Sirs: Partial anomalous pulmonary venous drainage (PAPVD) of the left upper pulmonary vein (LUPV) is a rare congenital anomaly leading to left to right shunt. Embryogenesis is thought to consist of non-development of the connection between LUPV and left atrium and the persistence of a primitive “vertical” vein between LUPV and the left brachiocephalic (innominate) vein [1]. The lesion must not be confused with a persistent left superior caval vein with connection to the left atrium. Standard treatment is surgical redirection of the LUPV to the left atrium [2]. This is the first description of interventional treatment of this lesion.

Methods and patients

Retrospective observational report. Six patients with PAPVD of the LUPV (0.25%) were identified out of 2400 patients who underwent cardiac catheterizations for congenital heart disease performed from 01/2000 to 09/2006. Age ranged from 1.5 to 35 years (median 7.5 years). In three of the six patients (50%), there was an additional connection to the left atrium, which could be identified by contrast injection into the vertical vein after superior balloon occlusion. All three of them had larger left to right shunts (Qp:Qs > 2:1) than to be expected from PAPVD alone. No patient had pulmonary hypertension.

Echocardiography showed enlarged right atria and ventricles in the absence of atrial septal defects in all patients. Dilated superior vena cava and abnormal inflow into the left brachiocephalic vein allowed echocardiographic diagnosis in four, but were initially missed in the other two. Magnetic resonance imaging (MRI) performed in one patient demonstrated beautifully the pathologic pulmonary venous connection.

Management

All three patients with PAPVD of the LUPV without connection to the left atrium (Fig. 1) were treated surgically by redirection of the pulmonary vein to the left atrial appendage. There was no development of pulmonary venous stenosis during follow-up as confirmed by echocardiography and chest radiography.

Surgical ligation of the vertical vein was performed in the first patient with an additional connection to the left atrium. Transcatheter device closure of the superior part of the vertical vein led to successful cor-
Fig. 1 (frontal views) 7-year-old girl. Qp:Qs 2:1. a The left upper pulmonary vein drains via a mildly stenotic vertical vein into the innominate vein, but is also connected to the left atrium. b Balloon occlusion (arrow) of the connecting vertical vein with an endopen Berman catheter showed no increase of pressure in the pulmonary vein. c Coil occlusion (asterisk) at the stenosis of the vertical vein abolished the left to right shunt completely. BCV left brachiocephalic (innominate) vein; LA left atrium; LUPV left upper pulmonary vein; VV vertical vein

Fig. 2 (frontal views) 3-year-old boy. Qp:Qs 3:1. a Contrast injection into the vertical vein showed flow to the innominate vein, the superior caval vein and the right atrium. b Contrast injection into the left atrium. The left upper pulmonary veins are connected to the vertical vein and to the left atrium. Despite a mild stenosis (asterisk) there is significant left to right shunt. c An Amplatzer duct occluder was used to occlude the vertical vein just caudal to the stenosis. d A pulmonary angiogram confirmed normal pulmonary venous return and complete occlusion of the vertical vein. ADO Amplatzer duct occluder; BCV left brachiocephalic (innominate) vein; LA left atrium; LUPV left upper pulmonary vein; RA right atrium; SVC superior caval vein; VV vertical vein