Airway surgery in children under extracorporeal circulatory support

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Contributions: (I) Conception and design: All authors; (II) Administrative support: None; (III) Provision of study materials or patients: None; (IV) Collection and assembly of data: None; (V) Data analysis and interpretation: None; (VI) Manuscript writing: All authors; (VII) Final approval of manuscript: All authors.

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Abstract: Airway surgery can be challenging in children. With increasing awareness of congenital lesions, expertise in diagnostic modalities and recent advances in surgical technique, complex airway malformations are increasingly better managed. Physiological and anatomical differences, combined with specific features of surgical problems seen in childhood, necessitate a different approach to perioperative care compared with adults. An often essential component for efficient and complete surgical correction is the appropriate utilisation of extracorporeal circulatory support. This article details the key differences between the paediatric and adult patient with respect to airway surgery, before discussing the role and applications of extracorporeal circulatory support in various airway reconstructions in children. We discuss the potential alternate options including use of extra-corporeal membrane oxygenation versus standard cardiopulmonary bypass (CPB) during surgery.

Keywords: Airway surgery; cardiopulmonary bypass (CPB); tracheal reconstruction

Received: 12 January 2021. Accepted: 12 March 2021.
doi: 10.21037/ccts-21-9
View this article at: http://dx.doi.org/10.21037/ccts-21-9

Introduction

Airway surgery in paediatric patients presents distinct challenges compared with the adult population due to significant differences in cardiorespiratory physiology and anatomy. These differences also predispose neonates and infants to an increased risk of perioperative complications. The cardiovascular and respiratory systems undergo extensive changes from foetal life to later childhood; these changes, combined with the technical challenges of surgery, render younger patients more vulnerable to cardiorespiratory insufficiency during airway surgery. This disparity between adult and infant patients means that extracorporeal circulatory support is rarely required in adult airway surgery; however, in paediatric patients it has become an essential component of safe and effective perioperative management.

Unique characteristics of paediatric airway physiology and anatomy

Several notable characteristics of the paediatric respiratory system reduce functional reserve and predispose these patients to respiratory decompensation and collapse in the perioperative period. In terms of physiology these include: immature ventilatory control; inefficient respiratory muscles; predisposition to small airway closure (due to reduced functional residual capacity and increased closing volume); and higher oxygen consumption due to the metabolic requirements of growth and temperature homeostasis (1). The use of increased inspired oxygen concentrations to mitigate these issues can potentially depress ventilatory effort and precipitate retinopathy and/or bronchopulmonary dysplasia. Neonates exhibit a blunted response to hypercapnia and therefore are less able to mitigate against type 2 respiratory failure.

Regarding anatomical considerations, the trachea in infants is short and narrow, with immature and unstable cartilages. Endobronchial displacement, accidental extubation of the tracheal tube or obstruction to ventilation can therefore occur easily during surgical manipulation. The smaller cross-sectional area of the airway produces higher resistance to airflow (2) and any further reduction in diameter (e.g., mucosal oedema, stenosis of any part of airways including larynx or trachea, presence of blood clots or secretions and/or placement of an inappropriately small tracheal tube) is extremely poorly tolerated.

The accessory respiratory muscles are less effective and immature in younger children. The skeletal configuration of infants, with horizontally placed ribs, limiting the inspiratory increase in cross-sectional area of the thorax seen in adults (3).

Unique characteristics of paediatric cardiovascular physiology:

The transitional circulation, which persists for days to several weeks, can be extremely vulnerable to changes in pulmonary vascular resistance. Rises in pulmonary vascular resistance due to hypoxia, hypothermia, hypercarbia and acidosis may reopen the ductus arteriosus and foramen ovale and may regress the transitional circulation to a persistent foetal circulation (4). The neonatal myocardium operates at near maximal contractility because of a relatively high concentration of endogenous catecholamines and thyroid hormones released during late gestation and at birth (5,6). The immature, poorly compliant neonatal myocardium has a lower capacity to increase cardiac output in the event of an increase in preload (7). Hence the newborn's heart is less able to compensate for the significant changes in loading conditions that can occur during anaesthesia and surgery (8). Depression of myocardial contractility caused by anaesthetic agents or other physiological derangement (e.g., hypoxia, hypovolaemia, arrhythmia, acidosis and electrolyte imbalance) may cause sudden catastrophic circulatory collapse (9,10).

