Unsuspected concentric tracheal rings in a 14-year-old with scoliosis

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A 14-yr-old boy was first found to have tracheal stenosis when anaesthesia was induced for extensive scoliosis surgery in the prone position. There are no guidelines for airway management under these conditions. We describe how we managed the problem and suggest some useful modifications.

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A 14-yr-old boy was scheduled for correction of scoliosis. He was obese, with a large head, coarse hair, short neck and a familial short stature (weight 59 kg, height 125 cm). His upper to lower body segment ratio was high and he demonstrated limb shortening.

As a young child, the patient had had repeated episodes of bronchitis and asthma, but had been symptom-free without respiratory problems or shortness of breath on exertion since 9 yr of age. On preoperative examination, there was no wheezing or stridor, breath sounds were normal and the lungs were clear. Pulmonary function tests and echocardiography were normal. There was no history of previous anaesthetic or surgical intervention.

Anaesthesia was induced uneventfully with a mask and circle system using a mixture of oxygen 30% and nitrous oxide 70%, with sevo-urane increasing to 8%. Once anaesthesia had been established, sevo-urane was replaced with iso-urane 1.5%. Intravenous access was established, and fentanyl 100 µg and rocuronium 60 mg were administered. Tracheal intubation was undertaken under direct laryngoscopy using a number 3 Macintosh blade and a 6.0 mm cuffed Portex tube. The tracheal tube was passed easily through apparently normal vocal cords, but met resistance 2–3 cm below. Neither a 5.5 mm nor a 5.0 mm cuffed or uncuffed tracheal tube could be inserted past the obstruction. There was, however, no difficulty with mask ventilation between intubation attempts and breath sounds were normal. Peak airway pressure reached 40 cm H₂O when ventilation was performed through a 5.0 mm uncuffed tracheal tube wedged in the trachea. At this time, arterial blood gases with a \( F_{\text{IO}_2} \) of 1.0 showed pH 7.24, \( P_{\text{aCO}_2} \) 7.5 kPa, and \( P_{\text{aO}_2} \) 34.3 kPa. A diagnosis of tracheal stenosis of unknown aetiology was made and an otorhinolaryngologist was consulted in the operating room. Rigid and flexible bronchoscopies were performed. A 2 cm long stenosis of the midsegment of the trachea was noted, suggestive of concentric tracheal rings. The carina and bronchi were normal. It was decided to let the patient emerge from anaesthesia, re-evaluate the tracheal stenosis and devise a plan for subsequent management. Recovery from anaesthesia was uneventful.

Magnification x-ray views of the airway were obtained, which confirmed a 2 cm stenotic area in the midsegment of the trachea (Fig. 1). A CT scan of the chest showed a narrowing (3.5 mm width) of the trachea from the level of the clavicular heads to approximately 2 cm above the carina (Fig. 2A and B). Both the magnification views and the CT scan were reported as consistent with complete tracheal rings. There were no bronchial abnormalities.

The medical staff did not think that this asymptomatic patient needed surgical correction of his tracheal stenosis at the present time, but the orthopaedic surgeon felt that his rapidly progressive scoliosis required early spinal stabilization. A lengthy surgical procedure was planned. Hence, we decided upon a trial of anaesthesia without surgery. Anaesthesia was induced with a mask and circle system using a mixture of oxygen 30% and nitrous oxide 70%, with sevoflurane increasing to 8%. Once anaesthesia had been established, sevoflurane was replaced with isoflurane 0.5%. Intravenous access was established, and fentanyl 100 µg and rocuronium 60 mg were administered. Under fibre-optic vision, a 5.5 mm cuffed tracheal tube was inserted through the vocal cords, with its tip positioned just above the obstruction and the cuff inflated to seal the airway. The
patient was ventilated in the prone position for 3 h, with a peak airway pressure stable at 30 cm H$_2$O. End-tidal carbon dioxide tension was 4.7±5.3 kPa, and arterial oxygen saturation by pulse oximetry was 95–99%. Vital signs remained stable. The patient made an uneventful recovery and was discharged home.

