UNIVERSITI MALAYA

INAUGURAL LECTURE

"CHALLENGES IN PAEDIATRIC ANAESTHESIA"

Professor Dr. Lucy Chan @ Helen Chan Kam Wan
Department of Anaesthesiology,
Faculty of Medicine

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PROFESSOR DR. LUCY CHAN @ HELEN KAM WAN
DEPARTMENT OF ANAESTHESIOLOGY, FACULTY OF MEDICINE
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Syarahan Perdana ini di adakan sebagai pengiktirafan jawatan Profesor di Universiti Malaya.

PROFESSOR DR. LUCY @ HELEN CHAN KAM WAN

MBBS (SINGAPORE), FANZCA (AUSTRALIA & NEW ZEALAND)

BIODATA

Professor Dr. Lucy Chan received her early education in Convent schools in Ipoh and Batu Gajah, Perak. She entered St Michael's Institution to complete her Upper and Lower Sixth. It was from this good background that she found an interest in books and learning.

She had thought of a career as a librarian or a biochemist but a State Scholarship from Perak enabled her to pursue medical studies in Singapore University from where she graduated with a MBBS in 1972. She spent her early years as a young doctor in Kuala Lumpur Hospital, Kuantan General Hospital and Seremban General Hospital.

She found her career in anaesthesiology when she was posted to the Anaesthetic Unit as a medical officer in Kuantan General Hospital in 1973 and had not turned back since.

She passed her anaesthetics in FANZCA in 1983 and joined as a lecturer in the Department of Anaesthesiology, UMMC, University Malaya from 1998. She received her promotion as an Associate Professor in 1995 and became Professor in Anaesthesiology in 2003. Since she joined the academic staff in UMMC, she has been involved in the teaching of undergraduates and postgraduates students, providing anaesthetic services to the hospital in the operation theatres and intensive care unit, setting up research work and sharing administrative duties in the Department of Anaesthesiology, UMMC.

She is a member of the Malaysia Medical Association, Malaysian Society of Anaesthesiologists, Australia and New Zealand College of Anaesthetists, Academy of Medicine, Malaysian Association for the Study of Pain and Parenteral Enteral Nutrition Society of Malaysia. She is also a member of TIMA (Tzu Chi International Medical

Association) a merit/charity NGO that provides free medical care to the sick and underprivileged.

She is an active coordinator and examiner in both Part 1 and Part 2 of the Conjoint Master in Anaesthesiology i.e. M.Med (Anaesthesiology), UKM and Master of Anaesthesiology (UM).

She has published more than 32 articles in regional and international anaesthetic journals and there are citations in various Proceedings, Seminar and Workshop. Her writings include research into bacteriological contamination of a closed enteral feeding system, blood cholinesterase levels in Malaysians, use of the VMB Laryngeal Tube in anaesthesia, pharmacokinetics/pharmacodynamics of drugs and BIS monitoring during anaesthesia. Among scientific material presented are papers on Malignant Hyperthermia and airway in paediatrics. Both fields are her special interest, apart from Pharmacology and teaching.

She is a reviewer for 2 prestigious journals -*Asean Journal of Anaesthesiology* and *Anaesthesia and Intensive Care*. She has been awarded "excellent service award" by the University of Malaya in 1999 and 2002.

Recently she has been selected to be a Convenor to the Special Interest Group"- Paediatric Anaesthesia and Analgesia - under the College of Anaesthesiologists/ Academy of Medicine from 2003.

INAUGURAL LECTURE

Delivered on 22.9.03 by

Professor Dr. Lucy Chan @ Helen Chan Kam Wan

Ladies and Gentlemen, Today's presentation is entitled "Challenges in Paediatric Anaesthesia" It includes a brief mention of the past and future. The challenges to be described involve airway management, neonatal emergencies and separation of conjoined twins. Some of the examples are very rare but all are interesting encounters in children who required general anesthesia during my 2 years as a Medical Officer and 14 years as an academic staff in the Anesthetic Department, University Malaya Medical Centre. University Malaya.

THE PAST

The physician must know, what the physicians knew before him, lest he deceives himself and others" - Hippocrates .

