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CONGENITAL TRACHEO-OESOPHAGEAL FISTULA

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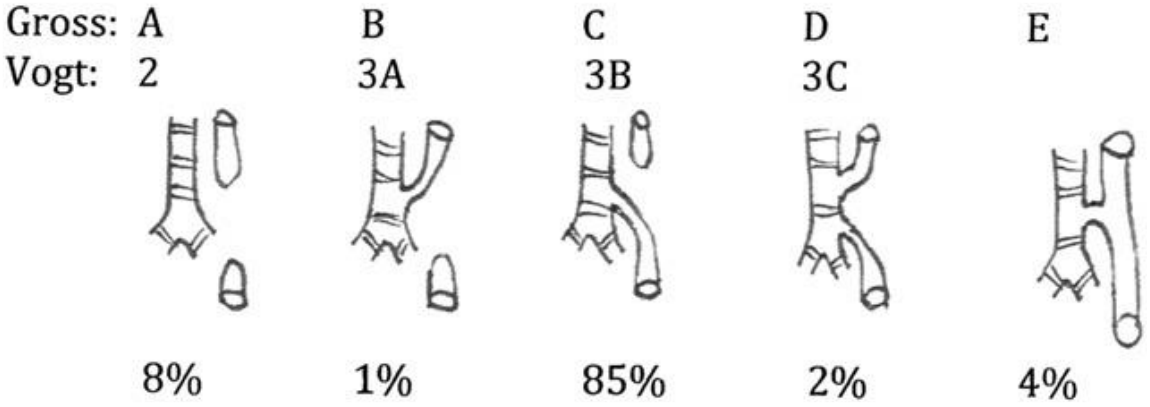
CONGENITAL TRACHEO-OESOPHAGEAL FISTULA

INTRODUCTION

Tracheo-oesophageal fistula (TOF) is a congenital abnormality, occurring in 1:3000-4500 neonates. Prior to the first successful TOF repair in 1939, mortality was high in this group of neonates. Survival rates have improved due to recent advances in anaesthesia, surgery and neonatology, however mortality and morbidity is still considered to be high in our setting.

Important to the anaesthetist with regard to these patients is the prevention of aspiration and dehydration, as well as overcoming difficulties with intubation and mechanical ventilation. These difficulties include inadvertent fistula intubation, ventilation of the stomach, atelectasis, resultant hypoxemia and shunt. It is also likely that these patients will present later in childhood, for correction of complications associated with fistula repair.



The Gross and Vogt Classifications of TOF

EMBRYOLOGY

TOF arises from failure of the primitive foregut to completely separate into the trachea and oesophagus. There are 5 variants according to the Vogt and Gross Classification (1):

- Type A (8%): isolated oesophageal atresia (OA) and no fistula formation.
- Type B (2%): OA with proximal fistula formation.
- Type C (80-85%): the most common malformation, with OA and distal fistula formation.
- Type D (1%): OA with proximal and distal fistulae.
- Type E (or H type) (4%): no atresia with isolated TOF in an H-formation.

Fistulae are most commonly located:

- >1cm above the carina (67%)
- <1cm above the carina (22%)
- At the level of the carina (11%).

ASSOCIATED ANOMALIES

50% of patients with TOF have other associated anomalies, especially midline/VACTERL (*vertebral, anorectal, cardiac, TOF/OA, radial, renal and limb*) abnormalities.

