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 hemodyna-
ic study was performed, which confirmed the exist-
tence 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 reve-
aled 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 dila-
ted. The patient was re-admitted 9 times during the fol-
lowing 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 pulmo-
nary vein with the left atrium. In some cases the atre-
sia is limited to the connection area or to a short seg-
ment. If the affected area is lobar vein, lobectomy may
resolve the situation. Thirty case reports have been pu-
lished, none of which had bilateral atresia.

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