

Fig. 1.

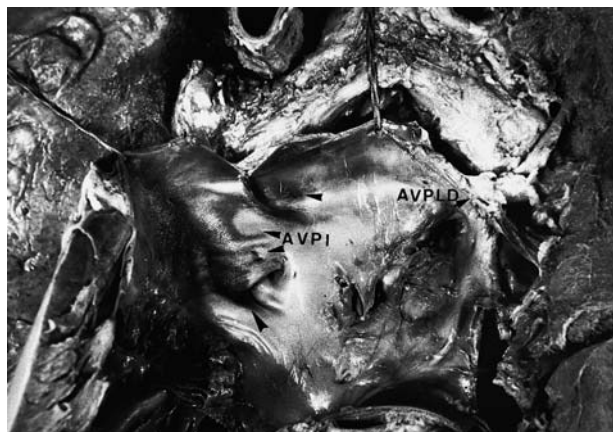


Fig. 2.

## Bilateral Pulmonary Veins Atresia

Congenital atresia of the pulmonary veins is a rare anomaly that is not associated with other heart disease. It has a poor prognosis and patients die secondary to pulmonary hypertension, recurrent hemoptysis, pulmonary edema, or congestive cardiac insufficiency.

We present a 5-year-old male patient admitted to the hospital for asthenia, anorexia, hemoptoic vomiting, and a severe cough. Marked dystrophy was observed, with a weight of less than the 3rd percentile, pale skin

and mucous, and bilateral nystagmus. Cardiovascular examination did not reveal murmurs and the second heart sound was strong. With respect to echocardiogram data, on 4-chamber projection the connections of the left pulmonary veins and the right superior vein lobe to the left atrium were not seen; in the short axis projection the great vessels were in a normal relationship, with the diameter of the pulmonary artery being greater than the diameter of the ascending aorta, the

right pulmonary branches and the inferior lobe were dilated, and there was a hypoplastic left pulmonary branch. In the days following admission, a hemodynamic study was performed, which confirmed the existence of moderate pulmonary hypertension (average pressure 29 mm Hg). The buried pulmonary pressure in the left artery was 16 mm Hg and was 26 mm Hg in the right superior lobe. Pulmonary arteriography revealed hypoplasia of the left pulmonary arteries and the right superior lobe vein with the left atrium (Figure 1B). The right inferior lobe pulmonary artery was dilated. The patient was re-admitted 9 times during the following year, dying during the last admission with congestive cardiac insufficiency that did not respond to treatment. The diagnosis was confirmed on autopsy. The right inferior lobe vein was dilated. There were no

connecting orifices between the left pulmonary veins (Figure 2; AVPI: arrow) left appendage orifice) and superior right lobe vein with the left atrium (Figure 2; AVPLD).

Congenital atresia of the pulmonary veins is caused by a defect in the incorporation of the common pulmonary vein with the left atrium. In some cases the atresia is limited to the connection area or to a short segment. If the affected area is lobar vein, lobectomy may resolve the situation. Thirty case reports have been published, none of which had bilateral atresia.

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