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Total anomalous connection of pulmonary veins to the portal vein. Value of multislice angiotomography. Report on three cases

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ABSTRACT

Objective: Present tomographic data from the patients with emphasis on structural characteristics.

Material and method: Retrospective study of three patients in whom the four pulmonary veins connected to the portal vein. This malformation is one of the varieties of total anomalous connection of pulmonary veins. In any of them, the pulmonary veins do not connect to the left atrium and blood from the lung reaches the right atrium directly or through its tributary systems. In such cases, arterialized pulmonary blood and systemic venous blood mix at the site of the anomalous connection and a short circuit is formed from the right atrium to the left atrium through an interauricular communication, allowing patients to survive.

Total anomalous connection of pulmonary veins accounts for between 0.4 and 2.0% of congenital heart diseases: it occurs in 6.8 of every 100,000 individuals. It is diagnosed in 68% of patients in neonatal stage, which reflects the severity of the condition. The infracardiac variety of total anomalous connection of pulmonary veins accounts for between 15 and 26% if all its varieties.

Multislice angiotomography allows us to view the blood vessels and adjacent organs under consideration and obtain high-definition anatomic information. In the patients in this study, total anomalous connection of pulmonary veins to the portal vein was viewed with three-dimensional volumetric tomographic reconstructions and their correlation with ultrasonography studies.

Key words: Total anomalous connection of pulmonary veins, congenital heart disease, angiotomography, ultrasonography.

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The first description of a case of total anomalous connection of pulmonary veins was made by Wilson in 1798. The first systematizations based on embryology were made by Nelly, Edwards and Helmoltz in 1956, Darling in 1957, and Van Praag in 1969.[1] Darling divided them in supracardiac, cardiac, infracardiac, and mixed. (Table 1)

Table 1. Total anomalous connection of pulmonary veins. Varieties and frequency by site

Supracardiac	50%
To innominate vein	40%
To right superior vena cava	10%
Cardiac	30%
To coronary sinus	20%
To right atrium	10%
Infracardiac	15%
To portal vein	8%
To venous conduit	4%
To inferior vena cava	2%
To hepatic veins	1%
Mixed	5%

Taken from: Marin Huerta E, Castaneda AR. Drenaje venoso pulmonar anómalo total. Cardiología Pediátrica Clínica y Cirugía. Mexico: Salvat, 1986;260.

The infracardiac varieties are usually the most serious. They all require early surgical treatment. Clinical diagnosis is difficult; establishing it requires highly specialized studies which provide precise data, as performed in our patients: multislice angiotomography and Doppler color ultrasound.

The severity of the malformation in our patients did not allow us to recur to surgical treatment; all three died despite medical treatment.

The purpose of this document is to present the tomographic data from our patients with emphasis on structural characteristics.

PATIENTS AND METHOD

A retrospective study of patients attended at the National Institute of Pediatrics Cardiology service between the years 2003 and 2011. We found three cases of girls with total anomalous connection of infracardiac pulmonary veins, with drainage to the portal vein, studied with angiotomography and ultrasound.

The tomographic studies were conducted using Siemens Sensation multislice tomography machines with 4 and 64 rows of detectors. Non-ionic water-soluble contrast material was administered in doses of 2 mL/kg through a peripheral vein in a lower limb, with bolus-tracking in craniocaudal (arterial phase) and caudo-cranial (venous) direction; slice thickness 1-1.25 mm, PICH 1.0, MSA: 120, KVP: 80. These parameters varied in relation to the patients' age and weight. The images were evaluated with maximum projection intensity and three-dimensional volumetric reconstructions. The ultrasound studies were performed with General Electric LOGIQ P9 and HDL 5000 machines.

CLINICAL CASES

Case 1. Girl age 2 months, without perinatal antecedents of interest. Five days before she presented dyspnea and cyanosis, which had initially been treated at another hospital as pneumonia. The echocardiogram showed dextro-isomerism, aorto-cava juxtaposition, two interauricular communications, a 6 mm ostium primum defect and a 7 mm ostium secundum defect. There was a venous collector with infradiaphragmatic drainage; the drainage site was not specific. Cardiac catheterization revealed an atrioventricular canal and pulmonary atresia. The abdominal ultrasonography showed, in a longitudinal subcostal slice, a 1 cm diameter collector descending in front of the abdominal aorta and emptying into the portal vein (Figure 1).

The angiotomography revealed that the liver was in horizontal position and the stomach was positioned to the right of the center line.

