Management considerations in congenital laryngotracheal stenosis

Congenital laryngotracheal stenosis poses significant management challenges, necessitating multidisciplinary input involving paediatric intensivists, neonatologists and paediatric otolaryngologists. Management decisions and patient outcomes are often compounded by coexisting factors including extreme prematurity, gestational age at time of diagnosis and low birth weight. The pathophysiology, treatment options and management considerations related to this complex pathology are discussed.

Isobel Rothera
MBBS, BSc, MRCS
Core Surgical trainee, Guys and St Thomas' NHS Trust
isobel.rothera@nhs.net

Mira Sadadcharam
FRCS (ORL-HNS), PhD
Consultant Paediatric ENT Surgeon, Royal Manchester Children's Hospital

Archana Mishra
MBBS, MD, MRCPCH
Consultant Neonatologist, Bolton NHS Foundation Trust

Richard J Hewitt
BSc, DOHNS, FRCS (ORL-HNS)
Consultant Paediatric ENT, Head and Neck and Tracheal Surgeon, Great Ormond Street Hospital for Children

Keywords

tracheal stenosis; congenital airway anomalies; bronchoscopy; tracheal surgery; laryngeal surgery; extreme prematurity

Key points

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1. Congenital tracheal anomalies are rare forms of laryngotracheal stenosis.
2. Surgery represents the ultimate form of treatment. Reconstructive techniques depend on the pattern of stenosis.
3. The decision-making process and clinical outcomes are often compounded by co-morbidities.
4. These cases should always be managed in a multidisciplinary setting with a treatment plan tailored to the child.

Case summary

An extremely preterm female infant, first of a set of dichorionic diamniotic twins, was born at 24 weeks' gestation weighing 590g. On the first intubation attempt at birth, the insertion of a size 2.5 endotracheal tube (ETT) was met with resistance at 6cm from the lip and any attempt to advance the ETT beyond this point was unsuccessful. However, ventilation was stable when the ETT was secured at 5.5cm at the lip. Similar difficulties were encountered on different occasions when intubation was attempted and a diagnosis of partial airway obstruction was suspected.

She was referred to the ear, nose and throat team at 26 weeks' corrected gestational age. Flexible bronchoscopy via the ETT revealed a tracheal stenosis immediately distal to the tip of the ETT. It was not possible to pass a 2.2mm bronchoscope into the distal tracheobronchial tree. She subsequently underwent a CT scan that confirmed a tracheal stenosis of 7-8mm in length, with the diameter varying between 1.3mm and 3mm. The scan also demonstrated a normal calibre distal trachea, carina and mainstem bronchi (**FIGURE 1**).

A tertiary paediatric centre in London reviewed the child's case. Due to her extreme prematurity and low birth weight she was initially managed conservatively. She has since been discharged home on room air where she is thriving and completely asymptomatic. Plans for surgical intervention in her case have yet to be finalised.

The principle challenges in this child's case included her extreme prematurity, gestational age at time of diagnosis and low birth weight. The salient points of the complexities of paediatric laryngotracheal stenosis and the therapeutic implications of this in directing management are discussed.

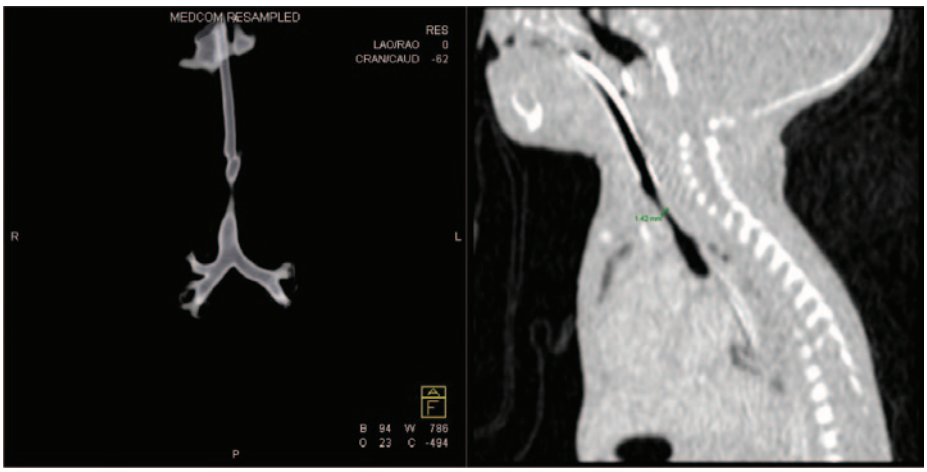

FIGURE 1 CT images showing congenital tracheal stenosis of 7-8mm and a normal distal trachea.