There is no profession that owes more to the past than does medicine. Around 2500 BC, ancient Egyptians were advanced medical practitioners who minimized screaming and fighting during surgeries by sedating patients with opium and herbal poisons like mandrake and henbane.

However, the art of medicine in the ancient world developed to its highest point in Greece between 500 BC and 50 AD and this creative period is symbolized by Hippocrates "Father of Medicine". Hippocrates' name has come to represent beauty, value and dignity of medicine for all times as embodie d in these words:

"Where there is love for mankind, there is love for the art of healing"

What was most convenient in ancient days was a direct but crude way of inducing a state of insensitivity: to knock the patient unconscious with a blow to the jaw. That was quick anaesthetic but it was not specific or complete and the surgeon had to finish the amputation before the patient recovered from the shock.

Today, surgery and anaesthesia are not carried out under such conditions for adults and definitely not for children. Anaesthetists work in a pleasant operation theatre and use sophiscated equipment. In the past, the 2 greatest obstacles to major operative procedures were pain and infection. It was Joseph Lister (1827 – 1912) who developed an antiseptic system for hospital surgery and anesthesia. Pain in surgery, in particular, had been a scourge upon mankind long before the dawn of history.

In 1803 FW Serturner isolated a powerful analgesic agent from crude opium and named it *morphine* after Morpheus the Greek god of dreams. It was not until the invention of the hypodermic syringe by Charles G Pravaz (French surgeon) and Alexander Wood (Scottish), that injections of morphine were used. The story of the successful development of surgical anesthesia began in 1840s. Four men played leading roles in bringing about the advent of practical anaesthesia.

1) Crawford W Long a physician who first used ether as an anesthetic 1842.
2) Horace Wells a dentist who used nitrous oxide as an anesthetic in his dental practice 1844.
3) William TG Morton a dentist studying medicine, successfully used ether in surgical operation 1846.
4) Charles T Jackson, physician and chemist, whose suggestions and challenges led Morton to experiment further with the sleep-inducing effects of ether in 1846.

Dentists were probably more highly motivated than any other practitioners to discover novel and powerful anesthetics. If a drug or technique could eliminate the excruciating pain of a rotting tooth, it could provide pain relief for other surgical operations.

The first recorded case of pediatric anesthesia was in 1842 when Crawford W Long of Georgia, administered ether to a 8 year old boy for amputation of the toe. Six years later, another toe job led to the first anesthetic death in 1848. Hannah Greener, fifteen year old, had a toenail removed under chloroform and probably aspirated on medicinal brandy given to her while she was semiconscious. The staff of Aesculap (fig 1), entwined by a snake, is regarded as a true symbol of our profession. The words, translated, mean "life is short, art is long, experience difficult". These ancient words aptly describe the struggle of the early years that have benefited us beyond comprehension.

Figure 1: ***Staff of Aesculap***

Ensuring that the surgeon operates on a nonmoving, painless and physiologically stable patient is the goal now, just as it was a century ago.

Today modern pediatric anaesthesia includes: new anesthetic agents, more advanced technology and equipment, innovative techniques, and special training that provides education in all aspects of pediatric anesthesia.

CHILDREN WITH AIRWAY PROBLEMS

"In skating over thin ice, our safety is in our speed""– Ralph Waldo Emerson

What then are some of these challenges? A major challenge is to anaesthetize a child with airway problem. Three scenarios will be used as examples: namely,

- children with anatomical changes to the airway undergoing surgery
- children with airway foreign body for removal under anaesthesia
- children for airway laser surgery

Children, unlike adults, present at different levels of development, psychologically and physically and have their own ailments. For example, the anatomy of the infant's airway is different. The narrowest point is the cricoid ring, in contrast to adults - which is the epiglottis. Due to the narrowing of the airway at the cricoid ring, the infant airway is funnel-shaped and this natural narrowing is the reason why uncuffed endotracheal tubes are used in infants.

Infants have a short trachea (2 inches)and small internal diameter, 4mm, compared with an adult trachea with diameter 20mm and length 5inches. The adult airway has an area 25 times larger than the infant airway! If the adult airway has 1/2 mm of edema - the area will decrease by 10%. If the infant airway has the same 1/2 mm of edema, the area of the airway decreases by over 50%! It clearly shows that infants will be more severely affected by changes in airway diameter than adults.

