was 3.5 ml/kg/h. The patient was removed from the pump on the first attempt with defibrillation at 1 J/kg, dopamine 8 mg/kg/min, dobutamine 8 mg/kg/min, milrinone 0.5 mg/kg/min and adrenaline 0.33 mg/kg/min. Aortic clamping time was 1 h 10 min. Time of circulatory arrest was 65 min. She was admitted to the surgical intensive therapy with AT 55 mmHg, CF 132/min, PVC 12 mmHg, and temperature 36.4 ºC. She presented with active bleeding via the orotracheal cannula, transfused red cell concentrate at 15 ml/kg/dose, fresh frozen plasma, cryoprecipitate and platelets.

Blood gases. pH 7.37, PaO_2 105, $PaCO_2$ 29.9, HCO_3 17.1, BE –6.7, $SatO_2$ 98.3%, anion gap 20.3, Lact 8.7, Na 140 mEq/l, K 2.9 mEq/l, Cl 105 mEq/l, Ca ionic 0.97 mEq/l.

Postoperative echocardiogram. Postoperative echocardiogram demonstrated severe right ventricular insufficiency. Liver was 4 cm from the right costal margin and hypotense. Vasopressin was initiated, orotracheal intubation, FiO_2 100%, PIM 20, PEEP 4, right hypoventilation, and the surgical wound was without signs of bleeding. Chest and mediastinal tube demonstrated bloody drainage.

Chest X-ray. Chest X-ray demonstrated total opacity of the right hemithorax with mild congestion. Abdomen was unremarkable, liver margin at 4-4-5 cm. The patient was sedated with midazolam and fentanyl, anisocoria with right mydriasis of 1.5 mm greater than the left. The patient presented with ventricular fibrillation managed with electrical activity without pulse, cardiopulmonary resuscitation for 27 min, six doses of adrenaline, and bicarbonate replacement on two occasions, without response to advanced resuscitation maneuvers.

Laboratory and imaging studies. Hb 10.5 g/dl, Hct 31%, Plaq 94,000, Leu 6,600 U/L, Neu 69.7%, Lymph 23.6%, Monocytes 6%, Ban 0%, PT 19.1 sec, PTT 39.7 sec.

Blood gases. pH 7.06, PaO_2 67.7, $PaCO_2$ 35.3, HCO_3 9.6, BE –19.2, $SatO_2$ 87.5%, anion gap 28.4, Lact 15, Na 139 mEq/l, K 5.2 mEq/l, Cl 106 mEq/l, Ca ionic 0.86 mEq/l.

CASE PRESENTATION

Hemodynamic Service (Dr. Jorge Luis Villatoro Hernández)

Images showing severe tricuspid insufficiency (Figure 1) and IVC (Figure 2) are presented.

DISCUSSION

Hemodynamic Service (Dr. Luís Alexis Arévalo Salas)

Today's case is open and the diagnoses are clearly described in the clinical history. In regard to this history, I will discuss the possible pathophysiological conditions that led to the patient's death.

There is no hereditary family history of note, but it is relevant that this patient was evaluated in another institution where the diagnosis of pulmonary vascular disease associated with IVC and PDA at 2 years of age was made which, although not improbable, is difficult that it would happen. There are many reviews in the medical literature dating back to the 1970s in which young children with IVC were studied and very interesting conclusions were made. For example, Drs. Kirklin and Barrat-Boyes reported in their classic book on Cardiac Surgery[1] that infants <1 year of age could have elevated pulmonary vascular resistance (PVR), even to >7 Wood units (Wu) without the histopathological changes of the pulmonary vasculature being compatible with pulmonary vascular disease, according to the classic description by Heath and Edwards.2 It should be mentioned that this description dates back to 1958 and remains at present because nothing would have