Introduction

Congenital tracheal anomalies are rare, accounting for a scant 0.3-1% of all laryngotracheal stenoses,[1] with an estimated incidence of approximately 1:60,000 live births.[2] Consequently, the published literature on the management of these conditions is both limited and diverse. Traditionally, an understanding of the relevant embryology provides a logical basis for understanding both the pathogenesis and physical/physiologic manifestations of the disease. This in turn allows for a broader knowledge base, facilitating the asking of more sophisticated questions. However, a unifying theory explaining the various structural anomalies of the tracheobronchial tree remains elusive.[3]

Embryology

The first indication of the tracheobronchial system appears in the primitive pharyngeal floor, early in the fourth week of embryonic life, as a longitudinal groove. This groove deepens and, with development, grows caudally below the pharynx to become a diverticulum – the laryngotracheal diverticulum (**FIGURE 2**). As the diverticulum grows, it becomes separated from the pharynx by a partition, the tracheoesophageal septum, which divides the foregut into the laryngotracheal tube anteriorly and the oesophagus posteriorly. This intimate embryologic development of the trachea, oesophagus and cardiovascular systems may account for the incidence of mediastinal anomalies associated with congenital tracheal stenosis (CTS).[4-9]

Classification

As a rule, congenital tracheal malformations may be intrinsic to the trachea itself or may represent external forces compressing the airway (**FIGURE 3**).

Often tracheal stenosis occurs due to complete cartilaginous rings, described as 'O' rings. However, disorganised segments of cartilage or, more rarely, complete cartilaginous segments may occur. In 2004, Grillo introduced a classification for congenital tracheal stenosis (**FIGURE 4**).[10]

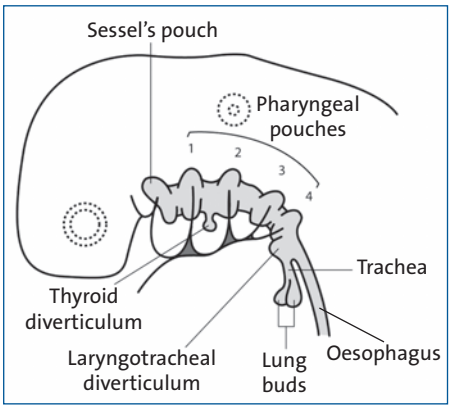

FIGURE 2 A schematic diagram of the laryngotracheal diverticulum – the embryonic precursor of the adult laryngotracheal tree.

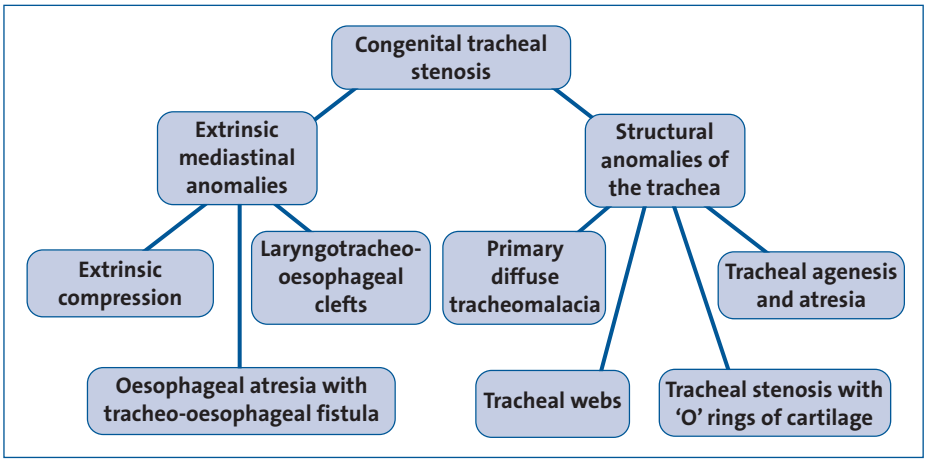

FIGURE 3 The various aetiologies causing congenital tracheal stenosis.