Therefore, any decrease in airway diameter due to secretions or inflammation can significantly increase resistance to breathing or ventilation. Neonates and children who require major surgeries or who are paralysed and ventilated require tracheal intubation.

Intubation ensures patency of airway and aspiration of gastric contents is avoided. The process of intubation should take minimum time and should smooth and atraumatic.

Anatomical changes to the airway

(1) acquired

A difficult airway may result from altered or distorted anatomy especially to the head and neck and a plan of action has to be ready prior to putting the child to sleep.

A large cystic hygroma can displace a central trachea far to the left. Holding a fitting face mask is not simple and as for intubation, adequate viewing of the glottic opening requires proper head and neck positioning. For intubation a laryngoscope is needed to view the laryngeal inlet and through this is passed an appropriate size pediatric uncuffed endotracheal tube into the trachea. The distal end of the tube should lie above the carina so that both lungs are equally ventilated. The infant's lungs are auscultated frequently and monitoring of oxygen by pulse oximetry is essential.

The infant's head is larger in proportion to body size when compared to adult. Therefore, there may be difficulty encountered at laryngoscopy and intubation in neurosurgery for a child with a large head and relatively short neck. Tumours of the CNS constitute the second most common neoplasm in children after leukemia. This child (Fig 2) has a brain tumour associated with hydrocephalus and is ready to be anaesthetized for excision of a recurrent astrocytoma. After the operation, the child is extubated when he is awake and vital signs are stable.

Figure 2: *Child with brain tumour*

Attention to positioning is also necessary with an occipital myeloencephaloceole protruding from the back of the head. Once the airway is secured with endotracheal intubation, it is important to fix the tube well and maintain its position to provide ventilation throughout operation especially when the surgery is to be performed with the child in the prone position.

(2) congenital

The above examples are children with airway difficulties having a normal laryngeal and tracheal airway but have an acquired abnormality affecting access to the airway. There are children who have airway problem as a result of a congenital abnormality. A not uncommon congenital syndrome is the Pierre Robin sequence.

The original text that describes this syndrome includes a small chin or micrognathia, a cleft palate accompanied by breathing problem which is both inspiratory and expiratory distress and a large tongue i.e. relative macroglossia. The most important anatomic feature of difficult airway is micrognathia (fig 3).

Figure 3: *Micrognathia in Pierre Robin child*

Hypoplasia of the jaw decreases the space in which soft tissue can be displaced by the laryngoscope. Tracheal intubation can be very difficult especially in the first few months of birth because of micrognathia and a more posteriorly placed tongue

The large tongue and small mouth can cause severe airway problem in the supine position from birth and the upper airway obstruction can lead to hypoxia, pulmonary hypertension, cor pulmonale or failure to thrive which is secondary to difficulty in swallowing and aspiration. Various methods of surgical fixation of the tongue have been attempted in order that the large tongue does not flop backwards in the oropharynx and obstruct breathing. A tongue-lip adhesion is performed to prevent the tongue falling back.The stitch is secured by 2 buttons, one at the chin and another on the tongue. The buttons are left in place for about 2 weeks during which time tongue is adhered to the lip (6 weeks to a year later or at time of palate repair, the tongue-lip adhesion is released)

In the worst cases of care of Pierre Robin child a tracheostomy may provide the only safe method of airway management. Anaesthesia in plastic surgery for cleft lip and palate repair is a challenging area. The congentital cleft lip is caused by failure of fusion in the medial and lateral nasal swellings which normally occurs by 35 days of intrauterine life.

This may subsequently impair the closure of the palatine shelves giving rise to the cleft palate. Isolated cleft palate has a higher incidence of association with other congenital anomalies such as congenital heart disease and umbilical hernia. A situation of difficult airway management occurs in holoprosencephaly, a rare craniofacial birth defect that includes: cleft lip, nasal and orbital malformation and severe mental retardation (fig 4).

Figure 4: ***Holoprosencephaly and bilateral cleft lip***

A detailed preoperative evaluation is required. Anesthetic management is complex because of the multiple congenital defects and physiological derangements. Potential airway problems include: difficulty in placing a fitting mask and intubation is not easy (some need awake intubation).