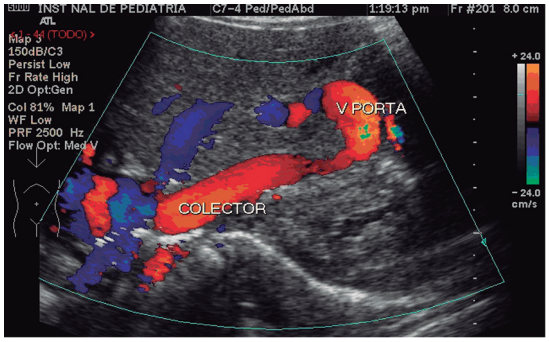

Figure 1. Sagittal ultrasound of the right hepatic lobe showing permeable collector of 1 cm in diameter which, before emptying into the portal vein, presents an area of stenosis (arrow), a probable site of intrinsic obstruction.

The pulmonary veins converged in a collector descending to the rights of the center line, crossed the diaphragm, and entered the liver to join the portal vein (Figure 2). Both lungs were trilobular and the patient lacked splenic function (anesplenia).

Case 2. Girl age 6 months, born by vaginal delivery; weight 3,400 g, APGAR 7-8. On arrival at the National Institute of Pediatrics she was found to have poor respiratory effort and cyanosis, with arterial oxygen saturation between 68 and 74%. Heart rate 140 beats per minute and respiratory rate 45 per minute. The echocardiogram showed aorto-cava juxtaposition, ostium primum interauricular communication, auriculoventricular canal, with right ventricle dominant; AV insufficiency, pulmonary atresia; confluent pulmonary artery branches, anomalous pulmonary vein connection, possibly mixed. A sagittal ultrasonography slice at the level of the epigastrium and taking the portal vein as reference, showed that it received an infradiaphragmatic collector 1 cm in diameter, with aneurismatic dilation in curled shape before connecting to the portal vein (Figure 3). There was also pulmonary dextroisomerism and anesplenia.

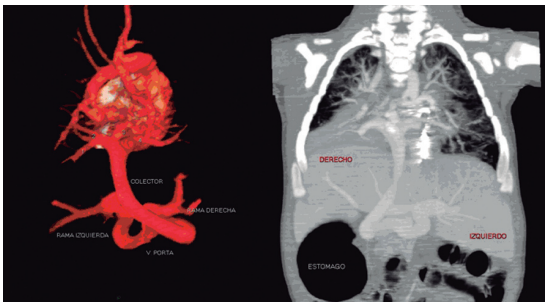

Figure 2. 3D postero-anterior reconstruction and maximum projection intensity reconstruction of coronal AP, identifying the four pulmonary veins converging in a tortuous vertical collector at the hepatic level which empties into the portal vein. Dextro-isomerism, liver horizontal, stomach to the right.

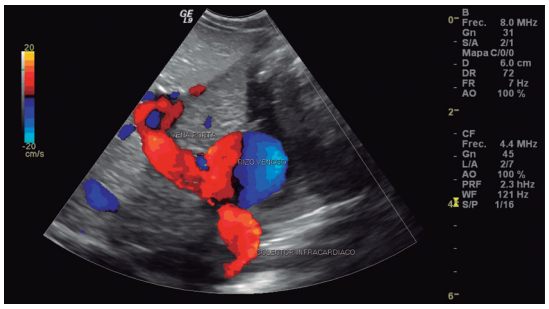

Figure 3. Abdominal ultrasound with presence of aneurismatic dilation of the collector before connecting to the portal vein.

The three-dimensional volumetric reconstructions of the angiotomography in postero-anterior projection showed the confluence of the four pulmonary veins to a long infracardiac collector, leading discretely to the left of the center line, of tortuous appearance and an aneurismatic dilation before connecting to the portal vein (Figure 4).

Case 3. Girl age 58 days, born from the first pregnancy. Six prenatal ultrasounds were taken; 4 were reported as normal, and the last two with oligohydramnios. Patient was born by vaginal

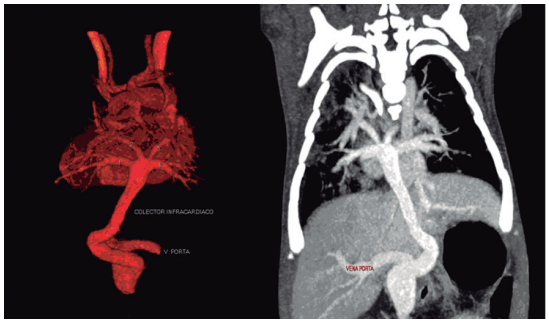

Figure 4. Three-dimensional volumetric postero-anterior reconstruction and maximum intensity projection A-P coronal reconstruction showing the four pulmonary veins converging in an infradiaphragmatic collector with the portal vein.

