Loeys-Dietz: Anything special for tomorrow's case?

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In this issue of the *Journal*, Zhang and colleagues¹ describe their experience anesthetizing 2 children with Loeys-Dietz Syndrome (LDS) for aortic aneurysm repair with circulatory arrest. This is the first report of anesthesia for pediatric patients with LDS undergoing aortic aneurysm surgery. Although this report lacks many of the fine details of the anesthetic that would allow close replication of their efforts, Zhang and colleagues¹ do highlight several challenging aspects of anesthetizing pediatric patients with LDS. Hemodynamic control is paramount to avoid rupture of these delicate aneurysms. Zhang and colleagues¹ achieved this with a seemingly routine opioid-based anesthetic and were prepared to administer 2 supplemental antihypertensive agents. In addition, this report highlights the utility of bronchial blockers for achieving single-lung ventilation in patients of insufficient size to use double-lumen endotracheal tubes. Finally, gentle airway management was justified by the concerns for joint laxity. The craniofacial manifestations of LDS, although worth mentioning, did not prohibit routine intubation.

LDS is a rare genetic connective tissue disorder first described in 2005 in which patients have characteristic craniofacial findings and may present with aneurysm formation throughout the arterial system at relatively young ages.² The observation that these aneurysms have a propensity to rupture even when small translates into the need for careful surveillance and often surgical intervention early in life.³

The bronchial blocker is positioned to achieve single-lung ventilation.

Central Message

Loeys-Dietz syndrome is a rare connective tissue disorder. Familiar and carefully conducted anesthesia techniques can lead to success.

See Article page e53.

Anesthesiologists may initially be justifiably dyspeptic when tasked with anesthetizing a young child with a rare, recently described connective tissue disorder and dramatic aortic aneurysm. Johnson and colleagues⁴ have previously provided a detailed account of their perioperative considerations when faced with management of a young adult with LDS undergoing aortic aneurysm repair. Zhang and colleagues¹ add to this with their experience caring for 2 pediatric patients. What we can surmise from these reports is that careful anesthesia with familiar techniques can result in successful outcomes for these patients.

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