The presence of apnoeic spells require careful postoperative respiratory monitoring. Temperature fluctuations during surgery can further complicate anaesthetic management. There are over 3,000 genetic syndromes and it is impossible for the physician to recognize all of them but the common ones. No syndrome has yet been identified for this boy (fig 5) who has a hypoplastic chin and bilateral club feet.

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Figure 5: *Child with hypoplastic chin*

He had 3 failed intubations for club feet surgery starting from age 9 months. At 5 years of age he was successfully intubated with a pediatric fibreoptic bronchoscope (FOB). The FOB (Fig 6) has in recent years become immensely useful and popular for all ages of children with difficult intubation.

Figure 6: *Fibreoptic bronchoscope*

The use of FOB is a skill that comes with usage under controlled conditions. Many children presenting for minor surgical procedures are in good health and pose no special problems from confounding medical conditions. However, there are children with complex medical conditions who present for seemingly minor surgical procedures such as hernia repair and pose a challenge for even the most experienced anaesthesiologist.A rare medical disorder that has serious airway implications is mucopolysaccharidosis or MPS.

It is a lysosomal storage disorder characterized by progressive and diffusive accumulation of mucopolpolysaccharides in lysosomes in bone, muscle, visceral organs and the soft tissues of the mouth and pharynx. The combination of mental deficiency with limited ability to cooperate, profuse airway secretions and increased soft tissue infiltration with mucopolsaccharides makes airway management and intubation a major anaesthetic problem, particularly in children past infancy. If muscle paralysis is required, for example in a laparotomy, intubation can only be performed with the aid of fibreoptic bronchoscopy.

However, surgery can proceed with spontaneous ventilation with the insertion of a laryngeal mask airway (LMA) for the repair of umbilical hernia.

Figure 7: *Laryngeal Mask Airway*

The LMA (fig 7) is one of the best inventions of the 20th century for the practice of airway management in anesthesia. It was designed by Archie Brain in 1981 and made commercially available in 1988. It is an ingenious alternative to a face mask and endotracheal tube and is an invaluable option in any failed intubation.

FOREIGN BODY IN THE AIRWAY

Another group consists of children with sudden obstruction of the airway such as foreign body aspiration which is potentially fatal. Children of almost any age can present with foreign bodies (FBs) in the airway, most common under 3 years of age. Children with airway FBs present as airway emergency. The severity of FB aspiration is determined by whether airway obstruction is complete or partial.

The anaesthetist who examines the child with FB aspirated has to consider the type of foreign body involved, the site of obstruction and the degree of airway compromise that determines urgency of removal. Foreign bodies aspirated are organic or inorganic. Food particles are common especially peanuts and seeds because of improper feeding. Hot dogs are a common cause of fatal food aspiration because of its large size. Examples of inorganic foreign bodies are coins and small batteries.

After initial coughing there may follow a history of cyanosis, shortness of breath wheezing or the child may remain quite asymptomatic with delayed presentation of secondary complications such as bronchiectasis. The anesthetist and surgeon share the airway and they work closely as a team.

The problems of the anaesthetist are: control of airway, a full stomach and a potential difficult airway. Anesthesia under such circumstances is often hazardous. Rigid bronchoscopy is the mainstay for retrieval of foreign in the respiratory tract by the surgeon. The traditional approach to anaesthetize the child is an inhalational induction with oxygen and a volatile anesthetic such as sevoflurane.

Under anaesthesia adverse airway events may occur: hypoxic episodes are common and there may also be hypoventilation, trauma, inflammation, oedema, barotrauma and bleeding in the airway.

In a "5 year retrospective review (1998-2002) of foreign bodies in the ear, nose, airway and oesophagus" which was undertaken together with medical students, FBs in the airway constituted 29% among the 4 locations studied. The study showed that all FB removal for children were carried out under general anaesthesia. Among the organic FB were ciku seed and peanuts and they were removed with the aid of a pediatric rigid bronchoscope under deep inhalational anaestheticThe commonest inorganic FB found were coins located at the post-cricoid or pyriform fossa.