Congenital abnormalities associated with OA/TOF (2)

Organ System	Incidence (%)	Examples
Cardiac	29	VSD, PDA, tetralogy of Fallot, ASD, right-sided aortic arch
Gastrointestinal	14	Duodenal atresia, imperforate anus, malrotation, pyloric stenosis, omphalocoele
Musculoskeletal	10	Radial limb abnormalities, polydactyly, lower limb defects, hemivertebrae, rib defects, Scoliosis
Genitourinary	14	Renal agenesis, hypospadius, horseshoe/polycystic kidney, ureteric/urethral abnormalities
VATER syndrome (VACTERL)	10	Vertebral, anorectal, tracheoesophageal, renal or radial anomalies (expanded to include cardiac and limb defects) Other associated syndromes include: CHARGE (coloboma, heart defects, atresia choanal, retarded growth and development, genital hypoplasia and ear deformities), Potter's (pulmonary hypoplasia, bilateral renal agenesis, characteristic facies of intrauterine compression), SCHISIS (omphalocoele, cleft lip and/or palate, genital hypoplasia)
Respiratory	6	Tracheo-bronchomalacia, pulmonary hypoplasia, tracheal agenesis/stenosis, tracheal upper pouch
Genetic	4	Trisomy 21, Trisomy 18, 13q deletion

FACTORS AFFECTING MORTALITY AND OUTCOME

These include:

1. Prematurity
2. Presence of congenital heart disease
3. Position and size of the fistula
4. Pre-existing lung pathology
5. Delayed diagnosis

Prematurity and congenital cardiac disease have been shown to be independent predictors of peri-operative morbidity and mortality (1). 20-30% of neonates with TOF will be preterm with low birth weights. 30% of neonates will have congenital cardiac disease, most commonly Ventricular Septal Defect and Tetralogy of Fallot. Outcome in patients with cardiac anomalies including patent ductus arteriosus (PDA), is equivalent in ductal-dependent and -independent groups.

Large, peri-carinal fistulae have been associated with worse peri-operative outcome. Poor lung compliance due to chronic lung disease, respiratory distress syndrome and aspiration, pose a challenge to ventilation in these already compromised patients. Delayed diagnosis increases the risk for aspiration and complications thereof (2). The first prognostic classification was by Waterstone in the 1960's, where he found weight, pneumonia and associated congenital anomalies to be determinants of outcome.

Spitz then found in the 1990's that congenital anomalies affecting prognosis were cyanotic heart disease requiring palliative or corrective surgery, or noncyanotic heart disease requiring medical or surgical management. Pneumonia was removed from the classification. In 2009, Okamoto used statistical methods and ROC curves of birth weight to extrapolate **THAT** the 'at-risk' weight group **WAS** <2 kg. Babies with <2kg birth weight with congenital heart disease has a survival rate of 27%, whereas neonates weighing >2kg without cardiac anomalies have survival rates approaching 100%.

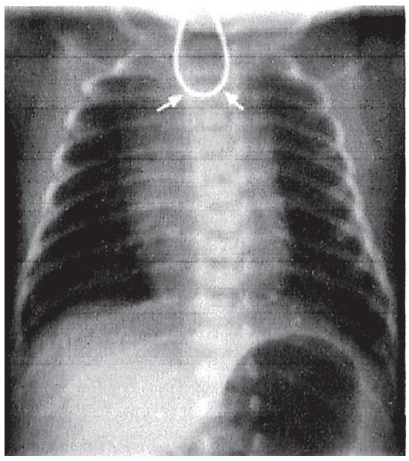
These classifications have also been used for timing of surgery and resuscitation, and whether surgery can be delayed or staged. (2)