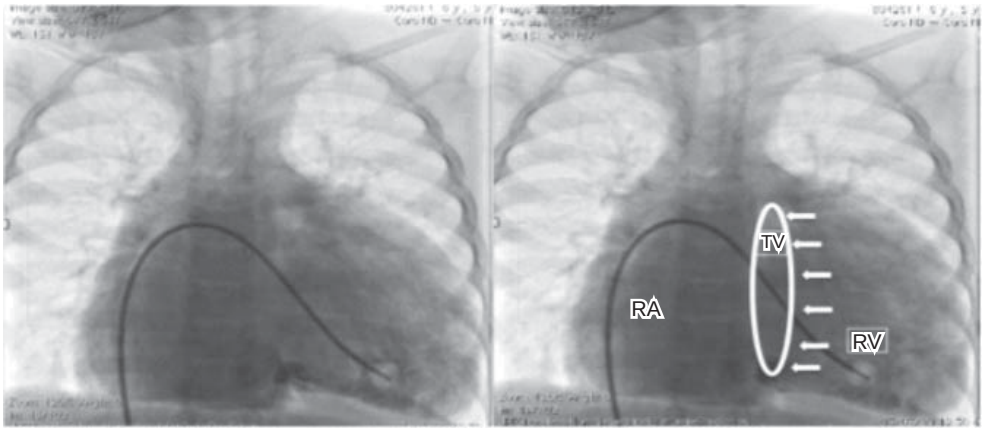

Figure 1.

Frontal projection where the catheter is located in the right ventricle. During administration of contrast media, the following are observed: significant dilatation of the tricuspid ring and right atrium and severe tricuspid insufficiency.

RA, right atrium;
TV, tricuspid valve;
RV, right ventricle

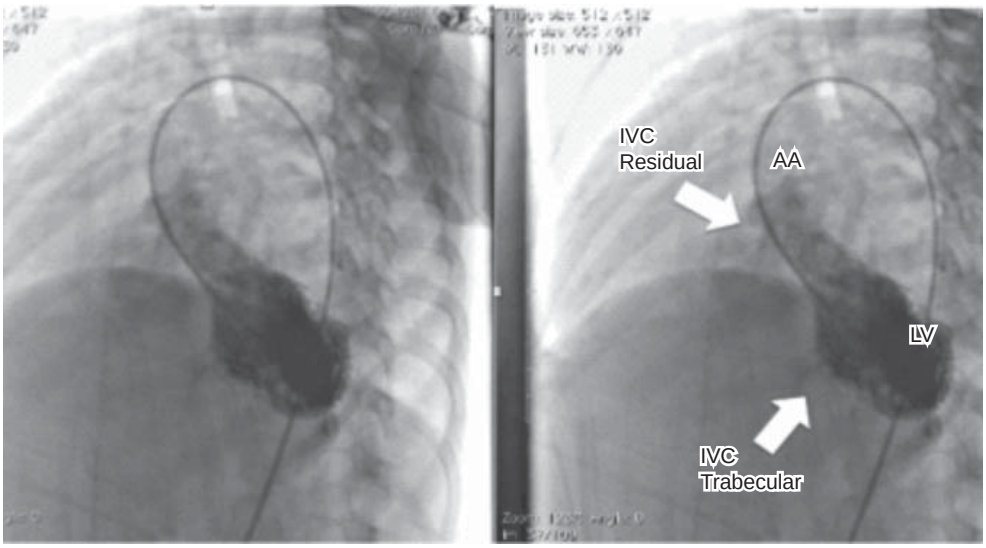

Figure 2.

Elongated axial projection allows visualization of the entire length of the interventricular septum. Two-mm short circuit is found in the patch corresponding to a residual IVC flow from left to right and another small IVC of 2 mm in the trabecular portion with left to right short circuit. Arrows show small and inconsequential residual IVC.

IVC, interventricular communication;
LV, left ventricle;
AA, ascending aorta

improved the histopathological classification of the pulmonary lesions to indicate pulmonary vascular disease. At present, it is necessary to mention an important condition. Frequently we speak of pulmonary artery hypertension (PAH), but it should be understood that it is the manifestation of the degree of PVR that is conditioned by the anatomic characteristics and the reactivity of the pulmonary vessels. Therefore, when there is peripheral vasoconstriction, an increase in the proximal pulmonary pressure is established, thereby causing PAH. If these pulmonary vessels preserve the ability to dilate, it should be considered as reactive with the principal stimuli being oxygen and certain drugs. At this time it is important to mention that pulmonary vascular disease is the irreversible damage of the pulmonary vasculature. It is marked by different histopathological changes that go from an increase in the thickness of the mid-muscular layer, accumulation of cells similar to fibroblasts that invade the elastic lamina, up to the development of fibrosis, plexiform lesions and necrosis with loss of vasodilation capacity. At this time, a classical condition called Eisenmenger syndrome is established in which the initial left to right shunts are inverted and cyanosis appears.