Clinical presentation and evaluation

Regarding the diagnosis of CTS, a high degree of clinical suspicion should be maintained in infants or children presenting with respiratory distress. It is important to bear in mind that although congenital tracheal malformations are, by definition, present at birth, affected children may be asymptomatic until far later in life. This is particularly true as children grow, where an increase in activity results in a corresponding increase in respiratory demand.

Clinical symptoms only arise when the tracheal lumen is compromised by greater than 50% or where there is a tracheo-oesophageal fistula or laryngotracheo-oesophageal clefts. Classically, children present with a biphasic stridor with a considerably prolonged expiratory phase. The inspiratory component of the stridor occurs due to stenosis in the cervical trachea. In contrast, the expiratory component results from stenosis/collapse of the thoracic trachea. Other symptoms include a barking cough, 'washing machine' breathing, and evidence of increased work of breathing, including inter/subcostal recession, nasal flaring, episodes of respiratory distress and apnoeic/cyanotic spells.

The mode of presentation may vary depending on the age of the infant/child.

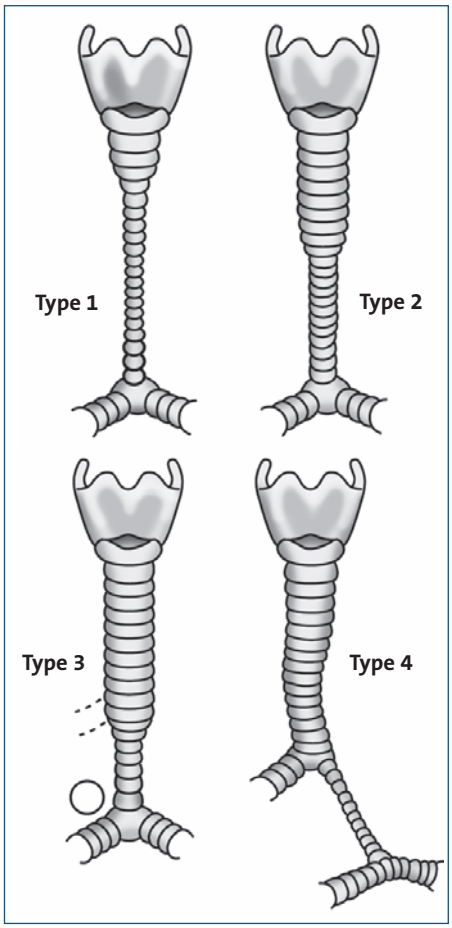

FIGURE 4 Classification of congenital tracheal stenosis due to complete 'O' rings. Type 1 describes a long segment stenosis where all, or the majority, of the trachea is involved with normal morphology noted in only the first to third tracheal rings. Type 2 describes a funnel-shaped stenotic segment, which may vary in length and location. Type 3 refers to a short segmental stenosis, which may occur at variable levels of the trachea, sometimes associated with an anomalous right upper lobe bronchus. Type 4 represents a variation of Type 3 whereby there is an anomalous right upper lobe bronchus near the carina. Here, the stenotic segment connects to the proximal trachea via horizontally branching bronchi.

In infants, hyperextension of the neck may occur in an attempt to maximise the calibre of a compromised airway. A history of failure to thrive and episodes of cyanosis/respiratory distress when feeding should also raise clinical suspicion for an underlying airway anomaly. In older children, a lower respiratory tract infection with resultant respiratory deterioration may result in the diagnosis of a previously undiscovered tracheal anomaly. Children with 'asthma' who fail to respond to treatment should also be suspected of having an underlying airway anomaly. In children with a previous history of ventilation, an organic airway stenosis should be the working diagnosis until proven otherwise.

Radiologic evaluation using CT and/or MRI may offer valuable information in regards to the length of the stenotic segment. However, these are often unable to distinguish between the true luminal diameter and inspissated secretions. The gold standard for evaluating children with laryngotracheal stenosis is direct laryngotracheoscopy.