Performing airway surgery in paediatric patients with these physiological characteristics is therefore a challenging task, and would be impossible in many cases without the use of some form of extra-corporeal circulatory support. This support can be achieved by either conventional full cardiopulmonary bypass (CPB) or extracorporeal membrane oxygenation (ECMO). What follows is an overview of the application of these techniques in our practice.

CPB is widely used in most of our tracheal surgeries. Most common surgeries include slide tracheoplasty for long segment tracheal stenosis and resection of trachea with end-to-end anastomosis for shorter segmental lesions. Other surgery carried out under CPB include reconstruction of the trachea and/or bronchi including the carina for various tumours, repair following failed or complex tracheo-esophageal fistula repair, and post-traumatic tracheal and bronchial reconstruction (e.g., following blunt-force trauma to the chest or following button battery ingestion).

Role of CPB in long segment congenital tracheal stenosis (LSCTS)

Management of LSCTS caused by the presence of complete cartilaginous tracheal rings has been revolutionised by the advent of the slide tracheoplasty technique (11). Since then, increasing numbers of slide tracheoplasties are being performed worldwide for this condition in children under CPB.

Clinical presentation with LSCTS in small babies is largely determined by the severity of airway stenosis. The majority of these children present with some form of respiratory distress; in our experience, at least 12% present with acute respiratory failure needing extra-corporeal support in the form of ECMO. An additional 20% of them will need invasive ventilation before arriving at a specialised centre for surgical repair. With this severity of disease, it is natural to expect these unstable children to require CPB for further surgery.

Quite often tracheal intubation involves use of an appropriately sized tube at, or just below the vocal cords, to facilitate maintenance of ventilation while these babies are transported to a surgical centre. CPB facilitates changing these tubes to appropriate sized tube at correct position as well as managing them after surgery in a stable manner.

In addition to pre-operative factors influencing the use of CPB, there are other intraoperative considerations that make the use of CPB beneficial or essential. One key factor is that, particularly with LSCTS, the stenosis is often too severe to pass a tracheal tube through. In adult tracheal surgery, it is often possible to maintain a route for ventilation by intubating the distal trachea or a main bronchus; in severe LSCTS this is impossible, and extracorporeal circulation becomes the only way to maintain gas exchange during surgery.

Another factor is the length of the stenotic segment, including extension of complete rings to either of the bronchi. At surgery, extensive dissection is often required for adequate mobilisation of the airways. This includes release of subcarinal and paratracheal tissues; this enhanced mobility is essential for a secure and tension-free anastomosis, while also allowing vascularity to be maintained. Adequate gas exchange needs to be maintained during these mobilisation maneuvers, and this would often be extremely difficult or impossible to achieve using standard ventilation. Use of CPB enables us to dissect, loop and retract the ascending aorta for the carinal and left main bronchus dissection, and the same for the right pulmonary artery and superior vena cava for right main bronchus dissection. While on CPB the tracheal tube may be removed and changed to an appropriately sized tube without any oxygenation compromise. At the end of reconstruction, this chosen tube is positioned at the correct depth under direct vision for postoperative care.

Finally, associated lesions are very common in this situation, in particular the left pulmonary artery sling. Correction of these lesions as well as intracardiac defects is facilitated greatly by the use of CPB (12).

Role of CPB in complex tracheobronchial reconstructions following tracheo-esophageal fistula repair

Complex airway pathologies are not uncommon in children. These include surgical failure following previous repair of tracheo-esophageal fistula and some form of injury to the airway following button battery ingestion; the latter can cause rapid and severe damage to the oesophagus and tracheobronchial tree.