The patient had two left to right shunts, one of the ductus arteriosus and the other of the IVC, which caused a pulmonary overflow—the main stimuli for the development of the initial damage of the pulmonary vasculature and, thereby, of the PAH. However, when this patient was admitted, there was no cyanosis and echocardiograms reported left to right shunts for both lesions. There were no clinical or echocardiographic criteria to sustain inoperability. On the face of severe PAH, it is very difficult to clinically and hemodynamically establish if there is a continuing disease process in progress even when the defect has been repaired.

Upon review of this patient's clinical data, it is noted that there is prevalence of a holosystolic murmur with a 2/6 intensity and hepatomegaly. The presence of an intense murmur indicates a massive left to right shunt because, when the pulmonary pressure reaches the level of the systemic pressure in response to an increase in PVR, ventricular pressures equalize and the murmur tends to disappear but causes a very intense second sound not reported in the clinical history. The presence of an apical reverberation is of great clinical relevance for the cardiologist. The translation of this sign is that, when there is an increase in the pulmonary overflow due to large shunts, an increase in the pulmonary venous return is established and the blood volume in the left atrium increases. At this time the mitral valve becomes relatively stenotic and a diastolic murmur is established. When the vascular resistance is elevated and thereby the pulmonary arterial pressure is high, the capacity for vasodilation is lost and the pulmonary flow decreases. Consequently, the pulmonary venous return is less. The presence of an apical reverberation then suggests that the pulmonary vessels still preserve their capacity for vasodilation. Until now, we can assure that this patient did not have pulmonary vascular disease because she had no cyanosis and echocardiograms showed left to right shunts.

At 4 years of age and with the condition already mentioned, it was decided to subject the patient to surgery for closure of the defects. It should be mentioned that, from the first echocardiogram, certain alterations in the tricuspid valve are described, specifically a prolapse. It is very important to mention this at this time because tricuspid anatomic lesions are unusual in the pediatric age. This is not so for functional alterations, i.e., there is a certain degree of tricuspid insufficiency in an anatomically competent valve, for example, in neonates with elevated PVR or in congenital heart diseases that increase the right ventricular pressure. In this case an anatomic disorder was seen that caused tricuspid insufficiency and consideration should have been given for its repair during the surgical intervention.

There was a postoperative event that can be considered to have been a hypertensive crisis. The child had PAH and remained desaturated. For this reason it was necessary to treat her with mechanical ventilation and pulmonary vasodilators in order to end the crisis. It should be mentioned that this is not an unusual event because it is the most frequent cause for this complication. Because IVC is one of the most frequently operated on congenital heart disease worldwide, it has also been described with an incomplete A-V channel, truncus arteriosus, total anomalous connection of the pulmonary veins or in the D-transposition of the great arteries.[3] In addition to the already known PAH, an aortic clamping is described of just over 1 h, which can be considered to be prolonged. When both conditions are combined, the secretion of vasoconstrictors is promoted such as thromboxane, leukotrienes and other inflammatory agents that maintain a state of hypertension. In this case it was resolved with conventional management based

on oxygen and vasodilators. In fact, the baseline echocardiogram in the ICU documented a pulmonary pressure of 42 mmHg. Based on this progress, it can be inferred that the elevated pulmonary pressure was reactive because the PVR was not corrected.

Postoperative evolution describes a clinical picture of hepatitis described with positive A antibodies, but that appears to be inconsequential in the patient's clinical history. What is totally abnormal is that the patient would continue with hepatomegaly and maintained even after correction of the defects. Whereas the association of hepatomegaly with cardiomegaly indicates cardiac insufficiency, in this case once the heart lesions were corrected, the hepatomegaly should have disappeared. In this situation it was decided to perform a cardiac catheterization, which reported a normal PVR, considered to be between 2 and 4 Wu— with elevated pulmonary pressure and two small IVCs that caused a pulmonary output of 1.2 (which may be considered to be of little significance because when it is >1.5 the child tends to be asymptomatic). One defect was located in the margins of the surgical patch and the other in the trabecular septum away from the surgical patch, but the pathology was really based on a severe tricuspid insufficiency.