Management

The last 20 years have seen a steady advance in surgical and endoscopic techniques resulting in dramatically improved patient outcomes. Prior to the advent of current surgical techniques, mortality rates associated with CTS were as high as 79%, mainly due to complications associated with acute airway compromise and lack of effective medical management.[11] Current challenges lie more in reducing the morbidity associated with surgical intervention rather than achieving survival. The management of paediatric laryngotracheal stenosis, whether acquired or congenital is challenging. Other potential coexistent co-morbidities will need to be excluded. The choice of surgical reconstruction will be dependent upon several variables including the location of the stenotic segment, duration, severity, aetiology and the degree of functional impairment. Having established this, not every patient with CTS will require operative intervention and asymptomatic children may be managed expectantly.[12-14]

Endoscopic techniques

Endoscopic treatment options for the management of CTS with complete circular rings are limited. Although previously advocated,[15] balloon dilatation associated with posterior division of the circular cartilaginous rings is dangerous as it entails stenting of an already compromised airway. The use of metal stents previously proposed by Maeda et al[16] has since been deemed unacceptable.[17] Presently, there are several papers in the published literature that appear to testify that stenting and dilatation have no role to play in the management of CTS with complete tracheal rings. This is particularly true considering the existing success rates associated with surgical repair.[18]

Surgical techniques

Advances in both surgical technology and technique have enabled far better management of both congenital and acquired tracheal stenosis. In addition, cardiopulmonary bypass and extracorporeal membrane oxygenation have helped in optimising patient conditions both intra- and post-operatively. Wherever possible, single-stage surgery is the preferred option for reconstruction and coexisting cardiovascular anomalies (such as pulmonary artery slings) should be repaired concurrently. Each case should be processed through a multidisciplinary forum and, regardless of the reconstructive technique selected, vigilant post-operative surveillance for complications is essential.

Presently, there are five open surgical approaches that have been used in the management of CTS with complete tracheal rings.

1. Primary resection with end-to-end anastomosis

This technique is reserved for stenotic segments that do not exceed a third of the tracheal length (**FIGURE 5A**). The technique involved is the same regardless of whether the stenosis is congenital or acquired. For high or mid-segment tracheal stenoses, the entire procedure may be carried out through a horizontal cervical incision, as the paediatric larynx is located high in the neck. For low/intra-thoracic stenotic segments, or in children with coexisting mediastinal anomalies, a sternotomy with cardiopulmonary bypass offers optimal intra-operative conditions. The procedure begins with conventional endotracheal intubation and ventilation. This is followed by midline division and lateral retraction of the strap muscles, thyroid isthmus and thymus. Dissection is limited to the anterolateral surface of the trachea with preservation of the vascular supply arising from both trachea-oesophageal grooves. A stay suture is passed distal to the stenotic segment and an inferiorly-placed transverse incision is made between the tracheal rings. At this stage, a second ETT is used to intubate the distal tracheal stump (**FIGURE 5B**). The tube is temporarily sutured into position and connected to a second sterile ventilation circuit. A proximally-placed transverse incision is then made. The stenotic segment is then mobilised and resected. The proximal and distal tracheal stumps are primarily reapproximated. Once reapproximation is nearly complete, the ETT in the distal tracheal stump is removed and the original ETT is advanced distally into the tracheal stump (**FIGURE 5C**). An airtight seal is confirmed by flooding the field with sterile saline and checking for an air leak. Drain insertion and wound closure are then carried out in the conventional fashion.

2. Slide tracheoplasty

This technique, originally described in 1989[19] has emerged as one of the best surgical options for reconstructing long-segment congenital tracheal stenosis (LSCTS).[20-24] The objective of a slide tracheoplasty is to achieve doubling of the tracheal circumference at the stenotic segment. Surgery begins by placing a horizontal incision at the midpoint of the stenotic segment followed by vertical slits sited both anteriorly and posteriorly over the upper and lower stenotic segments (**FIGURE 5D**). Gentle traction is then applied to slide the upper and lower segments over one another (**FIGURE 5E**). An oblique anastomosis is fashioned (**FIGURE 5F**) resulting in a trachea roughly half its original length with a four-fold increase in diameter.

