

VARIATIONS OF PULMONARY VEINS AND THEIR CLINICAL RELEVANCE

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Variations of pulmonary veins (PV) were previously considered rare, only documented as isolated case reports. Recent cadaveric, autopsy, CT, MDCT and MRI studies have revealed substantial variations with respect to their number and drainage pattern into the left atrium (Marom et al., 2004; Calkins et al., 2007; Wannasopha et al., 2012; Harbi et al., 2014; Rajguru and Fulzele, 2016). The most commonly described variations include supernumerary or less pulmonary veins and the corresponding arterial ostia. The article in this issue (Kinfemichael and Dawit, 2016) presents a case of 5th pulmonary vein originating from the middle lobe of the right lung. This is consistent with previous reports of supernumerary pulmonary veins and ostia (Marom et al., 2004; Calkins et al., 2007; Prasanna et al., 2014). These vary from 2 – 7 (Wei et al., 2014; Yuan et al., 2015; Rajguru and Fulzele, 2016; Kumzel – Piotrowska et al., 2016). The commonest cause of such variations is the right middle pulmonary vein (Calkins et al., 2007; Klimek – Piotrowska et al., 2016). These variations have informed various classifications based on number of pulmonary veins and ostia (Marom et al., 2004). One of the most straight forward classifications was proposed by Shukla et al (2012). The principle of this classification in the table below may be extended to include, say type VI or VII depending on the number of veins.

Classification of pulmonary venous system [Shukla et al., 2012]

Type [Left / Right]	Description
I	1 PV with 1 ostium
II A	2 PV with 2 Ostia
II B	2 PV with 1 ostium
III A	3 PV with 3 ostia
III B	3 PV with less than 3 ostia
IV A	4 PV with 4 ostia
IV B	4 PV with less than 4 ostia
V A	5 PV with 5 ostia
V B	5 PV with less than 5 ostia

These variations have an embryological basis. Pulmonary veins develop initially as vascular plexi, by vasculogenesis and angiogenesis, from mesenchyme surrounding lung buds and their derivatives. The plexi are remodeled to form venous channels which coalesce to form intraparenchymal vessels that drain into four definitive PVs (Webs et al., 2001; Hall et al., 2002). These venous systems establish connection with the primitive left atrium (LA) initially through a common pulmonary vein (CPV). This CPV develops from solid pharyngeal strands of mesenchyme referred to as mediastinal mesocardium which canalizes secondarily (Webs et al., 2001; Moorman, 2003; Moorman and Anderson, 2011). As the cavity of the LA expands, the CPV merges with the wall of the LA and the four PVs eventually become incorporated into the LA, giving the 4 pulmonary ostia (Sherif, 2013). The pulmonary venous system is, thus, developmentally and morphologically distinct from systemic veins (Moorman and Anderson, 2011; Sherif, 2013). Variations may arise from abnormal fusion, coalescence and incorporation of the pulmonary venous system into the atrial chambers. Supernumerary veins with independent ostia, for example, arise by over – incorporation of pulmonary veins beyond their first division (Marom et al., 2004; Prasanna et al., 2014)

Variations of PV are important because of their arrhythmogenic potential (Marom et al., 2004; Calkins et al., 2007; Buttner et al., 2011; Harbi et al., 2014). The variant veins are a source of ectopic beats which may predispose to thromboembolic complications, left ventricular overload, atrial remodeling and cardiac dysfunction (Rajeshwari and Ranganath, 2012). For radiofrequency, catheter and balloon based pulmonary vein ablation in atrial

fibrillation, they are also important to guide mapping, preablation planning and evaluation of post ablation complications (Rajeshwari and Ranganath, 2012; Aktan et al., 2014; Mayor, 2015; Nalliah et al., 2015). Further, their knowledge is critical to avoid inadvertent injury during general cardiothoracic surgery, thoracoscopic pulmonary lobectomy, cardiac valve replacement and to minimize misinterpretation of pulmonary venograms; inadvertent PV ablation and perioperative bleeding (Arslan et al., 2008; Shukla et al., 2012). Indeed, variability in pulmonary venous anatomy can substantially alter the success rate of radiofrequency cardiac ablation (Wellens, 2015). There is therefore need for pre – procedure CT and MRI evaluation of the pulmonary venous system.

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