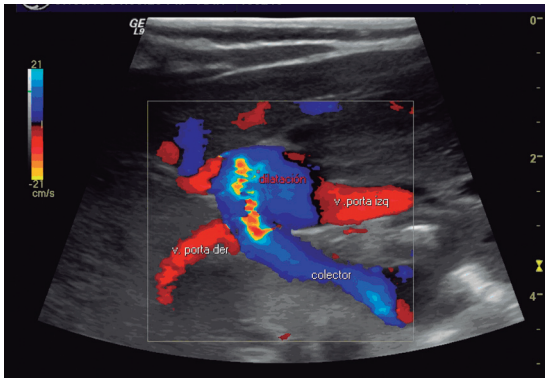

Figure 5. Abdominal Doppler color ultrasound showed the presence of an infracardiac collector draining to the portal vein.

delivery at 38 weeks of gestation with weight 2,600 g. APGAR and Silverman data unknown. Apparently the patient cried and breathed at birth without the need for resuscitation maneuvers. She left the hospital at 24 hours, and for one week had central cyanosis and dyspnea after eating. She was evaluated by doctors of the National Institute of Pediatrics Cardiology Service. The echocardiogram showed persistence of arterial conduit, situs solitus, AV canal, probable anomalous drainage of pulmonary veins and accentuated pulmonary valve stenosis. The Doppler color ultrasound (Figure 5) showed a collector connecting to the portal vein, a finding corroborated in coronal and three-dimensional volumetric reconstructions of the multislice tomography; she also had pulmonary dextroisomerism and absence of splenic function. (Figure 6).

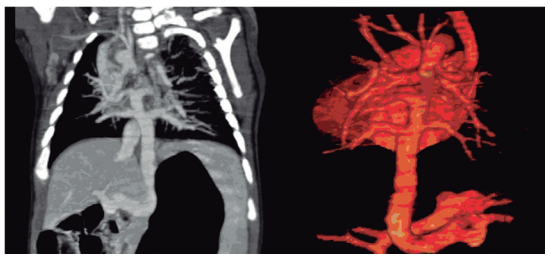

Figure 6. Maximum intensity projection coronal reconstruction showing convergence of the four pulmonary veins to a descending vein (collector) which passed through the diaphragm and ends in the portal vein. The three-dimensional postero-anterior volumetric reconstruction shows in detail the tortuous dilation of the collector (arrow) prior to its connection to the portal vein.

Analysis

In total anomalous connection of pulmonary veins, the right atrium receives blood from both vena cava and oxygenated blood from the four pulmonary veins. The clinical manifestations of this heart condition depend on: *1)* Presence or absence of an obstruction of the collector and *2)* size of the interauricular communication, whether an oval, wide or narrow, ostium primum, ostium secundum or persistent AV canal.

Symptoms may appear hours after birth or days or weeks later, depending on the site and magnitude of the obstruction in the collector; data related to pulmonary hyper-flow, such as: dyspnea, tachypnea, respiratory difficulty and fatigue during eating. There may be a degree of cyanosis.

Most patients have symptoms in the first week of life. While the arterial conduit remains open, cyanosis will predominate over pulmonary congestion because pulmonary pressure will not be higher than systemic pressure, which will delay the appearance of symptoms for several weeks.[5]

Patients may worsen rapidly, with increase in cyanosis and appearance of metabolic acidosis; even when they are not extreme, the clinical situation may remain stable for days, but worsens abruptly when the arterial conduit closes. In that case cyanosis may diminish, but respiratory insufficiency increases; tachypnea and intercostal strain, hepatomegaly and weak pulses appear. Patients appear mildly cyanotic and malnourished. There is enlargement of the precordium, with a marked impulse from the right ventricle. In the second left intercostal space, on the sternal edge, a grade 2/4 systolic ejection murmur can be heard; the second pulmonary sound is accentuated and constantly reduplicated. The chest x-ray shows congestion of the pulmonary vasculature, above all at the level of the strands; the pulmonary fields appear veiled" due to the edema. The cardiac sillhouette is enlarged.[1-6]

When the collector is long and has a blockage, as occurs in drainages to the portal vein, the pulmonary pressure rises and oxygen saturation drops considerably because in that case the pulmonary flow diminishes and the venous capillary pressure rises leading to pulmonary edema. If the collector is not obstructed, arterial saturation may be between 90 and 92% given that the right atrium receives only 3 of 5 parts of saturated blood from the pulmonary veins and passes to the right ventricle and to the pulmonary artery; this overloads the right cardiac cavities and causes pulmonary hyper-flow and causes right heart failure.[1,6,7]

Eighty to ninety percent of patients die within the first year of life if they are not surgically treated, or die in the first months of life if there is a blockage of the collector.[3-6]

In total anomalous connection of the infracardiac pulmonary veins the most common site of anomalous emptying is the portal vein. Cases reported with emptying in the venous duct, in the hepatic veins, in the vena cava inferior, in the splenic vein, or in the upper mesenteric vein are very rare. In this kind of anomalous connection the blockage of the collector may be anatomic or functional. The anatomic type may be extrinsic (esophageal hiatus) or intrinsic, due to narrowness of the collector. Functional blockage is due to the resistance of the intrahepatic blood flow, which introduces itself between the site of the connection to the portal vein and the entry of blood to the heart.[2-6-7]

The diagnosis of this heart condition is made, initially, by echocardiography (a highly sensitive study for morphological and heart function evaluation). Its primary limitations are: *1)* The experience and skill of the echocardiographist. *2)* The coexistence of not always adequate acoustic windows. *3)* The difficulty of satisfactorily evaluating the extracardiac structures, the drainage site of the drainage from the infracardiac collector and the associated bronchopulmonary malformations.

