We report a case of Kommerell diverticulum with right-sided aortic arch and anomalous origin of neck vessels which presented as recurrent apneic spells and choking attacks after feeding.

**CASE DETAILS**

**Clinical presentation**

A 3-week-old neonate presented with history of apneic spell twice each lasting more than one minute associated with floppiness and bluish discoloration all over the body. Her mother gave history of recurrent choking attacks and vomiting after feeding since birth. The vomiting is non-projectile, nor blood or bile tinged. Her birth history was unremarkable (spontaneous vaginal delivery, birth weight 2.9 kg). No postnatal complications were noted. Currently, she is on combination of breastfeeding with formula feed.

On arrival, she was afebrile, heart rate 130/min, respiratory rate 42/min, SpO2 99% and weight 3.8 kg. On clinical examination she was alert, not in distress. All biochemical and haematological investigations were normal.

**DIAGNOSTIC WORKUP**

Chest X-ray in emergency department showed straightening of the left cardiac border with loss of left aortic contour and prominent right para tracheal soft tissue. (Figure 1)

Decision was made to admit the patient for close observation and workup for apneic episodes including echocardiography, upper GI contrast study and CT angiography of chest.

Echocardiography showed normal left aortic arch with aberrant innominate artery from proximal descending aorta. Upper GI contrast study showed postero-lateral indentation on the contrast column at the oesophagus just above the level of tracheal bifurcation opposite 5th/6th thoracic vertebra. However, normal distal flow of the contrast was noted (Figure 2).

CT angiography revealed right-sided aortic arch with aberrant origin of left subclavian artery which arises from a dilatation at its origin (diverticulum of Kommerell) and significant narrowing of proximal part of the left subclavian artery. Left common carotid artery is arising from the right-sided aortic arch assuming a transverse oblique course anterior to the trachea. Aberrant left subclavian artery courses posterior to oesophagus seen on the left side-making a vascular ring. (Figure 3).

Preoperative flexible bronchoscopy and upper GI endoscopy was also performed to rule out any intraluminal pathology which were unremarkable.
Diagnosis was made of right-sided aortic arch with aberrant left subclavian artery arising from diverticulum of Kommerell and ligamentum arteriosum with tracheal and oesophageal compression. Surgery was planned and through left lateral thoracotomy division of ligamentum arteriosum, excision of Kommerell diverticulum, aortopexy and esophageal release was done. Baby was stable after the surgery, stayed in ICU for close cardiorespiratory monitoring and discharged on 2nd post-operative day in stable condition.

DISCUSSION
Kommerell Diverticulum is the remnant of 4th dorsal aortic arch first described by Burckhard Kommerell, as aneurysmal dilatation of descending aorta at the origin of aberrant left subclavian artery (ALSA).1,2

Three types of aortic arch diverticulum have been described in literature.3

1. Aortic Diverticulum in Left Aortic Arch with Aberrant Right Subclavian Artery
2. Aortic Diverticulum in Right Aortic Arch with Aberrant Left Subclavian Artery
3. Aortic Diverticulum at the Aortoductal Junction

Based on Edwards classification, right-sided aortic arch can be further classified into three major subgroups4:

1. Type I includes right-sided aortic arch with mirror image arch branches.
2. Type II includes right-sided aortic arch with aberrant left subclavian artery and Kommerell diverticulum.
3. Type III includes right-sided aortic arch with isolated left subclavian artery communicating with the pulmonary artery.

Kommerell diverticulum (KD) with right-sided aortic arch (RAA) usually seen in 0.05–0.1% of the population.1 The left
ligamentum arteriosum (LLA) joins the root of aberrant left subclavian artery (ALSA) to the left pulmonary artery (LPA). This along with RAA forms a vascular ring and typically seen with normal intracardiac anatomy. Our case was type II with significant stenosis at the origin of ALSA and aberrant origin of neck vessels from RAA. No intracardiac abnormalities were detected. Depending on severity, this vascular ring can cause symptoms of tracheal or oesophageal compression. In paediatric population, due to pliability of immature tracheal rings respiratory symptoms predominate. However in adults, common presentation is after the age of 40 years when aortic wall becomes atherosclerotic and rigid which can result in dysphagia, chest pain, cough and lower respiratory infection.