It is timely at this time to mention the role of the right ventricle. It manages the same volume as the left; however, it has profound anatomic and functional differences. If we keep in mind the anatomy of the left ventricle, it has a conical, muscular appearance, which gives it a circumferential contractile ability and allows maintaining pressures capable of enduring a systemic resistance of between 12 and 16 Wu versus the 2-4 Wu that the right ventricle maintains under normal conditions. In turn, the latter consists of three parts called the inlet, trabecular and outlet, forming a helical structure. Therefore, its contractility is longitudinal, forming a low pressure chamber that allows passage of a similar volume to the left ventricle. When the cardiac insufficiency is severe (as in this case), the ventricle loses its contractile capacity in great measure. It has less capacity to manage the volume towards the pulmonary artery and a blood bed is established between the pulmonary artery and the right atrium, causing the hepatomegaly experienced by this patient. At present, the tricuspid insufficiency should be considered as a factor of death associated with the correction of cardiac defects, especially when a concomitant correction is not carried out.

Finally, due to the condition of this patient with severe tricuspid insufficiency, it was mandatory that she undergo tricuspid valve replacement. However, postsurgical outcome was predictable based on the long-term presence of right ventricular dysfunction. My final diagnoses are as follows:

- Severe tricuspid insufficiency causing right ventricular dysfunction
- Moderate residual arterial hypertension with low PVR without evidence of pulmonary vascular disease
- Two small IVCs: one residual and the other congenital, without clinical consequences

Pathology Department (Dr. Mario Pérezpeña-Díazconti)

Postmorten anthropomorphic data revealed marked chronic malnutrition with weight of 14.9 kg versus an expected weight of 22 kg. Height was 104 cm versus a reference height for her age of 117 cm. In the anterior chest over the mediastinum there was a sutured wound of 12.5 cm in length. On opening the chest cavity, the right pleural cavity contained 10 mL of serosanguineous fluid, and the left cavity contained 20 cc of fluid with the same characteristics. A 4 x 3 cm hematoma was removed from the mediastinum. The pericardium (Figure 3) demonstrated multiple adhesions and blood clots. In that study the main organ is the heart, which was increased in size and weight. The cardiopulmonary block weighed 450 g, with a reference weight of 343 g. The atria were dilated. In the posterior view of the heart, a markedly dilated inferior vena cava was seen (Figure 4). Moreover, the surface of the lungs was light gray except for the bases (which were a reddish color). At the base of the right lung we observed two blisters that did not represent much change. Trachea and aorta showed no change. The heart cut revealed the prosthetic right atrioventricular valve, 2.5 cm in diameter (Figure 5). Both ventricles were hypertrophied; the right also was dilated, and there was an infundibular IVC of 0.1 cm. The ductus arteriosus was occluded surgically. Histologically, the ventricles showed hypertrophy with mild disorganization of the cardiomyofibrils, the nuclei were hyperchromatic with variations in form (Figure 6). As a result of heart disease, there were changes observed in the lungs with alveolar spaces with hemosiderin-laden macrophages, which

represented repeated bouts of edema and bleeding and minor changes apparent in the superior lobes associated with left heart insufficiency. Vascular damage was significant where the walls of the vessels were thickened due to pulmonary hypertension. Changes were also seen that were most apparent in the small and medium-sized vessels with wall muscularization (Figure 7), findings that were not uniform in all vessels. The damage was more pronounced in other fields, with fibroblast proliferation, muscularization and intimal proliferation arranged concentrically (Figure 8). In all vessels studied, we did not find plexiform lesions or wall necrosis. Staining of the elastic fibers showed damage to the vessel wall (Figure 9). These changes are included in grade III Heath and Edwards classification (Table 1), with changes in different stages in different vessels.

