

Pulmonary Hypertension Secondary to Pulmonary Artery Branch Stenosis in a 2-Year-Old Infant

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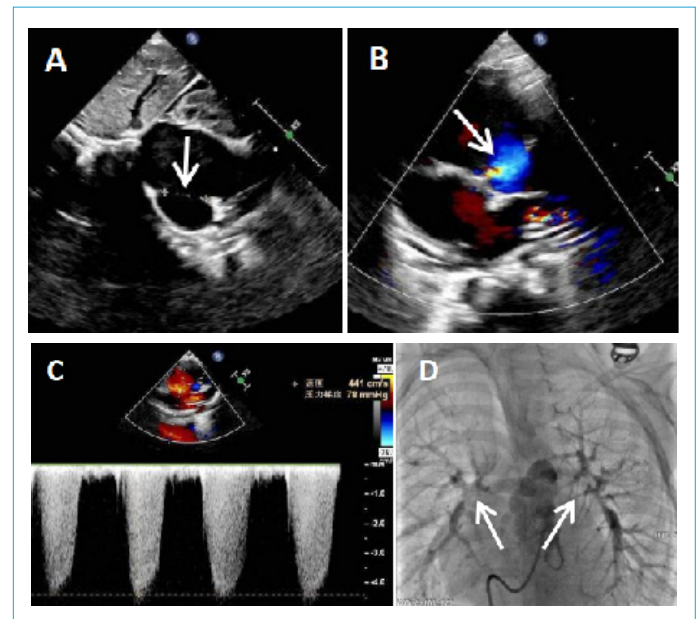
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Description

The patient admitted to the hospital is a 2-year-old girl with cyanosis. The SPO_2 is about 87% (in-room air) across all four limbs. Echocardiography showed Atrial Septal Defect (ASD, 17 mm, Panel A, arrow), ventricular septal defect (VSD, 8.5 mm, Panel B, arrow), and severe pulmonary hypertension (Panel C). Electrocardiogram reveals high voltage in the right ventricle. Other laboratory tests yield normal results. The cardiac catheterization revealed multiple segment stenosis in the left and right pulmonary artery branches (Panel D, arrow). Total pulmonary vascular resistance is calculated at 6.81 Wood Units per square meter. The whole exome gene test identified a copy number deletion of approximately 1.42 Mb on the long arm of chromosome 7q11.23 (copy number = 1), suggesting a pathogenic mutation associated with Williams-Beuren Syndrome (WBS).

Based on auxiliary examinations, it was determined that severe pulmonary hypertension resulted not only from VSD and ASD but primarily from pulmonary artery stenosis. Surgical repair for VSD and ASD is not suitable for patients with severe pulmonary hypertension caused by multiple segment pulmonary artery branch stenosis. Balloon dilation can be performed to alleviate obstruction caused by pulmonary stenosis.

In children, secondary pulmonary hypertension is less common compared to adults. However, it can easily go undiagnosed when combined with large left-to-right shunt Congenital Heart Disease (CHD). Therefore, evaluating secondary factors such as pulmonary artery stenosis is crucial in cases involving severe pressure within the pulmonary artery due to large left-to-right shunt CHD.



Declarations

All authors declare no conflict of interest regarding this contribution.

The corresponding author holds all data sets used and analyzed during this study.