

Diagnosis and Therapeutics of Pulmonary Arteriovenous Fistula in Childhood. Case Report and Review of the Literature

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We report the case of a 3-year-old female patient, who, since birth, had cyanosis difficult to explain with usual diagnostic tests. The only findings on physical examination were cyanosis and clubbing of her fingers. Chest computerized tomography showed images of excessive attenuation in the right lung, which resembled arteriovenous fistulae that were later confirmed on cardiac catheterization. The fistulous trajectories were then embolized with 7 Gianturco coils, which resulted in an immediate increase in the arterial saturation of blood oxygen.

During the embryological development of the vascular system, which occurs between the 5th and 10th weeks of intrauterine life, a continuous differentiation of the vascular bed occurs, resulting in the creation of separate arterial and venous channels, interconnected by capillaries. When a mistake or halt occurs in this process of vascular differentiation, vascular malformations will appear at different anatomical sites and with variable morphology (fig. 1), depending on the stage of differentiation¹. We report the diagnosis and interventional therapeutics of arteriovenous fistulae in the lungs of a 3-year-old child.

Pulmonary arteriovenous fistulae are characterized by right-to-left shunt of variable magnitude, and the effect of these communications depends on the size of the vessels involved. If the anastomoses affect peripheral arterioles and venules, a small telangiectasia will result, and it usually remains small, not causing hemodynamic alterations in the pulmonary circulation. If larger veins and arteries are affected or if massive involvement of the pulmonary capillaries takes place, an increase in the size of the vessels may occur², resulting in severe hemodynamic alterations. Arterial desaturation of oxygen, cyanosis, clubbing of the fingers, and po-

lycythemia may occur, secondary to the intrapulmonary shunt. Multiple neurological sequelae have been reported due to a variety of infectious, embolic, and hemorrhagic complications³⁻⁵.

The etiology of pulmonary arteriovenous fistulae may be congenital or acquired. The congenital form has 2 varieties: a) cavernous angioma, usually fed by 1 or more tortuous and dilated branches of the pulmonary artery and b) capillary telangiectasia, which forms a net of capillaries and is usually associated with hereditary hemorrhagic telangiectasia⁶ or Rendu-Osler-Weber syndrome. This syndrome is an autosomal dominant vascular dysplasia with variable penetrance, has multiple telangiectasias, and is characterized by recurring hemorrhages. The levels of the coagulation factors are normal and the coagulation mechanism is intact. Approximately 1/3 of the patients diagnosed with this syndrome presents pulmonary vascular malformations. Some studies together with the scientific division of the Hereditary Hemorrhagic Telangiectasia (HHT) Foundation International, Inc. reached a clinical consensus on the diagnostic criteria for this disease; 4 criteria should be sought: epistaxis, telangiectasias, visceral lesions, and a compatible familial history. The diagnosis of Rendu-Osler-Weber disease is established if 3 of the above-cited criteria are present. If less than 2 criteria are present, the diagnosis of the disease is very unlikely, even though children of parents affected should be considered at risk due to the fact that the penetrance varies with age. The clinical investigation of our patient showed only 1 of the cited criteria, which was pulmonary telangiectasia, making the diagnosis of Rendu-Osler-Weber syndrome unlikely.

Pulmonary arteriovenous fistula in its acquired form usually occurs in juvenile cirrhosis, but has also been reported in patients with metastases of thyroid carcinoma and pulmonary schistosomiasis. In the latter, the pulmonary arteriovenous fistula occurs because of the vascular neofor- mation secondary to chronic vasculitis triggered by *Schistosoma* eggs or their metabolic or degrading products.