A relatively frequent finding is the presence of plastic material in the lumen of the vessels, which could correspond to catheter fragments, accompanied by a giant multinucleated cell that may be attributed to a foreign body. The liver demonstrated a nutmeg appearance (Figure 10). It was increased in weight and size (800 g versus a refer-

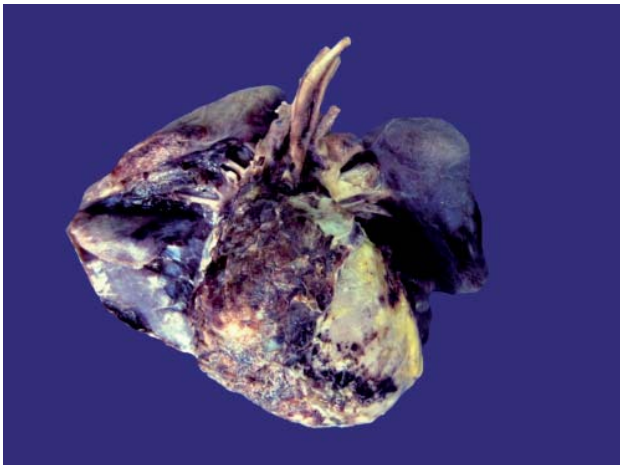

Figure 3. Pericardial surface showing fibrous aspect with attached clots. The heart is increased in size and weight.

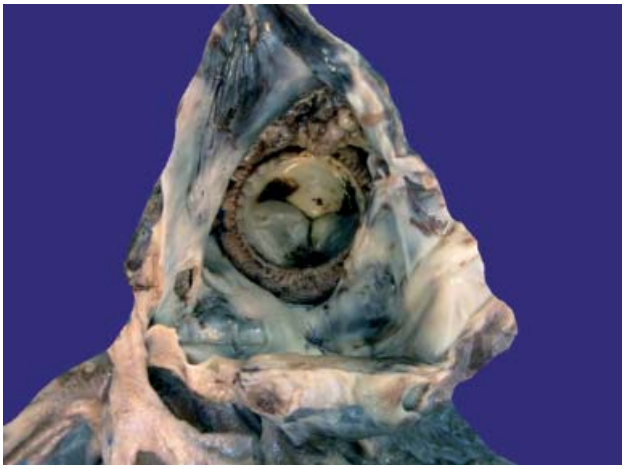

Figure 5. Prosthetic valve without alterations. Sutures were found to be integral.

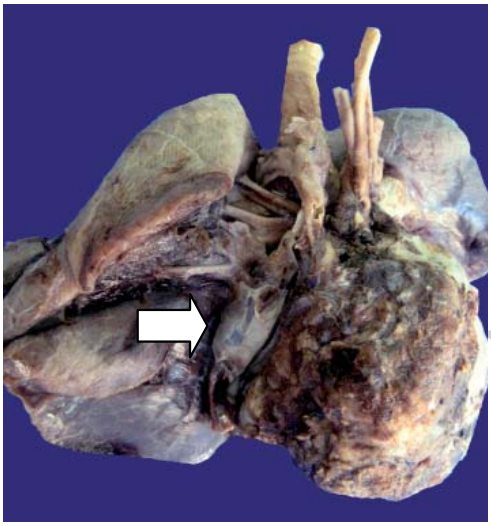

Figure 4. Dilated vena cava (arrow).

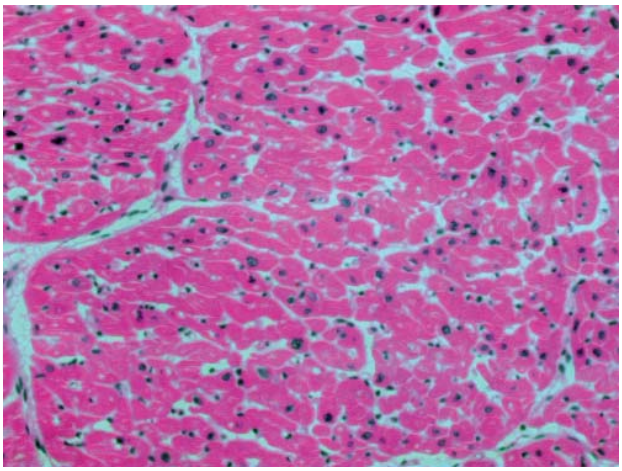

Figure 6. Histologically, both ventricles show hypertrophic changes with disorganization of the myofibrils, pleomorphism and nuclear hyperchromatism.