Management of the complication of post-oesophagectomy tracheo-bronchial-oesophageal fistula (PETEF) complication is uncertain and not standardised. The location of the oesophagogastric anastomosis next to the membranous wall of the trachea and mainstem bronchi and intimate relationship of the airway and proximal oesophagus predispose the airway to injury during an esophagectomy (13).

From various reported case series, fistulae with small defects and minimal mediastinal adhesions due to mediastinitis are accessed through cervical or cervicomediastinal approaches or posterolateral right thoracotomy. More extensive defects, following multiple reoperations, defects with frozen anatomy and complex abnormal vascular anatomy remain challenging to repair, and the optimal conditions provided by CPB via sternotomy may be required (14). In cases with complex abnormal vascular anatomy, use of 3D reconstruction of the images facilitates planning of the surgical approach. Under CPB, extremely vascular adhesions can be dealt with confidently, and the true extent of the defect safely defined. Estimating the true extent of the defect after extensive dissection helps in preventing failure of the repair requiring subsequent re-intervention. Simultaneously such extensive defects can be repaired with autologous pedicled pericardial patch.

Role of ECMO and CPB in tracheal and bronchial reconstruction after injury due to button battery ingestion and chest trauma

A significant proportion of children referred to our unit (under the UK national tracheal service for complex airway surgery) present with complex airway injuries. ECMO is a valuable tool in stabilising such patients initially, as these patients often arrive in extremely fragile condition with a decompensated cardiorespiratory status. The choice of ECMO can be either veno-veno or veno-arterial depending on the clinical picture. Once stabilised, these children undergo an array of investigations on the airway, including contrast-enhanced computed tomography (CT) scan and bronchoscopy and bronchogram. Optical coherence tomography (OCT) in particular, is very valuable in showing the contrast quality of the tracheobronchial wall.

Children with unstable airways can potentially be at further risk at induction of anaesthesia (assuming ECMO has not been instituted preoperatively). Tracheal intubation can result in further trauma to the defect, and positive pressure ventilation can cause life-threatening tension pneumomediastinum. One strategy for managing this situation is to maintain spontaneous ventilation via a supra-glottic airway device (SAD), such as a laryngeal mask airway (LMA). This can be used to maintain oxygenation until sternotomy is completed and CPB instituted (gentle positive pressure will be needed once the chest is open). This strategy clearly requires close communication and collaboration between surgeon and anaesthetist at all times.

In situations of caustic injury to the airways, additional debridement of the airway becomes an important step to prevent ongoing necrosis of the tissues while reconstruction (often using an autologous pericardial patch) is attempted. Use of CPB can further facilitate this step, in addition to repair of the esophagus at the same time via an anterior approach.

Role of CPB in tracheal and carinal tumour resection

Tumours, including both malignant and benign lesions can need further extra-corporeal support for surgery. As described above, tracheal tumours that extend beyond a short segment may need complex surgical reconstruction of the airway. Carinal tumours, especially carcinoid and proximal bronchial adenoids, where there are no resection margins to the carina for complete clearance, usually require CPB (15). Inflammatory myofibroblastic tumours are the commonest childhood airway tumour in our experience. Resection is by wide local excision with clear surgical margins. In such situations, the use of CPB can facilitate complete surgical cure in these children (16-18).

A comparison of the merits of CPB versus ECMO

Although CPB and ECMO have made complex airway lesions surgically correctable, there are significant deleterious effects to consider. Contact with the extracorporeal circuit activates an inflammatory response which, in combination with particulate emboli, multiple gaseous and/or lipoproteins generated due to disruption of the glycocalyx during CPB may all contribute to postoperative multiorgan dysfunction (19). The systemic inflammatory response is produced by proinflammatory mediators, activated leucocytes, vascular endothelial cells, and platelets. Coagulopathy caused due to factor depletion, use of heparin and haemodilution during CPB results in coagulopathy, postoperative bleeding requiring surgical re-operation, and increased transfusion requirements (20).

