A 3-day-old full-term infant was transferred to our pediatric intensive care unit because of significant biphasic stridor with desaturation spells. The history revealed an uncomplicated pregnancy and normal spontaneous vaginal delivery. The mother was known to have hypothyroidism caused by Hashimoto thyroiditis (negative thyroid-stimulating immunoglobulin). Apgar scores were 6 and 9 after 1 and 5 minutes; no resuscitation was needed. Umbilical arterial and venous blood gas values showed marked acidosis (pH 6.81 and 6.88, respectively) without signs of encephalopathy. General and cardiovascular examinations were normal. In particular, there was no marfanoid habitus, heart sounds were normal, and peripheral pulses were preserved. Chest x-ray illustrated a pronounced cardiac silhouette with signs of a normal pulmonary circulation (Figure 1). Echocardiography showed a structurally normal heart, left aortic arch (without signs of airway compression), and no double aortic arch. However, the entrance orifice of the ductus arteriosus was enlarged, tapering to a small tortuous channel at the site of pulmonary artery insertion where the exit orifice was small, with restrictive left-to-right shunt (Figure 2). Hence, a diagnosis of ductus arteriosus aneurysm (DAA) was made. Computed tomography angiography scan confirmed this finding, with a ductal diameter of 9 mm and length of 11 mm (Figure 3). Fiberoptic laryngoscopy under general anesthesia and spontaneous breathing was performed and revealed left and partial right vocal cord paralysis, possibly resulting from compression of the left recurrent laryngeal nerve by the DAA (Movie I in the online-only Data Supplement). The patient was intubated and ventilated. Tracheotomy was performed, and ventilation was weaned. In the following weeks, echocardiography demonstrated closing of the ductus arteriosus and regression of the aneurysm size. Three months later, repeated laryngoscopy revealed normal movement of the vocal cords.

Congenital DAA, a rare condition caused by saccular dilatation of the ductus arteriosus, can potentially be fatal because of the risk of infection, dissection, rupture, or thrombus formation and embolization. It can also compress adjacent structures such as the recurrent laryngeal nerve. Signs and symptoms are usually scarce and discrete. If symptoms occur, the child usually presents before 2 months of age. The incidence of DAA is somewhat unclear, with a reported incidence of 0.3% to 1% in neonatal autopsies. Surgical resection could be considered if DAA remains patent beyond the neonatal period, if DAA is associated with connective tissue disease, if there is evidence of thromboembolism, or if significant compression of adjacent structures exists. Surgical excision, however, carries additional risk for (further) nerve damage. Ligation and decompression of DAA is another option. Spontaneous regression occurs, as seen previously and in the present case. Therefore, a “wait and see” approach can be a justified policy in some patients.

Disclosures
None.

References
Figure 2. Echocardiography illustrating the enlarged entrance orifice of the ductus arteriosus aneurysm (DAA) with the small tortuous channel at the site of left pulmonary artery. DA indicates descending aorta; EO, exit orifice of the ductus aneurysm to the pulmonary artery; and LPA, left pulmonary artery.

Figure 3. Computed tomographic angiography with 3-dimensional reconstruction (A), coronal (B), and sagittal (C) multiplanar reconstruction confirming the diagnosis of ductus arteriosus aneurysm (DAA).
Ductus Arteriosus Aneurysm and Vocal Cord Paralysis
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Circulation. 2015;131:1713-1714
doi: 10.1161/CIRCULATIONAHA.114.013568

The online version of this article, along with updated information and services, is located on the World Wide Web at:
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