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English version by Stela Maris C. e Gandour

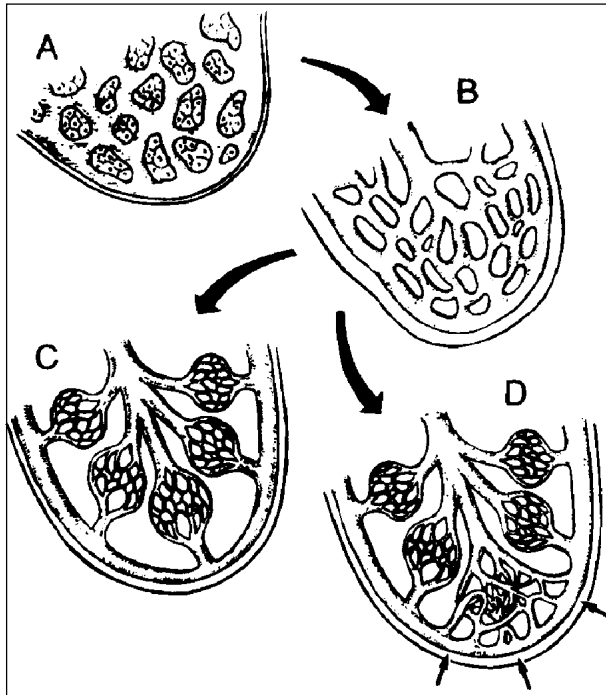


Fig. 1 – Normal stages of vascular development according to Woolard: a) primitive mesenchyma; b) differentiation in plexiform structures; c) disappearance of the primitive elements and differentiation into vascular structures; d) inadequate development during the embryogenesis of the vascular system with persistence of the primitive vascular communications or arteriovenous malformations (small arrows).

Case report

The patient is a 3-year-old white female child with progressive clubbing of her fingers and cyanosis since birth. Physical examination did not disclose other abnormalities and no additional clinical symptoms were reported. Analysis of the arterial gases at room temperature showed pH = 7.5; $PCO_2 = 26$ mmHg; $PO_2 = 36$ mmHg; $CO_2 T = 20.4$ mmol/L; bicarbonate = 19.4 mEq/L; saturation of $O_2 = 75\%$. Hematocrit = 48%; Hb = 15.5 g/dL; MCV = 91. Electrocardiography, echocardiography, and total abdominal echography were within the normal range. The chest X-ray showed an interstitial infiltrate in the right base and perihilar regions.

Continuing with the investigation, a chest computerized tomography was performed and revealed images of excessive attenuation in the right lung, close to the posterior wall, with confluence of the vessels. These alterations raised the possibility of pulmonary arteriovenous fistulae (fig. 2).

Considering that pulmonary arteriovenous fistulae might be present, a cardiac catheterization was performed to elucidate the diagnosis. The angiographic study showed a vascular net with diffuse malformations in the right middle and inferior pulmonary lobes (fig. 3 and 4). Pressures in the cardiac cavities were as follows (in mmHg): RA = 2 (mean); PA = 15 (systolic), 9 (diastolic), 19 (mean); LV = 77 (systolic); Ao = 63 (systolic), 38 (diastolic), 49 (mean). Pulmonary scintigraphy confirmed the results of catheterization.

Because the interventional therapy was chosen, pulmonary arteriovenous fistulae were embolized with catheteri-



Fig. 2 – Computed tomography depicting images of excessive attenuation in the right lung close to the posterior wall with confluence of the vessels determining the possibility of pulmonary arteriovenous fistula.

zation with 7 Gianturco coils, sizes 3 and 5 mm (figs. 5 and 6). The immediate result was an increase in oxygen saturation from 90% to 98%, with the patient sedated receiving oxygen by nasal catheter at the rate of 4 L/min (figs. 5 and 6). At the time of hospital discharge, oxygen saturation measured at room temperature with the pulse oximeter was 90%.

Prior to the embolization procedure of the fistulae, the right pulmonary artery was completely occluded with a balloon of an adequate size for investigating the existence of fistulae in the left lung. This hypothesis was discarded because the hemoglobin saturation suddenly increased from 90% to 100%.