Careful evaluation of aortic arch anatomy is crucial in planning for thoracic surgery and endovascular intervention, as well as to make decisions regarding repair of the coexisting lesions in the same procedure. At present, non-invasive modalities like echocardiography, cardiac magnetic resonance angiography (MRA) and CT angiography (CTA) has largely replaced invasive modalities like catheter angiograms in the diagnosis of vascular ring. In children, X-ray chest and echocardiography are considered as initial investigations in diagnosing congenital aortic arch anomalies. In patients presenting with dysphagia, contrast esophagography is performed while in children with respiratory symptoms bronchoscopy is used to rule out trauma or foreign body aspiration. However, evaluation by cross-sectional imaging for example CTA or MRA is mandatory in cases with complex anomalies, in older children and adults with inadequate or inconclusive echocardiographic findings.

In assessing cardiovascular morphology, MRA has the advantage due to lack of ionizing radiation, use of small volume of non-iodinated contrast and evaluation of flow patterns like flow changes, pressure gradients etc. On the other hand, CTA is fast and provides 3D reconstructed images for illustrating the complex anatomical relationship between the aortic arch structures, trachea and oesophagus that helps in surgical planning.

In our patient, after initial assessment CTA was planned as it was readily available and 3D images provided better understanding of vascular anatomy and its compression effects over the trachea and oesophagus. With the help of these imaging entities, KD can be diagnosed before lethal complications occur. Complications include aneurysm rupture, aortic dissection or recurrent pneumonia.

Backer et al suggested that any diverticulum more than 1.5 times the diameter of subclavian artery should be considered for surgical intervention due to the increase likelihood of rupture or tracheoesophageal compression. Surgical treatment options include open repair, hybrid endovascular and total endovascular repair. In a study done by Shinkawa et al, it was found that excision of KD with interposition of left subclavian to left common carotid artery and division of ligamentum arteriosum have good prognosis in eliminating residual symptoms and late complications.

**LEARNING POINTS**

1. Kommerell Diverticulum is a rare congenital anomaly which when occurs with right-sided aortic arch, aberrant left subclavian artery and ligamentum arteriosum forms a vascular ring.
2. Depending on severity, this vascular ring can cause symptoms of tracheal or oesophageal compression.
3. The role of imaging for evaluation of aortic arch anomalies and its associations is crucial for planning for surgery and endovascular intervention in symptomatic patients.
4. CT / MR angiography has gained more importance nowadays in non-invasive diagnosis of complex congenital anomalies.
5. If undiagnosed, lethal complications like aneurysm rupture and aortic dissection may occur.

