A one-day-old term female infant born via elective cesarean section develops noisy breathing after feeding in the newborn nursery. On exam, you notice biphasic stridor, suprasternal retractions and tachypnea with clear breath sounds, no murmur, and a preductal oxygen saturation of 98%. You administer CPAP of 5 cm H2O with improvement in her respiratory distress. The infant has a normal chest radiograph and echocardiogram, but a bronchoscopy demonstrates compression of the distal trachea, so she undergoes a computerized tomography angiogram (CTA) (Figure 1).

Figure 1: Kalane S, Santosh JA. Newborn with congenital stridor. Neoreviews. 2021;22(12):e854. 10.1542/neo.22-12-e8541

What is the most likely diagnosis?

A. Double aortic arch
B. Left aortic arch with aberrant right subclavian artery
C. Left pulmonary artery sling
D. Right aortic arch with aberrant left subclavian artery and left-sided ligamentum arteriosum

Explanation:

Answer: A. Double aortic arch

The infant in this vignette presents with biphasic stridor shortly after birth with CTA imaging findings consistent with a double aortic arch (DAA) (Option A). Stridor is a high-pitched respiratory sound suggestive of an airway anomaly. The causes of stridor in a newborn can be categorized into three groups: 1) extrathoracic pathologic conditions at the supraglottic level
that can present with inspiratory stridor (eg, laryngomalacia), 2) intrathoracic conditions at the glottic or subglottic level that can present with biphasic stridor (eg, subglottic stenosis, vascular ring), and 3) tracheal pathologic conditions that can present with expiratory stridor (eg, tracheomalacia). Our patient presented with biphasic stridor suggestive of a fixed proximal airway obstruction at the laryngeal or tracheobronchial level, which can be secondary to multiple causes such as a vascular ring, subglottic stenosis, subglottic hemangiomas, vocal cord paralysis, or laryngeal webs.

The infant in our vignette underwent a bronchoscopy that demonstrated extrinsic compression of her distal trachea prompting a CTA that demonstrated a complete vascular ring consistent with a DAA (Figure 2). Vascular rings occur in about 1 in 10,000 births with a mean age of diagnosis around six months. A DAA is the most common type of symptomatic vascular ring due to the persistence of the right fourth aortic arch and a hypoplastic or even atretic left arch that encircle the trachea and esophagus. Tracheal and/or esophageal compression leads to early symptoms in the newborn period or early infancy, including biphasic stridor, respiratory distress, and feeding difficulties. A DAA is typically an isolated anomaly but can be associated with intracardiac defects (eg, ventricular septal defects, transposition of the great arteries, truncus arteriosus, Tetralogy of Fallot, or coarctation of the aorta) in 1% of neonates. Surgical correction by ligating the duplicated arch is necessary and curative since affected patients are often symptomatic.

**Figure 2**  A) Coronal section of computed tomography angiogram demonstrating a double aortic arch (blue arrows) encircling the trachea (yellow arrow). (B) Computerized tomography reconstruction arteriography demonstrating the right aortic arch (red arrow) and left aortic arch (green arrow) forming a complete vascular ring. From: Kalane S, Santosh JA. Newborn with congenital stridor. *Neoreviews*. 2021;22(12):e854. 10.1542/neo.22-12-e854

A left aortic arch with aberrant right subclavian artery (Option B) is a type of incomplete vascular ring. Infants with this vascular anomaly are predominantly asymptomatic but can present with feeding difficulties such as dysphagia later in life secondary to extrinsic esophageal compression. This anomaly occurs following the regression of the right fourth aortic arch between the right common carotid and right subclavian artery rather than distal to the subclavian artery. The aberrant subclavian artery persists as a branch from the descending aorta and courses posterior to the esophagus. Up to 40% of patients with Trisomy 21 have this anomaly, which is also associated with congenital heart disease, including Tetralogy of Fallot, coarctation of the aorta, or interrupted aortic arch. Surgery is indicated in patients who are
symptomatic and involves reconstruction of the right subclavian artery to avoid a “subclavian steal” phenomenon. Our patient had an isolated vascular ring with respiratory and feeding symptoms notable at birth with imaging confirming a complete vascular ring, therefore ruling out a left arch with aberrant right subclavian artery.

A left pulmonary artery sling (Option C) is another type of an incomplete vascular ring that occurs when an aberrant left pulmonary artery arises from the right pulmonary artery, courses over the right mainstem bronchus, and then forms a sling around the trachea. As a result, compression of the right bronchus can lead to atelectasis or hyperinflation of the right lung due to a ball-valve phenomenon (Figure 3). In up to 50% of cases, pulmonary artery slings are associated with complete cartilaginous rings in the distal trachea, resulting in congenital tracheal stenosis known as the “ring-sling complex.” Most patients have respiratory signs by the first month of age, and more than 50% of patients have accompanying tracheobronchial anomalies such as tracheomalacia, airway stenosis, tracheal cartilage rings, or hypoplasia of tracheal segments. Intracardiac defects are present in 50% of patients and include atrial and ventricular septal defects, patent ductus arteriosus, single ventricle, aortic arch anomalies, and left superior vena cava. Surgery is curative to correct this lesion; however, almost 50% of patients continue to have some degree of long-term airway obstruction. The infant our vignette had a normal chest radiograph with no evidence of right-sided lung pathology or tracheal narrowing with imaging consistent with a DAA instead of an LPA sling.

Figure 3: Coronal view of computed tomography in a patient with a left pulmonary artery sling showing a) trachea, b) left pulmonary artery with an abnormal course around the trachea, c) stenosis of the right bronchus, d) areas of stenosis of the right main bronchus, e) atelectasis of upper lobe of the right lung. From: Dumpa V, Gupta P, Iqbal V, Nair J. Term neonate with respiratory distress. *Neoreviews*. 2017;18(10):e611–614

The second most common cause of a complete vascular ring following a DAA is a right aortic arch with aberrant left subclavian artery and left-sided ligament arteriosum (Option D) in which the trachea and esophagus are encircled by the ascending aorta anteriorly, the aortic arch on the right, the descending aorta posteriorly, and the ligamentum arteriosum and the left pulmonary artery on the left. Unlike a DAA, this type of vascular ring is looser and may not present with symptoms until after the neonatal period. A right aortic arch may initially be
identified on a chest radiograph if the aortic knob is noted to be on the right side of the sternum instead of the left (Figure 4), while a CTA or magnetic resonance angiography is diagnostic. Approximately 10% of patients have accompanying intracardiac defects such as Tetralogy of Fallot or truncus arteriosus. Surgical repair consists of ligation of the ligamentum arteriosum.

**Figure 4:** Anteroposterior chest radiograph of an infant with stridor who had a right aortic arch (blue arrow) found to have a vascular ring. From Baia C, Patra KP, Lambert B, Meyer R, Bajaj K, Samanich J, Dar P, Rosen O, Vyas H, Morrow WR. Index of suspicion in the nursery. *Neoreviews.* 2010;11(8):e447–452

**Did you know?**

- Chromosome 22q11 deletions occur in up to 20% of patients with vascular rings, thus routine screening with echocardiography or other imaging studies (eg, CT or MRI) should be considered to identify aortic arch sidedness and the branching pattern in these infants.

**What is the most common venous anomaly that can occur in infants with a double aortic arch?**

To find the answer, please refer to the following article: Gala M, Ghoneem A, Haffez M, El Bolkini M. An uncommon peripherally inserted central catheter position with an uncommon diagnosis. *Neoreviews.* 2020;21(11): e765–767

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