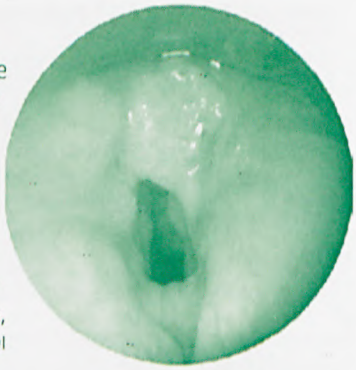
During the study period there were no complication following bronchoscope removal for airway FB except in a child who aspirated a light emitting diode bulb into the right main bronchus. Subsequently, thoracotomy was performed to remove the diode bulb.

AIRWAY LASER SURGERY

The last group requires laser surgery.

Children with laryngeal papillomatosis (fig 8) present with progressive airway obstruction that may acutely obstruct with an episode of upper respiratory tract infection.

Figure 8: Laryngeal papillomatosis



The only way for the ENT surgeon to provide the child with a clear airway is remove or reduce the bulk of laryngeal papillomatosis.

Because laser enables very precise excision and minimal oedema, it is particularly suitable for this type of airway surgery. The CO2 laser is most appropriate.

Bronchoscopic removal of FBs and airway laser surgery have one thing in common: in both instances, the airway is shared between anesthetist and surgeon and a close rapport is required in this teamwork.

However the use of laser has some inherent hazards. The laser beam can ignite flammable material such as endotracheal tubes, breathing circuits, drapes and lubricants resulting in fires or even explosions. Of particular concern to anesthetists is the problem of fire which may occur if the laser ignites flammable tracheal tubes. Human tissues can also be burned such as the skin if the laser beam is misdirected. The eyes are particularly vulnerable to laser related injuries.

Safety precautions can prevent most serious injuries. The OT doorway should be labeled with a warning sign "laser in use". Eye protection with goggles is required for OT personnel and patients' eyes are covered with wet gauzes. CO2 laser can cause corneal burns. Anaesthetic management during such a procedure includes the use of a small endotracheal tube or no tube at all. Surgery without a tube enables the surgeon to have a full view of the laryngeal inlet. If there is no tube, a fire is unlikely as no flammable material is in the airway.

Two common techniques to provide anaesthesia when there is no tube are to use jet ventilation and insufflation. Jet ventilation provides the oxygen while the child is kept immobile with anesthetic drugs. It is effective but contraindicated when major airway obstruction is present because of the increased risk of inadequate lung emptying and the resulting barotraumas and pneumothorax. It requires special equipment.

The insufflation technique requires simpler equipment. A gas inlet device can be attached to surgeon's endoscope or a catheter can be placed just above the glottic opening. With a catheter placed above the vocal cords, pharyngeal insufflation is possible and by insufflation, the child breathes a mixture of oxygen and volatile agent and remains anesthetized while laser surgery is in progress.

During lasering, an unprotected airway however has risks, such as hypoventilation and aspiration of gastric contents or surgical debri. The catheter can also burn if hit by laser beam.

The second option is to provide anaesthesia with the use of a tube. Tube selection is important. The conventional tubes are PVC, red rubber or silicone – they are readily ignitable and flammable and need wrapping with metallic tape. Special metal tubes are available such as the "laser-flex". It does not burn.

There are advantages if there is a tube: a secure airway, protection of lower airways from tissue debri and avoidance of OT room pollution from anaesthetic gases.

However there are disadvantages such as obstruction of surgeon's view, trauma and non-metal tubes are also combustible Children with laryngeal papillomatosis require repeated GA's over the years because there is no definitive cure.

A few severe and intractable cases require a surgical airway, that is, a long-term tracheostomy for safe and adequate management. Tracheostomy tubes for children are shorter with smaller diameter compared with adult tracheostomy tube. Under anaesthesia, the surgeon makes a hole in the trachea and inserts in the trachesotomy tube which is tied round the neck. The child breathes in the anaesthetic vapour through the tracheostomy.

It is important to stress that children are not small adults. Those with difficult airways are even more problematic than their adult counterparts who are difficult to intubate. For one thing, children will not cooperate. Consequently not only must airway equipment for difficult airway be smaller with a wider range of sizes, but a variety of techniques must be available as well. Important advances in the management of the difficult pediatric airway over the past decade include the fibreoptic bronchoscope and the laryngeal mask airway.

