



**Fig. 2.** Chest X-ray (A) showing a widened mediastinum, enlarged pulmonary hila and interstitial involvement. Chest computed tomography confirming presence of large mediastinal lymphadenopathies, and bilateral hilar (B, coronal reconstruction with intravenous contrast medium) and pulmonary parenchymal (C, axial image in lung window) involvement. Chest X-ray (D) obtained 6 months after starting corticosteroid treatment showing normalization of findings.

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## Pulmonary Arterial Hypertension in Congenital Heart Defects With Left-To-Right Shunt<sup>☆</sup>



### *Hipertensión arterial pulmonar en cardiopatías congénitas con cortocircuito izquierda-derecha*

To the Editor:

We read with interest the article published by Hernández Vázquez et al.<sup>1</sup> on partial anomalous pulmonary venous connection with pulmonary hypertension, and we would like to put forward some thoughts that may be of interest.

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Firstly, significant pulmonary arterial hypertension (PAH)<sup>2</sup> rarely develops in association with heart defects with left-to-right shunt and volume overload, such as partial anomalous pulmonary venous connection or drainage (PAPVD) and interatrial communication (IAC), in contrast to what the authors appear to claim in their discussion. PAPVD patients usually have normal pulmonary blood pressure, unless they have 2 pulmonary veins with abnormal return, or else sinus venous ICA with a pulmonary to systemic blood flow ratio of 1.5:1 (Qp:Qs>1.5:1). Even in this situation, severe PAH is very rarely observed (less than 5%), and is more related to genetic predisposition (as in primary PAH) than to the extent of the shunt itself.<sup>3</sup> In contrast, when the left-to-right shunt is associated with volume and pressure overload (interventricular communication, patent ductus arteriosus, aortopulmonary window or common arterial trunk), PAH frequently becomes severe if the defect is large and left uncorrected during the first year of life. Thus, shunts resulting in volume overload are more typically found

in asymptomatic or practically asymptomatic patients with right cavity dilation and mild or moderately increased pulmonary pressure, while systemic or suprasystemic pulmonary pressures with shunt reversal are found in shunts with both volume and pressure overload (Eisenmenger's syndrome).

Secondly, in PAPVD patients with significant PAH, other etiologies must be ruled out. These include respiratory disease, obstructive sleep apnea, connective tissue disease, chronic thromboembolism, portal hypertension, human immunodeficiency virus, use of appetite suppressants or toxic substances, and left heart disease. In the latter, patients with multiple cardiovascular risk factors, as discussed in the article, or coronary heart disease (not specified by the authors) may develop left ventricular diastolic dysfunction and secondary pulmonary arterial hypertension, even in the absence of evidence of heart failure.<sup>4</sup> Special precautions must be taken in these cases, since PAPVD can add to left atrial decompression and total correction of the defect may lead to a sudden increase in pulmonary venous pressure and the development of pulmonary edema.<sup>5</sup>

Finally, before PAPVD or IAC correction is attempted, pulmonary vascular and systemic resistances and pressures must be identified, in order to prevent converting an Eisenmenger's syndrome into a disease with a similar course as that of primary PAH, with its correspondingly poorer prognosis.<sup>3,5</sup>

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### Conflict of Interest

The authors declare that they had no conflict of interests.

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