ence weight of 680 g) due to congestion associated with right heart insufficiency. Histologically, the porta spaces did not present any alteration, and there were no data of hepatitis A, which had been diagnosed 2 years prior according to the clinical history. The spleen was congestive and increased in size with data of right heart insufficiency. Similarly, the kidneys were increased in size, with intense congestion in the cortex and medulla. The bladder wall demonstrated contraction bands. The cerebral vessels were congestive, although structurally no alterations were found. Histologically some neurons had data of hypoxia with intense dense red cytoplasm and hyperchromatic and condensed nuclei. The final anatomic diagnoses are enumerated in Table 2.

Patients with severe pulmonary hypertension frequently decompensate in the immediate postoperative period. The cause for this decompensation is found in the right ventricle, which is incapable of overcoming the increased peripheral resistance.

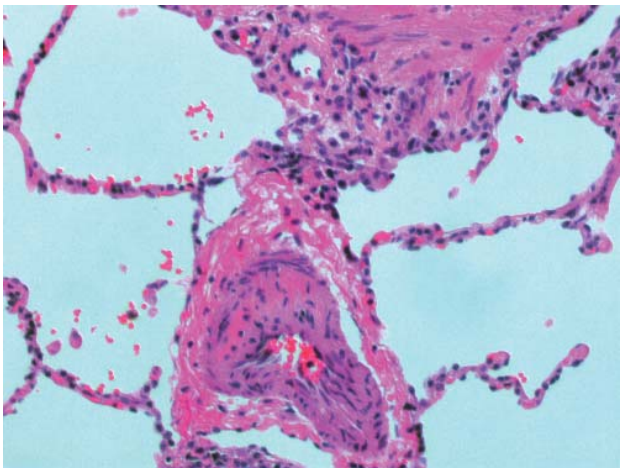

Figure 7. Vascular changes are apparent and affect small and medium-sized vessels. There is muscularization of the wall with the presence of collagen.

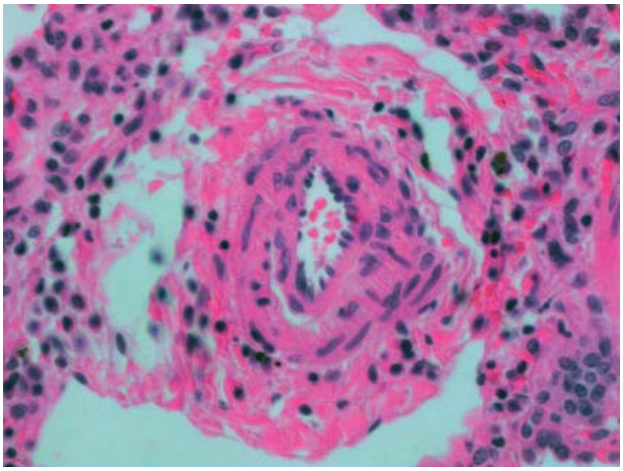

Figure 8. Damage seen in different states in the pulmonary vasculature with the presence of collagen and muscularization.

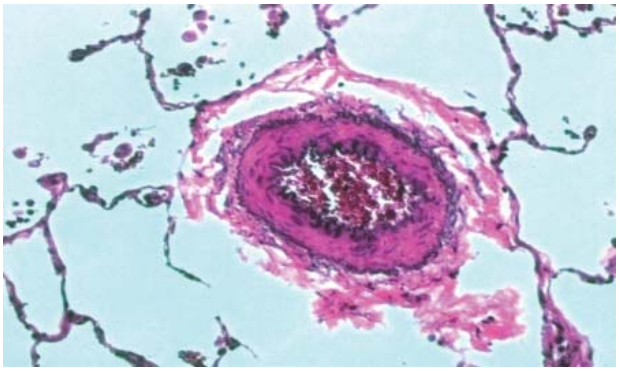

Figure 9. Staining of elastic fibers shows alteration in the vascular wall in the pulmonary vessels of the patient, where the elastic is lost and there are only a few traces of fibers due to the presence of collagen and fibroblast proliferation.

