CASE REPORT

The finding of an undiagnosed long segment tracheal stenosis in a cyanotic child undergoing an arterial switch operation and VSD closure

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SUMMARY

This report details the case of a 2-month-old baby boy with known cyanotic congenital heart disease (double outlet right ventricle with subpulmonary ventricular septal defect, VSD) in whom tracheal stenosis was undetected, being found later on failed intubation while undergoing anaesthesia for an arterial switch operation and VSD closure. As a result, the cardiac surgery was postponed. Such an association between congenital heart disease and tracheal stenosis has been reported but remains exceptionally rare. The baby subsequently underwent a slide tracheoplasty the next day and had major cardiac surgery a few days later. He was discharged approximately 3 weeks later after spending 8 days on the cardiac intensive care unit.

BACKGROUND

This combination of pathologies is rarely seen and only a few centres exist worldwide that can deal with such conditions. Advances in the treatment of both pathologies mean that the patient was successfully operated on and discharged in just 3 weeks. The anaesthetic management of the unidentified tracheal stenosis in a cyanotic child combined with the availability of expert surgical opinion has so far ensured a good outcome for this child. This case details the management of an unusual pathology that was rare and potentially disastrous for the baby concerned.

CASE PRESENTATION

We report the case of a full-term 2-month-old baby boy diagnosed antenatally with double outlet right ventricle with subpulmonary ventricular septal defect (VSD), with oxygen saturations of 82–94% on air. The patient was due to have an arterial switch procedure and VSD closure at Great Ormond Street Children's Hospital in April 2015. He had no history of stridor, wheeze, cough, difficulty in breathing or cyanotic episodes. After induction of anaesthesia via gas induction, it was noted that a 3.5 and 3.0 mm endotracheal tube (ET) could not be passed more than 1 cm through the vocal cords. No force was applied and further attempts at intubation were abandoned, leaving the ET tube placed only partly through the vocal cords. The baby was transferred into theatre where microlaryngoscopy and bronchoscopy were performed. This demonstrated a long segment tracheal stenosis. Cardiac surgery at this point was postponed and the baby remained intubated with the

ET tube just beyond the cords; gentle ventilation was continued and the patient transferred to the cardiac intensive care unit intubated and ventilated.

INVESTIGATIONS

After the initial bronchoscopy, the patient was transferred to interventional radiology for a full bronchogram, optical coherence tomography and repeat bronchoscopy (figure 1), the aim being to understand fully the anatomy and suitability for tracheal surgery. It was noted that the trachea measured 2.6 mm at its narrowest and 5.2 mm proximal to the carina; the length of the stenosis can be seen in figure 2.

DIFFERENTIAL DIAGNOSIS

Inability to pass the ET could have been caused by previous trauma or infection to the trachea, tumours such as chondromas or extrinsic compression by thyroid neoplasms. Compression via an anomalous subclavian artery has also been reported.

TREATMENT

It was decided to prioritise tracheal surgery over cardiac surgery, as the ET could not be safely secured; therefore preceding with cardiac surgery may have led to loss of the airway, potentially disastrous in a cyanotic child.

In this case, the tracheal stenosis affected over two-thirds of the trachea (so called long segment), therefore it was deemed to require sliding

Figure 1 A view from the bronchogram showing the tracheal stenosis.

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Figure 2 A reconstructed image from CT.

tracheoplasty surgery. This procedure was successfully carried out the next day under cardiopulmonary bypass. The baby's chest was left open between procedures, and he was transferred to the cardiac intensive care unit intubated and ventilated with inspiratory pressures of 13 cm H₂O and a positive end-expiratory pressure of 5 cm H₂O, requiring 40% inspired oxygen. Two days later, he was returned for an arterial switch procedure and VSD closure. Postprocedure, the child required epicardial pacing for 2 days and was successfully extubated on day 5.

At Great Ormond Street (a quaternary referral centre), we see around 8–10 children with long segment tracheal stenosis per year. This procedure is often carried out in the emergency setting, as children present with respiratory distress, usually following an upper respiratory infection. However, as this case demonstrates, diagnosis can sometimes be made only on induction of anaesthesia and failure to intubate. In short, the tracheal and cardiac surgery often both take place at the same time but in this case, the two procedures were not carried out simultaneously. This was due to the complex nature of the cardiac surgery, the length of the procedures and increased risk of a combined procedure.

OUTCOME AND FOLLOW-UP

Slide tracheoplasty is a relatively new procedure, having only been in use for the last 20 years. Postsurgery, children should be able to have a near normal life, attending school or nursery, but the initiation of feeding can be problematic and often dietetic input is required. The prognosis for children with combined cardiac and tracheal pathology is less clear, with so few patients presenting with this combination of problems. This child was successfully discharged home after 3 weeks in hospital, and to date continues to do well. A routine bronchoscopy on follow-up showed that the trachea was healing well and no further dilation was required. He will remain under review by the tracheal and cardiac teams for the foreseeable future.

With advances in tracheal surgery the outlook for such patients is much improved.

DISCUSSION

Combined tracheal and cardiac pathology remains exceptionally rare; however, an associated link has been described. A paper by Mainwaring *et al*¹ describes the cases of 27 patients with tracheal stenosis from 2003 to 2011, 21 of whom had combined cardiac and tracheal pathology. With two postoperative deaths

in children with single ventricles, the rest of the cohort did not require repeat surgery and the median duration of follow-up was 4 years. In a study by Wijeweera *et al*,² 11 children with congenital long segment stenosis from 1999 to 2008 were identified; 90.9% had an associated cardiac anomaly. A further paper by Loukanov *et al*³ describes the simultaneous management of tracheal stenosis and cardiac anomalies in nine infants, with one postoperative death; the rest of the group showed a stable and complication-free recovery. It is hoped that this report will add to the small body of evidence showing the successful management of the combination of cardiac and tracheal pathologies, a combination that, until recently, would have been regarded as non-survivable.

Learning points

- ▶ Cardiac anomalies increase the risk of tracheal stenosis.
- ▶ Unidentified tracheal stenosis may lead to being unable to ventilate and/or intubate on induction of anaesthesia. Ventilation of such children under anaesthesia remains the priority as intubation may be impossible; repeated attempts at intubation are best avoided.
- ▶ Owing to surgical advances, this combination of pathologies can now be successfully managed at a few specialist centres around the world.

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Competing interests None declared.

Patient consent Obtained.

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