Table 2: Prognostic classification of OA/TOF by Waterston, Spitz, Okamoto

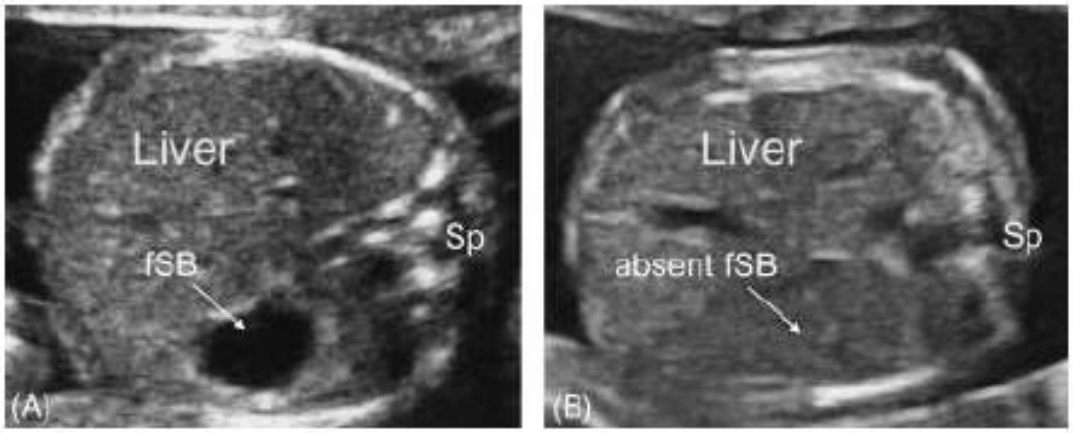
Prognostic Scheme	Class	Birth Weight	Anomaly	Survival Rate (%)
Waterston Classification 1962	A	>2.5 kg	Nil	95
	B	>2.5kg	Lobar pneumonia mild – moderate congenital anomaly	68
	C	1.8-2.5kg	Severe pneumonia Severe congenital anomaly, or multiple moderate anomalies	
		1.8-2.5kg	Nil	6
		<1.8 kg		
Spitz Classification 1994	I	>1.5 kg	Nil	97
	II	>1.5 kg	Major cardiac anomaly	59
		<1.5 kg	Nil	
	III	<1.5 kg	Major cardiac anomaly	22
Okamoto Classification 2009	I	>2kg	Nil	100
	II	<2kg	Nil	81
	III	>2kg	Major cardiac anomaly	72
	IV	<2kg	Major cardiac anomaly	27

DIAGNOSIS

TOF has been associated with non-specific prenatal signs such as polyhydramnios as well as reduced filling or absence of the stomach on ultrasound (3); however the positive predictive value of these findings is only 44% (2). The diagnosis is usually made at birth or within the first few days of life. Signs include choking and respiratory distress during feeds, and the inability to pass a naso-gastric tube (NGT) further than 9-10 cm. This NGT is visualized on the chest xray at T2-4 in the blind ending oesophageal pouch. The presence of a gastric bubble confirms distal TOF. Later signs include recurrent LRTI and chronic aspiration (common in the H type).



CXR: Curled NGT in blind ending oesophageal pouch



Ultrasound: Reduced stomach filling

PRE-OPERATIVE MANAGEMENT

The main pre-operative goals are to prevent aspiration and dehydration.(3) Once a diagnosis is made, a Replogle tube (radiolucent, double lumen tube) is passed into the oesophagus to aspirate saliva and gastric contents. Fluid and electrolyte losses via this suction need to be replaced. Hypogycemia should also be avoided. A babygram is done to confirm the diagnosis, to assess the lung fields in the case of aspiration pneumonitis/chronic lung disease, and to exclude other VACTERL anomalies.

These infants need to be nursed in a 30 degree head up/lateral position to avoid further aspiration. Prophylactic antibiotics are suggested in many texts; however a high index of suspicion for pneumonia and sepsis should be maintained, in which case broad spectrum antibiotics should be started, with de-escalation of antimicrobials once an organism has been identified. Some sources suggest starting empiric antibiotics because of the high risk of aspiration (11). A pre-operative echocardiogram needs to be done to exclude congenital cardiac disease, as well as to identify the position of the aortic arch (1).

INTRA-OPERATIVE MANAGEMENT: SURGICAL CONSIDERATIONS

Surgical approach

Surgery usually occurs on day 1 or day 2 of life. It involves ligation of the fistula, and anastomosis of the oesophagus (1). The primary aim is to avoid aspiration. Palliative/corrective cardiac procedures are usually done after ligation/repair of the fistula (2). Long gap atresias (gap extending over 2 vertebral bodies) may be repaired in a staged procedure, where a gastrostomy is performed following ligation of the fistula, and division and anastomosis occurs later once the 2 ends have grown closer together.