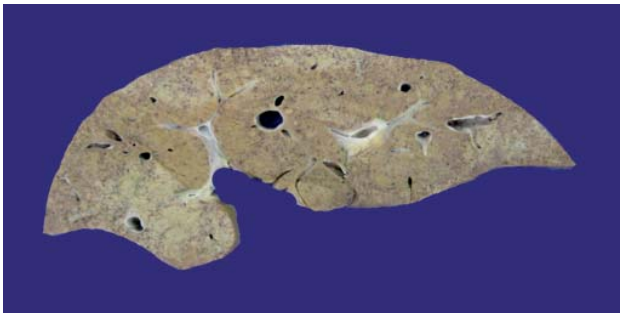

Figure 10. Liver with nutmeg appearance due to right cardiac insufficiency.

Table 1. Classification of Heath and Edwards of plexogenic arteriopathy

Grade 1. Muscularization of the pulmonary arteries
Grade 2. Migration of obscure muscle cells to the arterial intima
Grade 3. Transformation of intimal cells in myofibroblasts to form muscle, collagen and elastin
Grade 4. Plexiform lesions and dilatations
Grade 5. Rupture of dilated vessels with hemorrhage and exudative lesions
Grade 6. Necrotizing arteritis

Table 2. Final anatomic diagnoses

Principle disease
Congenital cyanogenic cardiopathy characterized by dysplasia of the posterior valve of the tricuspid valve with prolapse Trabecular IVC
Concomitant alterations
• Status post-closure of infundibular IVC, closure of PDA and tricuspid valvuloplasty with plication with fixation of the tendinous cords of the anterior leaflet • Clinical history of residual IVC and moderate to significant tricuspid insufficiency • Status post-replacement of the right prosthetic atrioventricular valve • Prolapse of the tricuspid posterior valve • Apical IVCs of 2 and 6 mm • Status post-replacement of right atrioventricular valve • Closure of residual IVC • Cardiomegaly • Biventricular hypertrophy and dilatation of the right ventricle • Pericardial-epicardial adhesions • Hematoma in mediastinum (4 x 3 cm) • Old and recent pulmonary hemorrhages • Grade 3 pulmonary vascular disease of Heath and Edwards with concentric laminar proliferation of the muscular and intima caps and transformation of myofibroblasts in the intima
Data of cardiac insufficiency
Bilateral pleural effusion (10 cc right, 20 cc left, serous) Congestive hepatomegaly (OW 800 g/EW 680 g) Severe malnutrition (OW 14.9 kg/EW 22 kg)
Cause of death
Cardiogenic shock

IVC, interventricular communication; PDA, persistence of patent ductus arteriosus; cc, cubic centimeters; OW, observed weight; EW, expected weight.

FINAL COMMENTS

Intensive Care Service (Dra. Ma. de Lourdes Marroquín Yáñez)

Patients who, for correction of heart disease, require the use of extracorporeal circulation may present as a complication low cardiac output syndrome (LCOS) consisting mainly of a cardiac dysfunction. Its peak presentation is from 16 to 18 h postoperative. Its incidence in U.S. hospitals is ~27%.[5] In our institution, the reported incidence is ~35% with a mortality of 6%. Our patient presented, as the principal factor for developing this syndrome, chronic right ventricular failure due to tricuspid insufficiency which, added to a profuse postoperative hemorrhage requiring large quantities of fluid for volume replacement and some elevated pulmonary resistance caused a biventricular failure resulting in the patient's death. Treatment of LCOS requires the use of diverse vasoactive medications. The general worldwide consensus agrees on the use of milrinone, a vasodilator with effects both on the pulmonary as well as systemic vasculature and improves diastolic function.[6] In a multicentric study in the U.S., the prophylactic use of milrinone was reported, which reduced the risk of LCOS by 64%. If the ventricular failure is irreversible, the final option is the use of extracorporeal membrane oxygenation (ECMO).