Three weeks later, the patient was readmitted for the definitive procedure. Anaesthesia was provided in the same manner as in the trial. In addition, the patient received i.v. fentanyl 2 μg kg$^{-1}$ h$^{-1}$ and sufficient vecuronium to maintain two muscle twitches in a train-of-four for the duration of surgery, which lasted 7 h. Expired carbon dioxide tension varied between 4.4 and 4.8 kPa and the arterial oxygen saturation between 95 and 100%. Intermittent arterial blood gases remained within normal limits. At the conclusion of surgery, the child emerged from anaesthesia, was extubated and breathed spontaneously without difficulty. His subsequent recovery was uneventful.

Discussion

Congenital tracheal stenosis is rare. Benjamin and colleagues$^1$ reported 21 cases, all in infants, and classified congenital tracheal stenosis into three types: (i) diffuse narrowing of the trachea from just below the cricoid cartilage to the carina; (ii) funnel-like stenosis that may occur anywhere along the trachea and involve varying lengths; and (iii) a short segmental length of tracheal stenosis, usually in the middle or lower third. Concentric tracheal rings characterize many of these lesions.

Chen and Holinger have proposed that formation of complete or near complete tracheal rings arises from disproportionate growth of the cartilage relative to the posterior tracheal pars membrane.$^2$ Voland and colleagues have suggested an intrinsic field defect in the cervical splanchnic mesenchyme for the formation of concentric tracheal rings.$^3$ Because of this association, concentric tracheal rings are seen in patients with cervical chondrogenic anomalies, a foreshortened neck and trachea, pulmonary agenesis and abnormal vasculature. Our patient showed some evidence of a defect in the cervical splanchnic mesenchyme.

The incidence of concentric tracheal ring stenosis in older children or adults has not been determined. We found only four reported cases of anaesthetic management of patients with concentric tracheal rings,$^4$ and only one of these patients was managed in the prone position.$^7$ Goddard and Hughes$^7$ describe a failed intubation in a 16-yr-old child who presented for surgical correction of idiopathic scoliosis. The narrow segment began at the level of the cricoid and extended to just above the carina. The authors were able to wedge a 5.0 mm tracheal tube into the narrow segment of the trachea. However, there was no report of peak airway pressures, end-expired carbon dioxide tension or blood gas analysis, and the patient developed postoperative bronchospasm which required i.v. aminophylline therapy. Ours is a second such patient, but the two are not truly comparable. There are similarities, namely, scoliosis, age, history of repeated childhood respiratory infection, and eventual successful outcome after surgical repair, but there are also dissimilarities between the two cases. Our patient...
had a 2 cm midsegment tracheal ring stenosis, whereas Goddard and Hughes report that their patient’s tracheal ring stenosis extended from just below the vocal cords down to the carina. Our patient had dysmorphic features not present in the other. Pulmonary function testing also differed. Their patient had signs of fixed extrathoracic airway obstruction, which was partially obscured by mild asthma, whereas pulmonary function tests in our patient demonstrated normal flow-volume loops and FEV<sub>1</sub> 98%, FEV<sub>1</sub>/FVC 86%, PEFR 93%, PEF<sub>25-75</sub> 83%, VC 101% and IC 107% of predicted values. Hence there was no evidence of fixed extrathoracic airway obstruction.

Perhaps we were overzealous in our endeavour to be cautious by undertaking a trial of anaesthesia. We feared that airway instrumentation and the trauma of a tracheal tube rubbing against or wedging into a stenotic tracheal orifice might induce catastrophic airway obstruction. One could argue that our trial was not even a true one, for it lacked the operative manipulation of the vertebral column and chest.

We now favour induction of anaesthesia and placement of the tracheal tube as described above, followed by turning the patient into the prone position. If respiratory mechanics and blood gases remain stable, definitive surgery should commence, thus obviating the need for a separate trial of anaesthesia.

Even though undiagnosed concentric tracheal ring stenosis is a rare finding, there are diagnostic clues which, if pursued, may obviate surgical delay or postponement. Though we know of no relationship between scoliosis and concentric tracheal ring stenosis, there are now two such reported cases. We propose that if a patient has a history of repeated respiratory infections in childhood and exhibits dysmorphic features or has even mildly abnormal pulmonary function tests, then preoperative evaluation of such patients should include magnification x-ray views of the upper airway followed by CT scan and bronchoscopy if indicated.

References