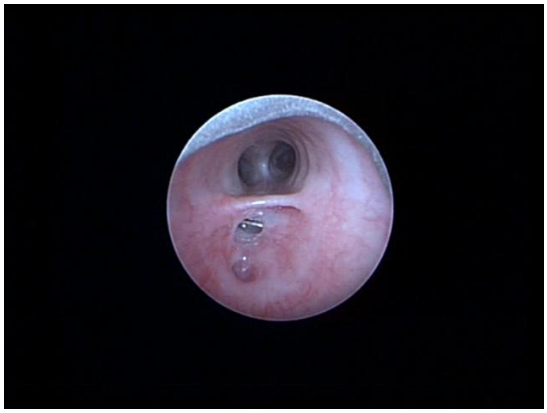
Premature infants with respiratory distress syndrome may only tolerate emergency ligation of the fistula, with oesophageal anastomosis a few days later when the patient is more stable. Minimally invasive thoracoscopic repairs are being done more frequently using strict patient criteria. Previously gastrostomies were performed pre-ligation in an attempt to avoid gastric distension, but have been found to cause broncho-cutaneous fistulae during ventilation.

Positioning

Most commonly, the patient is in the left lateral decubitus position with a right thoracotomy. Patients with a right sided aortic arch are placed in the right lateral position. H type fistulae are repaired in the supine position. It is important to ensure well-secured lines and tubes, and appropriate padding and protection of pressure points during positioning.

The role of bronchoscopy in TOF

It has become standard of care (4) to perform rigid bronchoscopy on spontaneously breathing anaesthetized patients before attempting intubation. This aids the anaesthetist in visualizing the airway and identifying the position of the fistula, as well as multiple or missed fistulae. It allows areas of tracheomalacia and bronchial abnormalities to be identified, which will also impact on the timing of extubation post-operatively. The bronchoscope can also be used to guide fogarty catheters and bronchial blockers, which will be discussed later. Bronchoscopy is also useful for the surgeon to characterize the previously unknown mediastinal anatomy, and decide on the surgical approach and course. Limitations to use of bronchoscopy include size in preterm infants, as well as poor functional reserve during the bronchoscopy in spontaneously breathing premature infants. Other complications of bronchoscopy include trauma, laryngospasm, bronchospasm, hypoxia and coughing.



Bronchoscopic view of TOF

INTRA-OPERATIVE MANAGEMENT: ANAESTHETIC CONSIDERATIONS

Monitoring

The pulse oximeter should be placed on the right hand; and in the case of patent ductus arteriosus, monitors should applied to the right hand and foot to measure pre- and post-ductal saturations. An arterial line should be inserted to measure invasive blood pressure during periods of respiratory and haemodynamic instability (12) during the surgery, including test occlusion of the azygous vein. Central venous catheter insertion has been described in terms of monitoring filling pressures of the right atrium (12), as a secure line for administration of drugs and fluids in infants with difficult intravenous access or haemodynamic instability; but is not essential in every case.

Challenges

The main anesthetic challenge is that of inadequate ventilation due to: (3)
Intubation of the fistula
Gastric distension
Pre-morbid aspiration and pneumonia/pneumonitis
Respiratory distress syndromes and respiratory disease of prematurity
Other congenital anomalies, especially cardiac.

Large (>3mm) peri-carinal fistulae pose the greatest anaesthetic challenge, and lend themselves to increased gastric distension and ventilation difficulties. Gastric distension occurs during positive pressure ventilation (PPV), where air moves down the path of least resistance (via the fistula in to the oesophagus and stomach). The consequences of this distension include splinting of the diaphragm, atelectasis, raised intra-abdominal and airway pressures and increasingly difficult ventilation. It has also been known to cause gastric rupture and pneumoperitioneum. These ventilation difficulties are compounded by co-existing cardiac and respiratory comorbidities. Numerous deaths have been associated with inadvertent intubation and/or ventilation of the fistula, aspiration, and significant hypoxemia (1).