Department of Hemodynamics (Dra. Begoña Segura Stanford)

There are three groups of patients with acynogenic heart disease of increased pulmonary flow: 1) those who clinically and radiographically have a left to right shunt, normal saturation and data of congestion with cardiomegaly and are perfectly operable: 2) those who already have pulmonary hypertension established by irreversible pulmonary vascular disease with desaturation, cyanosis, without cardiomegaly and decreased pulmonary flow who are not candidates for surgical correction, and 3) those patients who are halfway on the road towards pulmonary vascular disease with increased pulmonary pressure, but still do not have cardiomegaly, increased pulmonary flow and congestion. For these patients a vascular reactivity test is done in the hemodynamic laboratory that consists of the controlled administration of medications or vasodilating gases. Initially and up to now, oxygen at 100% is administered to cause a fall in the midpressure of the pulmonary artery with respect to systemic pressure of 10 mmHg or more. If there is not a good response to the oxygen, in our institution a double drug challenge is being carried out with levosimendan, which is a vasodilator with a pulmonary vasodilating effect and iloprost that is a selective pulmonary vasodilator with the intention of reducing the PVR <20% or if there is a fall in the mid-pressure of the pulmonary artery (<10 mmHg with respect to baseline). If these reductions present themselves, it is considered a positive test that indicates the operability of the patient. In other locations, tests with nitric oxide are done.

Cardiology Service
(Dr. Julio Erdmenger Orellana)

Based on the echocardiographic studies and hemodynamics, our patient was clearly operable with regard to the concept of pulmonary vascular disease. Even though the interventricular defects may be closed, once the mechanisms of pulmonary vascular damage have been established, it is not reversible. In the worst case scenario, disease progression of the pulmonary vascular disease continues. On autopsy, severe pulmonary damage is seen when compared with the hemodynamic evaluation. Therefore, I believe that even though the IVC was closed, the pulmonary vascular disease continued its progression. Tricuspid insufficiency generally is well tolerated (with the exception of severe postoperative insufficiency where the volume in the right ventricle is increased). It seems to me that the main damage experienced by this patient was right ventricular dysfunction aggravated by pulmonary vascular disease. From this case we have learned that it is well documented to carry out in our institution operative transesophageal echocardiogram for all patients who are subjected to valve surgery. If severe or moderate valvular insufficiency persists, re-intervention is done. Another important point is to reduce the time between the first and second surgery to avoid development of ventricular dysfunction. It is necessary that the institution have ECMO. Even though global mortality in heart surgery is ~10%, ECMO could help rescue an important number of patients who would otherwise die.

Correspondence: Dr. Sergio Ruíz González
E-mail: lulymarroquin@yahoo.com.mx

REFERENCES

1. Kouchoukos N, Blackstone E, Doty D, Hanley F, Karp R. Ventricular septal defect. In: Kouchoukos N, Blackstone E, Doty D, Hanley F, Harp R, eds. Kirklin/Barrat-Boyes Cardiac Surgery. Philadelphia: Churchill Livingstone;2003. pp. 850-909.
2. Heath D, Edwards JE. The pathology of hypertensive pulmonary vascular disease: a description of six grades of structural changes in the pulmonary arteries with special reference to congenital cardiac septal defects. Circulation 1958;18(4 Part 1):533-547.
3. Nichols D, Ungerleider R, Spevak P, Greeley W, Cameron D, Lappe D, Wetzel R. Critical Heart Disease in Infants and Children. Philadelphia: Mosby Elsevier; 2006.
4. Mascherbauer J, Maurer G. The forgotten valve: lessons to be learned in tricuspid regurgitation. Eur Heart J 2010;31:2841-2843.
5. Wessel DL. Managing low cardiac output syndrome after congenital heart surgery. Crit Care Med 2001;29(suppl 10):S220-S230.
6. Hoffman TM, Wernovsky G, Atz AM, Kulik TJ, Nelson DP, Chang AC, et al. Efficacy and safety of milrinone in preventing low cardiac output syndrome in infants and children after corrective surgery for congenital heart disease. Circulation 2003;107:996-1002.