Cardiac catheterization helps to evaluate the intra and extra cardiac vascular structures, and their functional aspects; however, it is an invasive and prolonged study. We need to bear in mind that these patients are almost always hemodynamically unstable at the time of the study.

Today, cardiac and extracardiac vascular structures can be viewed with multislice helical tomographies, in a short time, in arterial and venous phases. Table 2 shows the contribution and limitations of the different diagnostic methods.

Table 2. Advantages and disadvantages of diagnostic methods commonly used in evaluating pulmonary anomalies

Criterion	Echocardiogram	Magnetic resonance	CT Angiogram	Cardiac catheterization
Availability	Excellent	Variable	Excellent	Variable
Duration of study (not including anesthesia)	Long	Long	Short	Long
Study limited by acoustic window	Yes	No	No	No
Exposure to radiation	No	No	Yes	Yes
3D reconstruction capability	No	Yes	Yes	No
Provision of physiological information	Limited	Excellent (analysis of flow and potential volume)	Limited	Good (analysis of flow and identification of shunt)

Taken from: Himesh V Vays, S Bruce Greenberg. MR imaging and CT Evaluation of Congenital Pulmonary Vein Abnormalities in neonates and infants. Radiographics 2012;32:87-98.

Importance of the venous duct

During fetal life, 50% of the umbilical blood flow passes into hepatic capillary circulation through the venous duct. After birth, the umbilical blood flow stops and the pulmonary blood flow increases. While the venous duct remains permeable, the pulmonary venous flow passing through the collector vessel to the portal vein, can directly enter the vena cava inferior, and therefore causes neither pulmonary venous hypertension nor pulmonary edema.

Venous duct occlusion causes the venous blood to pass through the hepatic microcirculation, which raises pressure in the portal vein and pulmonary venous drainage, which is a cause of pulmonary edema.

When the venous duct closes, the portal venous system dilates and the blood flow is delayed in the descending vein, due to resistance to the flow through the long and narrow venous channels. If the venous duct is not permeable, the connection to the portal vein is blocked, as circulation through the hepatic parenchyma faces high resistance. If the descending vein connects with the vena cava inferior or the hepatic vein and there are no clinical or echocardiographic signs of obstruction, the patient can undergo surgery with less urgency than when it connects with the venous duct or the portal venous system.[6,7,10]

Our patients may have died due to blockage of the collector and pulmonary edema.

Surgical correction consists of ligating the descending vein as distally as possible (it may or may not be sectioned), following the direction of its largest tributary, then entering the posterior wall of the left atrium, parallel to the pulmonary venous confluence and performing latero-lateral end anastomososis.[6-11]

The surgical mortality of patients operated on by experts varies between 2 and 20%. This is due to the severity of the malformation, the young age of the patients in question, their low weight, and the rareness of the heart condition. Long-term survival depends largely on a well performed intervention. Death in the first months following surgery is uncommon, and is almost always caused by stenosis of the pulmonary veins.[1-6,12,13]

Between 5 and 10% of patients operated develop stenosis of the anastomosis, with a high incidence of mortality months or years after surgery.

CONCLUSIONS

The study of this condition has been conducted with conventional angiography and cardiac catheterization; the latter has helped to diagnose associated malformations.

Multislice helical computed tomography, with access to graphic processing stations, obtains three-dimensional volumetric images which reveal complex congenital anomalies with high precision. In our three patients the auriculo-ventricular canal and dextroisomerism were corroborated.[14]

The advantages of tomography are that the study is performed in a short time, of 15 to 20 seconds (inspiratory apnea); 320-detector multislice tomographs or dual machines are even faster; among others, angiographic images in arterial and venous phases are obtained, which can be evaluated in different planes; information is obtained on adjacent structures such as: lungs, the lower neck and upper abdomen, all with a single injection of 1-2 mL/kg of body weight of contrast medium. The limitations of multislice tomography are: radiation, although in smaller doses than in angiographic studies, anesthesia, and the use pf contrast medium (minimal amounts). Cases of allergy to water-soluble contrast mediums are rare.[15]

The studies should be interpreted by pediatric radiologists with experience in this kind of abnormalities, as they were in our patients.

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