Physiology of neonatal thoracic surgery in the lateral decubitus position

Effects of thoracic surgery on respiratory physiology:

- Reduced Functional Residual Capacity.
- Impaired Hypoxic Pulmonary Vasoconstriction caused by inhalational agents, and other drugs.
- Surgical retraction of the lung, or the need for one lung ventilation causes shunt and hypoxemia (5).

Effects of the lateral decubitus position on the neonate:
In adults and older children, spontaneous ventilation results in maximal ventilation and perfusion of the dependent lung zones. In the lateral decubitus position, adults and older children display better ventilation and perfusion in the dependent lung when the diseased lung is in the non-dependent position.

In infants, ventilation perfusion mismatch occurs more readily in this position due to:

- A very compliant rib cage that does not support the dependent lung, resulting in atelectasis and worsening of the shunt.
- A decreased hydrostatic pressure gradient between the dependent and non-dependent lungs, resulting in the loss of benefit of increased perfusion to the dependent lung.
- A reduced abdominal hydrostatic pressure gradient, and reduced mechanical load of the dependent diaphragm on the lungs.

Induction

Induction goals prior to rigid bronchoscopy include the prevention of aspiration and maintenance of spontaneous ventilation. The maintenance of negative intrathoracic pressure during spontaneous ventilation allows gas to move preferentially into the trachea and the [1]lungs. Inhalational techniques have been favourably quoted for induction; although intravenous Ketamine has been used. Both techniques have disadvantages, including coughing and hypoventilation associated with inhalational agents, and increased secretions with the use of Ketamine.

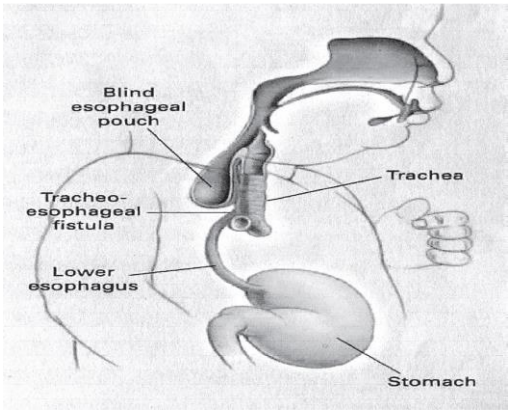
Intubation

Once bronchoscopy is complete, endotracheal intubation is a critical part of the anaesthetic. Awake intubations have become historical due to the resultant stress response, desaturation, raised intracranial pressure and increased incidence of associated intraventricular haemorrhage.

Maintenance of spontaneous respiration has been advocated for intubation for the aforementioned reasons, as well as easier wake up in the case of difficulty.

Apnoeic intubation has also been successfully described, requiring the use of short-acting opiates such as fentanyl, neuromuscular blockade and a ‘gentle bagging’ technique (1). This may not be feasible in patients with large fistulae, or poorly compliant lungs, where gas flow down the fistula causes distension of the stomach.

The most critical point in the procedure is prior to ligation of the fistula where risk of hypoxemia, aspiration and displacement of the endotracheal tube (ETT) is highest. Optimal position of the ETT would be distal to the fistula, and above the carina. This is a problem once again in large peri-carinal fistulae. It must be noted that once the patient is intubated, there is a high risk of displacement of the ETT into the fistula, or deeper into the airway once the patient has been positioned.



Type C TOF

Ventilation strategies

Various strategies have been documented to improve ventilation prior to ligation of the fistula. These include:

- ✓ High *fi*O2 of up to 100% has been advocated in term infants to maximize oxygenation. Permissive
- ✓ hypercapnoea prior to ligation of the fistula can be allowed; however it should be avoided in patients with congenital cardiac disease where an increase in pulmonary vascular resistance can worsen a right to left shunt.
- ✓ The use of a single lumen ETT, the position of which is confirmed by auscultation of bilateral breath sounds in the correct position relative to the fistula. (This is the most common practice in our setting, seeing as other airway devices are inappropriately sized or unavailable for such small patients).
- ✓ Rotation of the ETT so that the bevel is anterior to occlude the fistula.
- ✓ The use of a cuffed endotracheal tube, where the cuff is inflated over the area of the fistula to occlude it. Both of the above methods are not possible in low carinal fistulae.
- ✓ Left mainstem intubation to bypass the fistula, resulting in collapse of the right lung, and repositioning in the trachea once the fistula is ligated. The problem associated with this technique is using the correct tube size to prevent left bronchial oedema and significant leak in the tracheal position (2); as well as hypoxemia from collapse of the right lung, requiring periods of repositioning and 2lung ventilation, which can be cumbersome and difficult.
- ✓ A Fogarty embolectomy catheter can be inserted during rigid bronchoscopy, and inflated in the fistula. This has been described in large carinal fistulae. The problem is that the catheter can dislodge and obstruct the trachea, causing complete airway obstruction. The use of Bronchial Blockers has been similarly described, however in such small patients, this equipment is often unavailable, or becomes easily dislodged.
- ✓ A group from Zurich described the use of flexible tracheoscopy through a secured ETT to aid in positioning of the tube, and trans illumination of the fistula to hasten time to ligation. There are, however, many associated risks including accidental extubation, as well as lack of expertise in bronchoscopy in such a small airway (2).
- ✓ In acquired TOF in adults and older children, double lumen and Combivent tubes can be used to aid in suctioning and ventilation; however there are no small sizes for use in neonates (5).
- ✓ Maintenance of spontaneous respiration prior to ligation of the fistula avoids the use of PPV and inadvertent gastric distension, but usually is inadequate for thoracotomy.

In sick, small neonates with severe respiratory disease, emergency ligation can be performed. Once the fistula is ligated, normal PPV can continue to both lungs. The patient is then stabilized and brought back for definitive repair 8-10days later. Other emergency procedures include ligation of the oesophago-gastric junction (via an abdominal approach), and creating a cervical oesophagostomy, where the oesophagus is temporarily brought out to the skin of the chest wall.

Once the fistula is ligated either thoracoscopically or via a right thoracotomy, the endotracheal tube can be repositioned, with positive pressure ventilation applied to both lungs with neuromuscular blockade, for definitive repair of the oesophagus. Some centers perform gastrostomies following ligation of the fistula.

Division of the fistula and repair of the oesophagus requires one-lung ventilation and/or manual retraction of the lung for best surgical access. Seeing as these infants may have 2 healthy lungs, or 2 diseased lungs, one lung ventilation has a significant effect on ventilation/perfusion mismatching and shunt. Inadvertent damage to the phrenic nerve, great vessels, and tracheobronchial tree can occur.

Analgesic options

These are usually dictated by post-operative ventilation requirements, pre-existing comorbidities, duration of surgery and intra-operative course, post-operative pain, and tension on the oesophageal anastomosis (1). Systemic opiate infusions are more suitable for patients who will require post-operative ventilation. Neonatologists advocate the use of fentanyl rather than morphine, due to the effects of accumulation (1).

Epidural catheters have also been used as an 'opiate sparing' technique (2). These can be inserted via a caudal, lumbar or thoracic approach, and secured at a mid-high thoracic level (T6/7). Vertebral abnormalities must be excluded first. Its position can be confirmed radiologically (ultrasound or x-ray) or using electrical nerve stimulation. Regional anaesthesia in an otherwise healthy neonate, allows for early extubation.

It should be noted however, that especially in premature infants, the CYP450 system is immature and lower doses of bupivacaine should be administered for a maximum of 48hours post-operatively. Infusion rates of bupivacaine should not exceed 0.2mg/kg/hour. Local infiltration, paravertebral blocks and intrapleural infusion have also been described.

Surgical concerns for the anaesthetist intra operatively

The anaesthetist needs to be aware of the effects of left lateral positioning (discussed below), as well as the risk of accidental extubation and dislodging of devices occluding the fistula, kinking of the tube, as well as accidental intubation of the fistula. H type fistulae are usually repaired in the supine position with a cervical approach.