3. Anterior patch tracheoplasty

This technique is suitable for reconstruction of LSCTS and involves a midline sternotomy. A midline vertical incision is made over the anterior wall of the stenotic tracheal segment (**FIGURE 5G**). Airway control and maintenance of ventilation is achieved by advancing the ETT distally through the now open stenotic segment down to the carina. The ETT also serves to act as a stent allowing for assessment of the length and width of graft to allow for tracheoplasty. Costal cartilage is then harvested, shaped to the necessary dimensions (**FIGURE 5H**) and secured in place using interrupted sutures with the

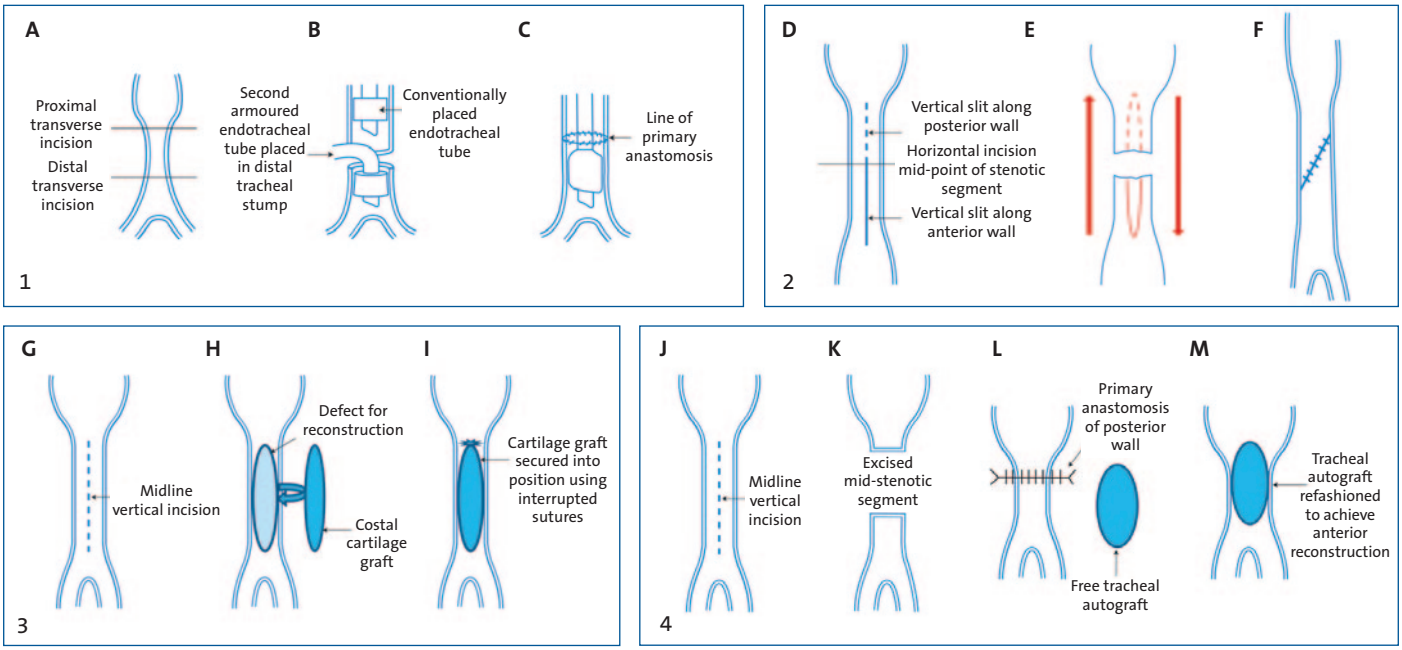

FIGURE 5 Schematic diagrams representing: (1) Primary resection and end-to-end anastomosis for short segment tracheal stenosis. (2) The technique involved in slide tracheoplasty. (3) The anterior tracheoplasty technique used for reconstructing long segment tracheal stenosis. (4) The stages involved in tracheal autografting.

perichondrial surface facing internally to the lumen of the trachea (**FIGURE 5I**).

4. Tracheal autograft

Similar to the anterior patch tracheoplasty, this procedure begins with a midline vertical incision made over the entire length of the stenotic segment (**FIGURE 5J**). The narrowest central portion of the stenosis is then resected (**FIGURE 5K**). It is important that the excised segment does not exceed 30% of the tracheal length. This excised portion can then be refashioned into a free tracheal autograft. The excised tracheal stumps are primarily anastomosed posteriorly (**FIGURE 5L**) and the tracheal autograft can then be reconfigured to reconstruct the residual defect (**FIGURE 5M**).