NEONATAL EMERGENCIES

"So long as little children are allowed to suffer, there is no true love in this world"
'– Isadora Duncan

A group of children that deserve mention are neonates: they are recognized as having special anatomic, physiologic and pharmacologic concerns. From an anaesthetic point of view, when a neonate presents for emergency surgery, understanding these differences along with the underlying pathology of the surgical emergency is essential.

Therefore, there must be detailed preanaesthetic evaluation and preparation for surgery. The anesthetic implications on the presence of related or unrelated congenital malformations need careful consideration

In particular, the preterm infants are at great risk for a list of problems, such as: hyaline membrane disease, intraventricular hemorrhage, hypothermia, hypoglycemia, apnoea, patent ductus arteriosus, hypocalcemia and retinopathy of prematurity. Fluids and electrolyte status require urgent assessment and correction especially where surgical emergencies involve the gastrointestinal tract. A possibility of a difficult airway is always there and a plan to ensure airway patency perioperatively is important. A period of stabilization is often possible in neonatal surgical emergencies and after the operation, in most instances, the neonate is cared for in the intensive care unit.

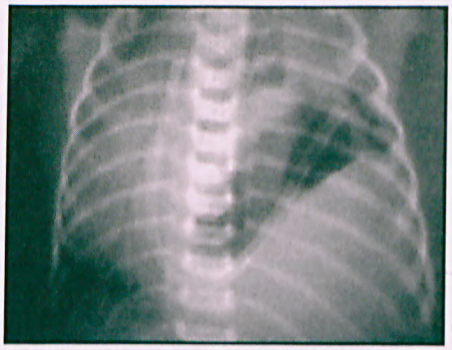
An important neonatal emergency is congenital diaphragmatic hernia (CDH) where congenital herniation of the abdominal viscera through the diaphragm happens. The most common location is the left posterolateral. It has an incidence of approximately 1:2500 live births. The hemithorax is filled with stomach and loops of intestines, rarely accompanied by the spleen, liver or kidneys. About 15% have associated hypolasia of the contralateral lung.

There is variable lung hypoplasia. When the newborn presents with signs of respiratory distress at birth, there is a greater degree of lung hypolasia and a greater mortality rate approaching 50%. When examined at birth, there is a scaphoid abdomen

and the neonate is cyanosed with tachypnoea, retraction and grunting. Intubation and ventilatory support for a few days before surgery may be required in a deteriorating neonate.

During surgery care is taken to avoid high positive pressure ventilation because this increases gastric distension and intrathoracic pressure and may result in pneumothorax to the contralateral lung. Unless hernia is exceptionally small, the newborn is left intubated and ventilated for transfer to the ICU. This is the chest x-ray of a newborn (fig 9) with CDH.

Figure 9: *Chest X-ray of a child with CDH*



His chest X-ray shows a catheter coiled up in the stomach in the left lung. Part of the liver is also in the left thoracic cavity. (Surgical closure of the large diaphragmatic hernia was performed on day 12 of birth).

An emergency condition where adequate fluid resuscitation is paramount before surgery begins is gastroschisis, a condition where there is a defect during development of the abdominal wall and there is no sac covering the exposed viscera. The viscera have been exposed to amniotic fluid and are usually oedematous and adherent from inflammation.

Primary closure can result in a tense and tight abdomen that does not provide adequate ventilation or the ability to clear secretions. Compressing too much bowel into the abdomen can also result in hypotension, decreased cardiac output, bowel ischemia and renal ischemia. Although it is desirable to have primary closure, a staged repair with a "silo" (fig 10) is used if the abdominal cavity is too small to accept all the viscera.