Surgical approach may be extra- or trans-pleural. Extra-pleural surgery allows retro-pleural anastomosis of the oesophagus, and also allows a leak to be better contained. Trans-pleural approaches may result in empyema secondary to an anastomotic leak. In such small patients, differentiation of mediastinal structures may be difficult, and the aorta, oesophagus trachea must be identified before attempts at ligation.

A test occlusion of the azygous vein before ligation and division of the fistula needs to be done to exclude haemodynamic changes and inadvertent clamping of the inferior vena cava. This necessitates the use of an arterial line for invasive monitoring.

Manipulation and retraction of the lungs, as well as the need for one lung ventilation for optimal surgical access may cause shunt and hypoxemia. This is particularly prominent during packing of the chest to mobilise and anastomose the oesophagus, and lung re-expansion may be necessary (3).

Thoracoscopic surgery necessitates neuromuscular blockade and positive pressure ventilation, as well as thoracic insufflation (although newer methods are being practiced). A nasogastric tube needs to be advanced into the distal oesophagus, around which the surgeon will perform the anastomosis. Once the anastomosis is complete, positive pressure is applied in a bubble leak test to exclude a leak from the suture lines.

THE ROLE OF THORACOSCOPIC SURGERY

These procedures are reserved for term, otherwise well infants with no underlying respiratory or cardiac comorbidities. These procedures require left lateral positioning and the insertion of ports into the chest. Thoracoscopic procedures usually necessitate neuromuscular blockade and positive pressure ventilation. One lung ventilation is not routinely employed, as the non-dependent lung is compressed by the insufflation of CO2 (5mmHg). Neonatal insufflators have been designed to try to overcome the problem of over distension of the chest cavity with CO2.

In large peri-carinal fistulae, left mainstem intubation may be necessary. The resultant hypoxemia can be addressed with use of high fi02 and gentle manual application of PPV to maintain oxygen saturation above 85% (4). ETCO2 reading will be unreliable during this period. Arterial CO2 will be raised, and must be noted in the setting of congenital cardiac disease and pulmonary hypertension.

Thoracoscopic surgery has been shown to reduce musculo-skeletal complications associated with open TOF repairs. These include winging of the scapula, chest wall asymmetry and thoracic scoliosis. Thoracoscopic approaches have been described as an 'opiate sparing' technique, with reduced post-operative pain. Studies have shown shorter extubation times and ICU stays; however surgical safety and complications are equivalent (2).

FACTORS AFFECTING EXTUBATION

Surgical

Ideally extubation should occur within the first 24hours to avoid pressure on the anastomosis. Spontaneous respiration reduces pressure on the suture line; however re-intubation poses the risk of damage to the repair. In patients with repairs that are performed under tension, they are usually ventilated with the use of neuromuscular blockade for a number of days to allow healing of the anastomosis. As mentioned, thoracoscopic repair decreases postoperative opiate requirements, and allows for earlier extubation. Patients undergoing repair of an H type fistula can usually be extubated early.

Patient

Patients with respiratory impairment (3) due to prematurity, pre-existing congenital cardiac disease, respiratory distress syndrome, aspiration pneumonitis and tracheomalacia usually require post-operative ventilation. Otherwise well, term neonates with uncomplicated repairs with epidural analgesia can be extubated in theater.

Anaesthetic

As mentioned, neuraxial/regional anaesthesia allows for early extubation in otherwise healthy infants. In patients who will be ventilated post operatively, a systemic opiate infusion can be used. Patients with an unstable intra operative course, should also be ventilated post operatively.

POST-OPERATIVE MANAGEMENT

All patients should be transferred to a neonatal ICU facility following TOF repair, for respiratory monitoring and pain control (1), irrespective of the need for mechanical ventilation. The chest drain is placed in enough water to seal it, and is not attached to suction (6). Antibiotics should be continued until the chest drain is removed.

