Cardiovascular Risks in Patients with Loeys–Dietz Syndrome

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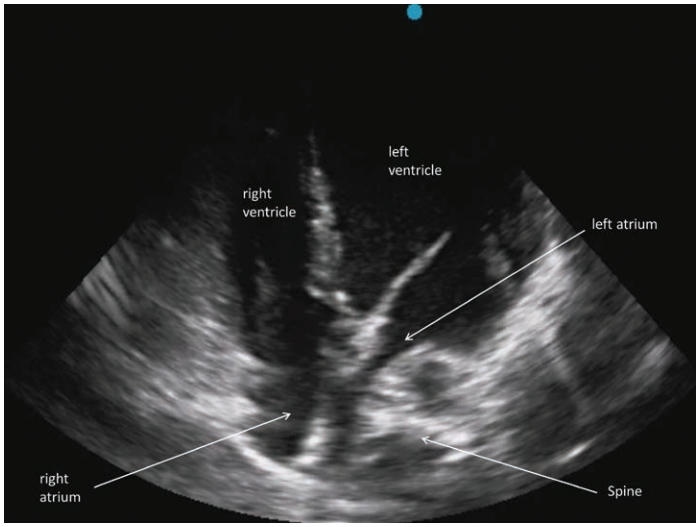

Compression of the left atrium is well described in cases of mediastinal masses, pericardial effusion, or aortic aneurysms. Here, we present a transthoracic apical four-chamber view of a patient with Loeys–Dietz syndrome that displays a severe slit-like compression of the left atrium between the sternum and the thoracic spine (Supplemental Digital Content, http://links.lww.com/ALN/C263). Loeys–Dietz syndrome is a Marfan-like autosomal dominant connective tissue disorder which typically includes aneurysmal aorta or adjacent arterial branches.[1] The vascular course is aggressive, with complications developing at smaller aortic dimensions compared to Marfan syndrome. Many patients die in their 20s due to aortic dissection. Up to 60% of patients with Loeys–Dietz syndrome show scoliosis, which can compress the heart, particularly when manifesting with concomitant *pectus excavatum*.[2] Apart from cardiovascular risks, airway management can be complicated in these patients by craniosynostosis, cleft palate, retrogenia, and cervical spine instability. Cervical spine manipulation should be minimized by using video laryngoscopy or fiberoptic intubation for airway management.[3]

In patients with Loeys–Dietz syndrome, complete aortic imaging is recommended at the time of the initial diagnosis and every 6 months thereafter.[1] In addition, preoperative transthoracic echocardiography for evaluation of aortic root and cardiac function should be strongly considered. In patients with severe cardiac compression, all maneuvers to improve cardiac preload should be employed. These include left lateral or Trendelenburg positioning, liberal fluid management, and avoidance of high mean airway pressures. Adequate ventricular filling can be sufficiently monitored by repeated intraoperative echocardiography.

Competing Interests

Dr. Magunia has received a speaker's honorarium from CSL Behring GmbH (Marburg, Germany). The other authors declare no competing interests.

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