Acute life-threatening episodes in an infant post-TOF repair

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DESCRIPTION

A 2-year-old girl presented with multiple acute life-threatening episodes (ALTEs) with cyanosis and apnoea, often occurring after feeds despite following a strict puréed diet. She had a complex neonatal course; born at 30 weeks' gestation with an antenatal diagnosis of Tetralogy of Fallot and neonatal diagnosis of tracheoesophageal fistula (TOF) and oesophageal atresia (OA)—repaired in early infancy.

Following her first ALTE at 6 months of age, bronchoscopy revealed mild tracheomalacia (TM) only, while CT angiogram (CTA) of the upper airway did not identify tracheal compression. This was performed with an endotracheal (ET) tube in situ (figure 1A).

Subsequent video fluoroscopy demonstrated pooling of contrast in the upper oesophagus, likely proximal to a degree of oesophageal narrowing and dysmotility. A diagnosis of anastomotic stricture (AS), the most common complication following OA/TOF repair, was considered in light of persistent failure to achieve feeding milestones complicated by enduring gastro-oesophageal reflux disease.[1] In light of this, she remained under multidisciplinary evaluation with ongoing AS screening for endoscopic dilation if symptoms did not resolve.

With progressive severity of symptoms in her second year of life, a repeat CTA, this time with a laryngeal mask in place, showed marked anterior tracheal compression by the innominate (brachiocephalic) artery just above the carina (figure 1B). The trachea was noted to be 'slit-like' with an accompanying saccular oesophagus (figure 1B). It became apparent that the puréed diet pooled in the oesophagus at this level, resulting in posterior tracheal compression, while the anomalous innominate artery pressed anteriorly on the trachea (figure 2). Nasogastric (NG) feeding was commenced prior to brachiocephalic arteriopexy and tracheopexy. At 3-month follow-up, she remains on NG feeds and is thriving with no further concerning episodes.

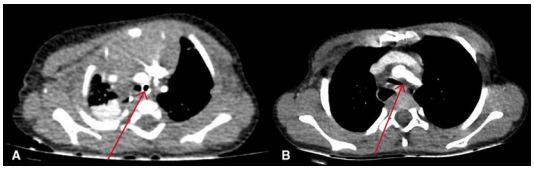

Figure 1 (A) Axial source CT image performed at 6 months of age at the level of the aortic great vessel origins. The tip of the endotracheal tube is seen within the trachea, immediately posterior to the brachiocephalic artery. Incidental note is made of consolidation in the apical segment of the right lower lobe. (B) Axial source CT image performed at 2 years of age at the level of the brachiocephalic artery. The trachea is flattened by the artery; it measured only 2 mm in the anteroposterior dimension.

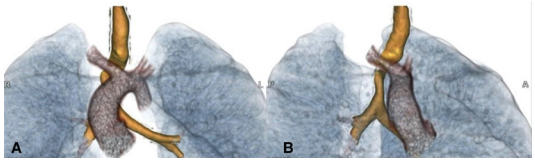

Figure 2 Three-dimensional airway reconstruction, anteroposterior and left lateral view, respectively, showing the trachea (beige) compressed by the brachiocephalic artery (purple), which passes immediately anterior to it. The aerated lung parenchyma is shown in blue.

Anterior tracheal compression by the anomalous innominate artery remains the most common vascular anomaly, first described in 1948.[2] Further mediastinal crowding was later considered, with posterior tracheal compression by the oesophagus in children post-OA/TOF repair identified.[3] Notably, reflex apnoea in association with feeding was deemed pathognomonic for this secondary phenomenon. It often presents with a variety of other respiratory symptoms including, noisy breathing, chronic cough, dyspnoea and recurrent respiratory infections.

Synchronous mediastinal lesions often coexist, in which case early recognition is of paramount importance.[4] A high index of suspicion for TM should be maintained in a child with a history of OA/TOF repair who presents with ALTE, the prevalence of which has been reported to range from 65% to 89%, with differences between neonates, young children and teenagers.[5 6] However, its clinical expression is variable with the degree of tracheal collapse relative to the complexity of mediastinal crowding.

The stenting effect of the ET tube on the initial CTA remains an important caveat and learning point from this case. It masked the presence of TM, which rendered the trachea particularly susceptible to luminal reduction induced by the innominate artery anteriorly and the oesophageal dilatation posteriorly, resulting in a delay in diagnosis.[7] Historically, urgent surgical correction to free the airway via arteriopexy was performed in children with reflex apnoea similar to our case. Suspension of the innominate artery to the sternum has proven to be a safe and effective means of symptom management.[8]

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Patient's perspective

For us, it has been one terrifying journey from the moment our daughter was born by emergency section at 30 weeks. We had known that she had a heart defect but we didn't know that her oesophagus hadn't completely formed and that would be the start of the complications.

She spent the first six months in hospital before going to England to have her heart surgery; which although it was a success still left some problems for us. She started having blue episodes where she would actually need CPR and this was the most terrifying thing—seeing our child lay there lifeless while a stranger or even at times myself had to work on her.

Every time she ended up in hospital, although she was well looked after, we kept getting a response that she had choked on something which we just knew that wasn't the case. Every day we lived on the edge not knowing if today was going to be a good day or if she would have another episode, and not having oxygen made it even more terrifying. Recently, it was spotted that she would need another operation and to our delight, she has not had any episodes since.

It has been the longest and most terrifying two and half years for myself, my wife and also our five kids. I must say the hospital staff at both hospitals have been our absolute heroes in making sure our baby is still with us today and we will be forever grateful.

Learning points

- Consider tracheal compression by the innominate artery anteriorly and dilated oesophagus posteriorly, in patients post-oesophageal atresia/tracheoesophageal fistula repair presenting with acute life-threatening episode, particularly in those with a history of post-prandial deterioration.
- Stenting by endotracheal tubes can mask tracheomalacia and tracheal compression warranting careful consideration of imaging modalities and the conditions under which they are performed.
- Urgent diagnostic evaluation and surgical referral must be considered in children with symptomatic tracheal compression.

Today, surgical indications have broadened to allow for subjective judgement based on diagnostic findings and correlation with symptom severity. Tracheal collapse greater than 75% is likely to be deemed significant and requires intervention. Traditionally, diagnostic evaluation included diagnostic laryngoscopy and bronchoscopy requiring anaesthesia. The introduction of less invasive modalities to include fibre-optic laryngoscopy as well as dynamic CT and MRI has offered comparable diagnostic information in paediatric populations.[9]

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