Discussion

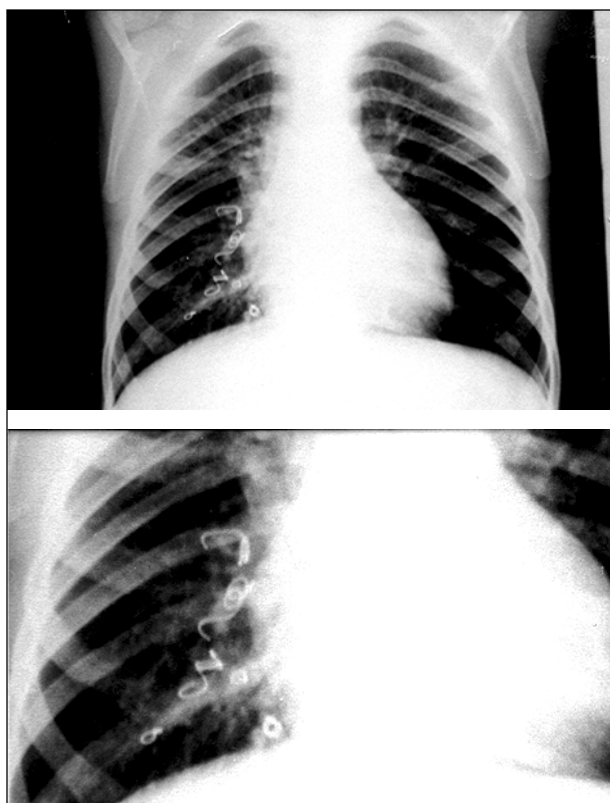
Pulmonary arteriovenous fistula is not easily diagnosed routinely, due to its rarity and its unspecific findings on routine examinations. However, this diagnostic hypothesis should always be considered when examining children with cyanosis, in whom the initial tests do not detect cardiac abnormalities.

Two other alternatives in the diagnostic investigation, which were not used in the present case, are the calculation of the shunt through the saturations measured with catheterization in the different cardiac chambers⁷ and contrast echocardiography^{8,9}, in which agitated saline solution, for example, can be injected in a peripheral vein. The microbubbles resulting from this process and detected in the left atrium determine a right-to-left shunt through the lungs. The capillary alveolar filter does not allow the passage of these microbubbles.

Tc-99m MAA total body imaging^{10,11} follows the same principle, because the macroaggregates of albumin labeled with technetium-99 measure from 20 to 60 μ m, but the diameter of the pulmonary capillary bed is smaller than 15 μ m. Contrast medium uptake in other organs, in addition to the lungs, would show the presence of the shunt, and the pulmonary area with no radionuclide would delimitate its site¹².



Figs. 3 and 4 – Pulmonary arteriography showing multiple pulmonary fistulae.



Figs. 5 and 6 – Chest X-ray showing the coils (left) and one widened image of the right inferior lobe (right).

In the case we report, the diagnosis was established by chest spiral computed tomography, pulmonary scintigraphy, and selective pulmonary angiography.

Interferon has been suggested as a less invasive therapeutic approach, but, until the present time, its efficacy for the treatment of pulmonary arteriovenous fistula has not been scientifically confirmed.

Another alternative would be occlusion of each pulmonary lobe with a balloon, and concomitant assessment of blood saturation, therefore checking whether selective embolizations with detachable balloon¹³⁻¹⁵ could replace pneumectomy¹⁶. Embolization with coils has also been successfully used¹⁷⁻²⁰, even in the patients with telangiectasia²¹.

Chest computed tomography performed after the procedure could serve to confirm the success of embolization²², and the quantification of the percentage of the reduction in the shunt could be done with pulmonary scintigraphy with Tc-99m MAA^{10,11}, comparing the rate of short-circuit established prior to the procedure.

In our case, we chose the interventional approach, with multiple embolizations with coils, because of the great number of fistulous trajectories, and the result was considered satisfactory. Andersen et al²³ and Shioyia et al²⁴ had similar results.

Finally, it is worth emphasizing that blood saturation may decrease with time as a consequence of opening new fistulae because of the progressive character of the disease.

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