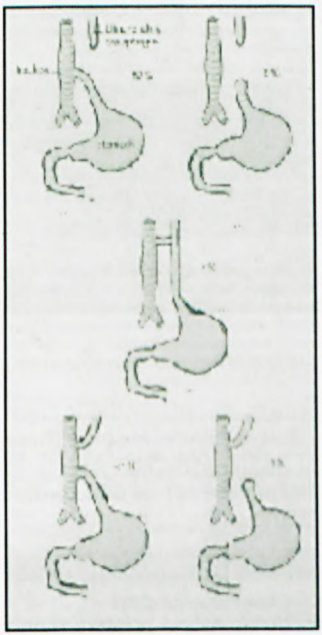
Figure 10: *Gastroschisis with "silo"*

The silo, an artificial sac of prosthetic material, is constructed around the exposed bowel and suspended to prevent excessive pressure on the underlying bowel and aortocaval system. The silo will be systematically reduced over days or weeks for oedema and inflammation to settle before surgical closure of the abdominal defect.

One of the more common conditions requiring surgical intervention in the newborn is tracheoesophageal fistula or TOF. It is considered a test of the anesthetist's skill with the airway. TOF occurs between 1:3000 and 1:4500 live births.

TOF occurs when normal separation of the trachea and oesophagus does not occur during fetal development. Several potential abnormalities can occur in TOF as shown (fig 11)

Figure 11: ***Tracheoesophageal fistula***



Survival is linked very closely to birth weight. The most important marker for mortality and morbidity in TOF is the presence of other major abnormalies especially cardiac and prematurity. An immature respiratory system is significant because aspiration pneumonitis from TOF may be complicated by infant respiratory distress system, making ventilation and oxygenation especially challenging.

The most common presentation is where the upper oesophagus ends in a blind pouch and a lower esophagus that connects to the trachea by a fistula. Breathing results in gastric distension because the neonate is breathing into the stomach and during feeding milk is aspirated into the airway leading to choking and coughing because the oesophagus ends in an upper pouch. The diagnosis is suspected by failure to pass a catheter into the stomach.

The key to successful anaesthetic management is correct endotracheal tube position: ideally the tip of the breathing tube lies between the fistula and carina so that anesthetic gases pass into lungs instead of stomach. The surgeon divides the fistula and primary oesophageal anastomosis is performed. These infants require 100% oxygen despite the risk of retinopathy of prematurity. Blood should be immediately available for transfusion.

Most TOF continue to require intubation and ventilation in the immediate postoperative period. Neck extension and suctioning of the esophagus may disrupt surgical repair and should be avoided.

Postoperative monitoring is important because complications can occur: gastroesophageal reflux, aspiration pneumonia, pneumothorax, tracheal compression and anastomotic leak

SEPARATION OF CONJOINED TWINS

"We are so fond of one another because our ailments are the same""– Jonathan Swift

An event although extremely rare that continues to fascinate physicians and lay public is the birth of conjoined twins. Since the first reported successful separation of conjoined twins by Konig in 1689, separation operation has continued to challenge anaesthetic and surgical teams

The incidence of conjoined twins is very low; 1 in every 100,000 to 200,000 births. Conjoined twins happen when an embryo starts to split into identical twins after conception but for some reason the separation process stops by the end of the second week and the fetus continues to develop growing into 2 babies that are fused together.

More than 600 sets of conjoined twins have been reported worldwide. The overwhelming majority are females 75%. The chance of postnatal survival is tiny: around half the births are delivered stillborn and another third survive just one day. Ideally separation should be delayed until after the neonatal period to allow tissue growth.

Conjoint twins are classified by their most prominent site of connection. Whether twins can or should be surgically separated depends where the conjointment is and the essential organs shared. If the twins have their own set of organs the chances of a successful operation are better than if they share the same heart or other vital organ. But when there are shared organs, in many cases the separation leads to the death of one or both twins. And even with successful separation, there may be lasting damage to the twins' health or they may require many surgical corrections thereafter. Currently there remain the insoluble challenges of surgery: finding substitutes for essential organs, dealing with tissue trauma and stemming blood loss.

What problems do anaesthetists face in separation surgery?

(1) Requires a multidisciplinary approach. Dedicated pediatric/surgical/anesthetic team management is vital to provide perioperative care. In the preoperative stage there are multiple meetings of the various teams. Surgical and anesthetic techniques are discussed. Mock trial and dress rehearsal are run before the actual day, so that everything proceeds smoothly and everyone involved knows his or her role.