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**REFERENCES**

1. Van Son JA, Konstantinov IE, Burckhard F. Kommerell and Kommerell’s diverticulum. Tex Heart Inst J 2002; 29: 109–12.
2. Robles TA, Srinivasan A, Mazur L, Gourishankar A. Kommerell’s Diverticulum with a Twist: A Case of Recurrent Wheeze in an 8-Year-Old Boy. Glob Pediatr Health 2019; 2019:2333794X19897506. Published.
3. Bhatt TC, Muralidharan CG, Singh G, Jain NK. Kommerell’s diverticulum: a rare aortic arch anomaly. Med J Armed Forces India 2016; 72(Suppl 1): S80–3. doi: https://doi.org/10.1016/j.mjaf.2016.09.003
4. Shuford WH, Sybers RG, Gordon JI, Baron MG, Carson GC. Circumflex retroesophageal right aortic arch simulating mediastinal tumor or dissecting aneurysm. American Journal of Roentgenology 1986; 146: 491–6. doi: https://doi.org/10.2214/ajr.146.3.491
5. Morishita A, Tomioka H, Katahira S, Hoshino T, Hanazawa K. Surgical treatment for Kommerell’s diverticulum associated with a right-sided aortic arch and an aberrant left subclavian artery: endovascular or hybrid. Ann Vasc Dis 2019; 12: 228–32. doi: https://doi.org/10.3400/avd.cr.18-00170
6. Berg C, Bender F, Soukup M, Geipel A, Axt-Fliedner R, Breuer J, et al. Right aortic arch detected in fetal life. Ultrasound Obstet Gynecol 2006; 28: 882–9. doi: https://doi.org/10.1002/uog.3883
7. Miranda JO, Callaghan N, Miller O, Simpson J, Sharland G. Right aortic arch diagnosed antenatally: associations and outcome in 98 fetuses. Heart 2014; 100: 54–9. doi: https://doi.org/10.1136/heartjnl-2013-304860
8. Cinà CS, Althani H, Pasenau J, Abouzahr L. Kommerell’s diverticulum and right-sided aortic arch: a cohort study and review of the literature. J Vasc Surg 2004; 39: 131–9. doi: https://doi.org/10.1016/j.jvs.2003.07.021
9. Hudgins PA, Siegel J, Jacobs I, Abramowsky C. The normal pediatric larynx on CT and Mr. AJNR Am J Neuroradiol 1997; 18: 239–45.
10. Jakanani GC, Adair W. Frequency of variations in aortic arch anatomy depicted
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11. Hanneman K, Newman B, Chan F. Congenital variants and anomalies of the aortic arch. Radiographics 2017; 37: 32–51. doi: https://doi.org/10.1148/rg.2017160033

12. Stojanovska APA, Jadranka MD, Cascade MS, Philip NMD, Chong SMD, Quint MS, et al. Sundaram, Baskaran MD. Embryology and imaging review of aortic arch anomalies. Journal of Thoracic Imaging 2012; 27(Issue 2): 73–84Volumep.

13. Backer CL, Mavroudis C. Congenital heart surgery Nomenclature and database project: vascular rings, tracheal stenosis, pectus excavatum. Ann Thorac Surg 2006; 69(suppl 4): 308–18. doi: https://doi.org/10.1016/S0003-4975(06)00179-5

14. van Son IA, Julsrud PR, Hagler DJ, Sim EK, Pairolero PC, Puga FJ, et al. Surgical treatment of vascular rings: the Mayo clinic experience. Mayo Clin Proc 1993; 68: 1056–63. doi: https://doi.org/10.1016/S0025-6196(12)60898-2

15. Bhat V, Al Kuwari M. Diverticulum of Kommerell: role of imaging revisited. Heart Views 2008;9:121–3 17.

16. Backer CL, Mavroudis C, Rigsby CK, Holinger LD. Trends in vascular ring surgery. J Thorac Cardiovasc Surg 2005; 129: 1339–47. doi: https://doi.org/10.1016/j.jtcvs.2004.10.044

17. Türkvatan A, Büyükbayraktar FG, Olçer T, Cumhur T. Congenital anomalies of the aortic arch: evaluation with the use of multidetector computed tomography. Korean J Radiol 2009; 10: 176–84. doi: https://doi.org/10.3348/kjr.2009.10.2.176

18. Backer CL, Russell HM, Wurlitzer KC, Rastatter JC, Rigsby CK. Primary resection of Kommerell diverticulum and left subclavian artery transfer. Ann Thorac Surg 2012; 94: 1612–7. Epub 2012 Aug 9PMID. doi: https://doi.org/10.1016/j.athoracsur.2012.05.101

19. Tanaka A, Milner R, Ota T. Kommerell’s diverticulum in the current era: a comprehensive review. Gen Thorac Cardiovasc Surg 2015; 63: 245–59. Epub 2015 Jan 3PMID. doi: https://doi.org/10.1007/s11748-015-0521-3

20. Shinkawa T, Greenberg SB, Jaquiss RDB, Imamura M. Primary translocation of aberrant left subclavian artery for children with symptomatic vascular ring. Ann Thorac Surg 2012; 93: 1262–5. doi: https://doi.org/10.1016/j.athoracsur.2011.12.030