Regular pharyngeal suctioning with a clearly marked suction catheter should occur, so that the catheter is not advanced far enough to cause trauma to the anastomosis. It is important to differentiate, label and secure the endotracheal and trans-anastomotic feeding tubes, to avoid reintubation as well as damage to the anastomosis and displacement of the feeding tube.

Controversy exists regarding the risk of prolonged intubation *vs* the potential damage caused by reintubation (3). It has been argued that early extubation avoids the effects of positive pressure ventilation on the anastomosis, as well as decreased pressure effects of the endotracheal tube on the repair site. Delayed extubation offers protection of a better healed anastomosis should reintubation be required, however it is associated with the well described risks of prolonged mechanical ventilation.

Controversy surrounding the routine use of neuromuscular blockade and post-operative mechanical ventilation also remains a point of discussion between disciplines. Case studies have shown that the use of paralysis and elective post-operative ventilation has improved outcomes in terms of anastomotic healing. There are opposing views in critical care literature with regard to prolonged recovery from neuromuscular blockade, as well as associated myopathies.

The aforemementioned surgical case series suggested that despite advanced surgical technique and new suture material used, the use of post-operative paralysis and elective ventilation contributes more to successful outcomes and anastomotic healing (7). Other local surgical studies have cited the importance of caudo-thoracic epidurals in decreasing the need for post-operative ventilation in a resource-constrained setting (8).

Another study suggested that timing of extubation is particularly important in TOF repairs in terms of the need for reintubation and risk of damaging the anastomosis; and therefore suggests extubation once the patient is no longer dependent on opiates for post-surgical pain control (9). In terms of paediatric critical care, it has been suggested that there is no need for prolonged ventilation in stable infants with satisfactory anastomoses (10). In infants with anastomoses under tension, they suggest 3-5 days of paralysis and elective ventilation.

Following extubation, upper airway oedema, respiratory arrest and recurrent laryngeal nerve injury have been described, necessitating re intubation (2). Failure to wean mechanical ventilation should alert one to the possibility of recurrent or missed fistulae.

Trans-anastomotic feeding should start 48 hours post repair, with enteral feeds being introduced around day 5, following a confirmatory barium swallow or other radiological investigation that shows integrity of the anastomosis (1).

COMPLICATIONS

Surgical postoperative complications include anastomotic leak, stricture, gastroesophageal reflux, tracheomalacia, and recurrent TOF (3).

Tracheomalacia may be severe enough to warrant reintubation in 10-20% of cases (1). In some cases, urgent aortopexy is required.

Anastomotic leaks occur in 15-20% of cases, the majority of which are small and heal spontaneously. Major leaks can manifest as saliva draining from the intercostal drain, or pneumothorax.

Anastomotic strictures occur in 30-50% of cases, most of which require dilatation. Stricture formation and anastomotic leak has also been linked to the gap length between the initial proximal and distal oesophageal ends, with gap lengths >3.5cm resulting in 100% risk of stricture formation and 80% of leak (1).

Recurrent fistula occur in 10% of cases.

Later complications include gastro-oesophageal reflux (GOR), and recurrent lower respiratory tract infections from chronic reflux and tracheal epithelial dysmotility (1). Half of these patients will present to theater for surgical management of their GOR. Others will present for recurrent oesophageal dilations; however most will outgrow these issues.

CONCLUSION

It is important to identify and diagnose make the diagnosis of tracheo-oesophageal fistula early. Pre-operative resuscitation and avoidance of aspiration is central to the emergency medical management of patients with TOF, prior to surgery. Associated comorbidities, especially cardiac and those associated with prematurity, affect the prognosis.

The major anaesthetic challenge is airway management prior to ligation of the fistula, especially large peri-carinal fistulae, due to the risk of the inadvertent fistula intubation and ventilation and distension of the stomach. Post-operative concerns relate to the timing of extubation with regards to anastomosis healing and the effects of prolonged mechanical ventilation.

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