5. Cadaveric tracheal homografts

This reconstructive technique is indicated for children with recurrent tracheal stenosis. It is not indicated for primary repair.[25]

Conclusion

The management of paediatric laryngotracheal stenosis continues to pose significant challenges to the otolaryngologist. Classically, surgery represents the ultimate form of treatment for this anomaly, and techniques of repair have evolved over time. The principle challenges in managing this child's case included her extreme prematurity, gestational age at time of diagnosis and low birth weight. This is particularly relevant when considering the significant morbidity associated with surgical intervention demonstrated by prolonged intubation times, intensive care requirements, inpatient stay, and the need for repeated surveillance bronchoscopies, stenting and possibly revision surgery.

The common denominator for nearly all post-operative morbidity is the formation of excessive granulation tissue at the repair site, resulting in varying degrees of airway obstruction, failure to clear secretions, atelectasis and respiratory distress. Again, this is particularly relevant in the context of preterm neonates with pre-existing reduced respiratory reserves. Additionally, grafts may get infected or suffer from necrosis or dehiscence, with the potential for air leak and mediastinal emphysema requiring revision surgery.

Due to the significant variability and not infrequent co-morbidity profile, these cases should always be managed in a multidisciplinary setting. Treatment plans should be individualised to each child, taking into consideration the site, severity, underlying aetiology, and the extent to which the child is symptomatic.

Close follow-up with bronchoscopy is mandatory and interventional procedures such as granulation tissue debridement, balloon dilation or intraluminal stenting should be available as required.

Future advances may evolve around optimising tracheal healing and meticulous control of inflammation, fibrosis and granulation tissue formation around the surgical repair site. Vascular endothelial growth factor and tissue engineering may provide major breakthroughs at a cellular level, improving clinical results and postoperative care following surgical repair of CTS.

Patient consent

The authors received written consent to publish this report from the patient's parents.

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Book review

Guidelines for Perinatal Care, eighth edition

Edited by Sarah Kilpatrick, Lu-Ann Papile, George Macones

American Academy of Pediatrics and The American College of Obstetricians and Gynecologists

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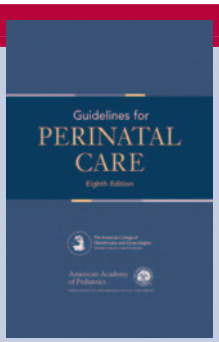

As obstetric and neonatal care becomes increasingly protocol driven, it is essential for clinicians to stay up-to-date with the most recent evidence to aid decision making. Jointly developed by the American Academy of Pediatrics and the American College of Obstetricians and Gynecologists, *Guidelines for Perinatal Care* offers a wide range of advice and reference material on perinatal care from obstetric and paediatric viewpoints.

It is rare to find a book that covers such a large array of topics: non-clinical aspects (eg communications skills and quality improvement) are discussed alongside the more clinical topics (eg nutritional requirements of the newborn infant, hypoglycaemia and hyperbilirubinaemia). Chapters cover organisational aspects such as public health, family planning and patient safety, through to suggested recommendations for perinatal infections (eg hepatitis), neonatal resuscitation and neonatal complications. There is a tendency for short summaries of each topic rather than an in-depth review, however there is a comprehensive bibliography and reference list at the end of each chapter for signposting sources of information and further reading.

This text is mainly aimed at those providing obstetric care and looking after newborn infants. As a neonatal doctor I have found it valuable in terms of understanding obstetric decision-making and also for making treatment decisions for the newborn babies under my care. As the book is endorsed by American institutes it was interesting to read about the differences in healthcare provision and systems between the US and the UK, although when referring to the guidance it is important to ensure that it doesn't contradict national and local advice, which may well differ.

Guidelines for Perinatal Care is probably intended as a quick-look manual rather than a book to sit down and ponder over. That said, it is probably too large to carry around the ward. As a paediatrician, I have found the book useful for adding to my knowledge of antenatal and postnatal management of routine complications during pregnancy, and particularly valuable when counselling expectant mothers. It is a book I will keep referring back to. It covers such a wide range of topics that I would recommend it for delivery suite, postnatal ward and neonatal unit staff.

Stefan Zalewski
Neonatal Higher Specialist Trainee
Northern Deanery