(2) Babies should be investigated thoroughly and optimized as far as possible unless emergency situations arise. Separation should be unhurried to permit infant growth. A variety of diagnostic studies may be necessary to define organ sharing and demonstrate coexisting anomalies. Conjoined twins may require GA for diagnostic and emergency procedures not involving separation.

(3) Risks are explained carefully to the parents and include long hours, massive blood loss, biochemical fluctuations, hypothermia and mortality.

(4) 2 teams of anaesthetists and nurses are necessary. A senior anesthetist coordinates all the anesthetic input for perioperative care.

(5) On the day of separation, technical problems are multiple. Special operation tables may be necessary and the operation theatre is prepared for 2 sets of equipment, colour-coded for each twin, such as warming devices, infusion pumps, anaesthetic machines and a variety of electronic monitors. Intubation is a great challenge and usually the weaker twin is anesthetized first, positioning the second twin may involve supporting it semi-prone while the other is intubated supine. Intubation and placing all necessary invasive monitoring devices may take several hours before definitive surgery can proceed. Efforts are made in placement of personnel and machinery to avoid room crowding. Doses of drugs are precalculated. However, cross circulation may result in the second twin falling asleep as the first twin is anaesthetized. The intensive care support for postoperative management includes optimized facilities for vital system support for 2 babies after separation.

What is the history of conjoined twin separation in UH?

I joined the Anaesthetic Department as a medical officer in July 1981 and therefore missed, by 4 months, the historic day of Malaysia's first set of conjoined twin separation in 15 March 1981. Professor AE Delilkan was the consultant anaesthetist in charge. The TOH twins were joined at the pelvis or ischiopagus twins (fig 12)

Figure 12: ***Toh conjoined twins***

One is alive today.

Subsequently, I was able to witness four and a half sets of conjoined twin separation. The half belonged to one set of conjoined twins who had liver separation only.

The second and third were both separated in 1982. The Salina twins were joined at the pelvis and both were separated April 1982. The twins are alive today but require genitourinary follow-up. The Navaratnam omphalopagus twins were joined at the abdomen and were separated in November 1982. One twin died on the 6th post-operative day.

Sixteen years later, in7 March 1998, the 4th set of twins, the PANG twins, joined at the pelvis (ischiopagus tetrapus) were separated. The Pang twins are leading as full a life as possible with long term follow-ups for bladder and bowel function.

More recently we have the Rosli twins, Ahmad and Mohamad. They were joined at the chest, abdomen and pelvis (thoracoomphaloischiopagus). The liver separation was performed at age 2 in February 2001 and took about 15 hours. They were fully separated in 17th September 2002 in Saudi Arabia.

Adam and Adiq the latest set of conjoined twins (fig 13), were joined at chest and abdomen, (thoracoomphalopagus) were separated in 13 August 2002 when they were 4 months of age and weighed 8.7kg. Operative time was about 24 hours.

Figure 13: ***Adam and Adiq conjoined twins***

Adam died 3 weeks after separation and Adiq passed away before his first birthday.

Successful anaesthetic management of conjoined twins undergoing separation requires timing and detailed preoperative preparation, teamwork, an appreciation of intraoperative risks involved and intensive intraoperative monitoring.

THE FUTURE

"The child sunned by love and security will be able to withstand the storms of illness and pain"" – WJ Potts

I am in this field because it is a rewarding lifelong experience and caring for a child is similar to caring for the whole life. We are in the process of building a new pediatric unit with its attached surgical wards and operation theatres where we will continue to give our support to children there. The physical structure is coming up and the training in pediatric anaesthesia has therefore to continue and improve in order to become a dynamic subspecialty.

Research in pediatric anaesthesia has lagged behind other disciplines. Hopefully, the future will see a greater role in clinical anaesthesia that will spur the development of even newer ideas and methods to meet challenges in this subspecialty. Anesthesiologists no longer administer anesthesia alone; they deliver sophisticated and complex intensive medical and critical care in the operating room and intensive care units.

Finally, to be a competent paediatric anaesthetist requires commitment, a patient caring attitude and long years of practice and hard work. Thus children of all ages will be safely taken through new and challenging surgical environments.

Thank you.

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