Several prospective studies have demonstrated a higher incidence of circulating TNF-α, (21,22). TNF-α is also a critical factor in initiating the cytokine cascade responsible for tissue ICAM-1 induction and subsequent neutrophil sequestration (23). In patients with comorbid conditions that are associated with postoperative renal dysfunction, CPB is associated with an increased risk of dialysis-dependent renal failure (24).

CPB is associated with significant pulmonary cytokine production (IL-8), neutrophil sequestration, and alveolar macrophage activation in clinical studies (25). This results in significant increases in static and dynamic compliance of the respiratory system and lung and work of breathing (26). The incidence of overt stroke, following cardiac surgery, ranges from 1% to 6% while the incidence of neurocognitive dysfunction is as high as 60% at 1 week with reduction to 25% to 30% at 8 weeks and 12 months (27).

Several technical innovations have resulted in less intraoperative hemolysis, blood activation, and the circulation of emboli. These include: biocompatible plastics; in-line filters; membrane oxygenators (*vs.* bubble oxygenators); and centrifugal blood pumps (*vs.* roller pumps) (28). Surface modification of circuits with heparin to mimic the native circulation and the use antifibrinolytics (e.g., aprotinin, tranexamic acid) have been shown to attenuate blood component activation, systemic inflammation, and organ dysfunction (29,30). Even with these developments, post-CPB inflammation and organ dysfunction are attenuated but not abolished (31).

There is some evidence that ECMO confers some advantages over CPB with regard to the inflammatory response. Firstly, the blood-air interface present in the venous reservoir in CPB is thought to be particularly important in activating the systemic inflammatory response. This is not present in the ECMO circuit. Similarly, haemodilution, which is known to contribute to coagulopathy and inflammatory activation, is less likely to occur during ECMO than CPB. Finally, the necessary use of protamine at the termination of CPB produces protamine-heparin complexes that are known to worsen the inflammatory response via complement activation (32). Despite these advantages of ECMO over CPB, there are several critical technical differences that make CPB a better strategy than ECMO during airway surgery. These are detailed in *Table 1*.

Conclusions

Complex airway surgery has evolved well beyond routine sliding tracheoplasty in children. Use of extra-corporeal support have greatly facilitated procedures, both in stabilising these children pre-operatively (using ECMO) and conducting safe and effective surgical repair (using CPB).

Table 1 Key differences between cardiopulmonary bypass as against ECMO

Factors	CPB	ECMO
Capillary pressures	Low	High
Circulation	Decompressed ventricles, making surgical manipulation easier	Circulation is often full – with less effective decompression. Needs additional Vent
Cannulae	Can be easier to manage – including introduction, mobilisation at surgery and control of flows	Can be very temperamental
Strategy	Dynamic bypass strategy with intermittent low flow can be made safe with systemic cooling	Lack of rapid systemic cooling means that safe dynamic low flow strategies are not possible
Bleeding during procedure	Retained under CPB using cardiotomy suckers	Lost from circulation

CPB, cardiopulmonary bypass; ECMO, extracorporeal membrane oxygenation.

Acknowledgments

Funding: None.

Footnote

Provenance and Peer Review: This article was commissioned by the Guest Editors (Andrea Dell'Amore and Nizar Asadi) for the series "Mechanical Extracorporeal Cardio-Respiratory Supports in General Thoracic Surgery" published in *Current Challenges in Thoracic Surgery*. The article has undergone external peer review.

Conflict of Interest: The authors have completed the ICMJE uniform disclosure form (available at http://dx.doi.org/10.21037/ccts-21-9). The series "Mechanical Extracorporeal Cardio-Respiratory Supports in General Thoracic Surgery" was commissioned by the editorial office without any funding or sponsorship. Dr. Muthialu serves as an unpaid editorial board member of *Current Challenges in Thoracic Surgery* from Sep 2019 to Aug 2021. The authors have no other conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

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doi: 10.21037/ccts-21-9
Cite this article as: Beeman A, Ramaswamy M, Robertson A, Ip J, Muthialu N. Airway surgery in children under extracorporeal circulatory support. Curr Chall Thorac Surg 2021.

