# Continual Professional Development. The developmental anatomy and physiology of the respiratory system.

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## Abstract

This article will review the embryology and the developmental anatomy of the respiratory system. It will provide an overview of the respiratory assessment process of the individual from birth to adolescence. The article will consider a range of the more common respiratory system congenital defects and disorders of the respiratory tract. An overview of care pathways will be provided as there is a focus on reducing the number of hospital admissions.

## Introduction

The respiratory system is a complex and diverse system and there are many conditions and diseases which can affect this system in the child. This article focuses on common conditions mainly with an underlying congenital origin.

The aim of this article is to provide the reader with an introduction to the embryology and the developmental anatomy of the respiratory system.

After reading this article, completing time out activities and visiting associated links the reader will be able to:

* Outline the embryology of the respiratory system
* Describe the components of a respiratory assessment and appreciate the importance of some basic observations.
* Describe some of the respiratory congenital problems.
* Reflect upon how the pathways of care for children with respiratory conditions have shifted from the hospital to the community.

## Embryogenesis and developmental A&P

The respiratory system does not carry out a physiological function until after birth, so it develops relatively late in embryological terms. Lung development involves several stages of development (see table 1). If an infant is born before 22 weeks gestation the lungs are not sufficiently mature to support life. The current age of viability in the UK is regarded as 24 weeks gestation. Gestation is a rather crude calculation as it is counted from the first day of the last menstrual period. Increasingly infants on the borderline of viability can be supported to survive, however the mechanisms used to support the life of the extremely premature can also have a lasting consequence for the individual and their family (O’Reilly 2018, NICE 2019a) there is a strong link between chronic lung disease and high oxygen requirements and positive pressure ventilation for example (Ambalavanan and Rosenkrantz 2014).

### Table 1 Development of the respiratory system before birth.

|  |  |
| --- | --- |
| Timeframe and stage  | Structural and tissue organisation |
| Post fertilization to week 16/17.Embryonic and pseudoglandular stage.  | The early cellular and tissue specialisation which will form the respiratory system initially emerges from an upper portion of the primitive gut, which is a tubular structure which extends from one end of the embryo to the other. From the head end of this tubular gut structure the respiratory tract buds emerge, see diagram 1 these form the respiratory diverticulum (Sharma and Kapoor 2018). Subdivisions of the diverticulum will become the right and left bronchial buds, see diagram 1. Failed or incomplete separation of these tubular structures, destined to become the trachea from the foregut will lead to the development of a congenital condition, such as tracheoesophageal fistula and /or oesophageal atresia (Sharma and Kapoor 2018).The bronchial buds will elongate and subdivide to become the main bronchi, bronchioles and with tissue specialisation will become the functional lung units. The right bronchial bud will form three main bronchi and three lobes, while the left will form two main bronchi and two lobes see diagram 1. During the pseudoglandular phase, the lungs resemble a gland, see diagram 2. At the end of this phase the precursors of the pneumocytes can be seen as cuboidal epithelium this produces secretions which contribute to the amniotic fluid. The thoracic cavity is separated from the abdominal cavity by the formation of the diaphragm (weeks 8-10). Failure of this process will lead to a congenital diaphragmatic hernia and possible lung hypoplasia (Chin and Sharma 2017).   |
| Canalicular weeks 17 to 24/25 | This stage involves dramatic changes in pulmonary epithelium. This stage includes the continued development of the respiratory bronchioles and each end in a small dilated bulge. These small enlargements will become the primitive alveoli and host specialist pneumocytes. Further refinement and maturation of these cells will result in the gossamer thin membranes through which gas will be exchanged. Type 2 alveolar cells will eventually produce surfactant and line the primitive alveoli.  |
| Saccular period weeks 24 to 36 | This stage includes the development of the elastic fibres which provide support to the structures of the respiratory tract. The phase is defined by the appearance of defined airspaces which appear at the end of each respiratory tract passage (sacculi). The sacculi become generously supplied with capillaries, both vital for gas exchange. The alveoli are immature until quite late in pregnancy, but their action can be matured artificially by the administration of steroids to the mother to accelerate the production of surfactant (NICE 2016a).  |
| Alveolar stage Weeks 36 to term | The alveolar period occurs from week 36 onwards, this is the final refinement of the developing alveoli before birth and the transition from foetal circulation to respiration, see diagram 2. The cells present are: Type one cells – cover the alveolar surface area important for gas exchange.Type two cells – produces surfactant.One function of surfactant is to lower surface tension in the alveoli. It also stabilises the alveolus and prevents alveolar collapse at the end of exhalation. By keeping the alveoli expanded the work of breathing is easier. Surfactant also contributes to the innate defence system and has been shown to possess anti-inflammatory properties. |

Crawford (2011a) Dixon and Crawford (2012) Sharma and Kapoor (2018), Chin and Sharma (2017), Respiratory System Development (see links) Embryology (see links)

The lungs and the structure of the airways continue to develop after birth to meet the oxygenation demands of the growing and active child and reflect the elongation of the neck. Shape and size of the mouth changes and increases reflecting the commencement of a solid diet and the modulation of speech. The vital capacity of the lungs increases, and this allows the respiratory rate to slow down as gas can be exchanged more efficiently. The chest wall becomes more stable with increasing rib ossification and musculature and this increasing maturity. This growth and maturation of the lungs and airways can be related to the changing signs of respiratory distress. Rib retraction and sternal tug are signs more commonly seen in infants because of the compliant chest wall. They are not seen in older children, because the chest wall is more stable however as there is more muscle in the airways an asthmatic trigger for example can result in bronchospasm, narrowing of the airways, creating airway turbulence and an audible wheeze. A wheeze is not common in infants but is a common symptom of Asthma which emerges in older children. See tables 2-4

### Table 2 development of the respiratory system after birth to completion.



Crawford (2011a) Dixon and Crawford (2012) Auerback and Nimavat (2016) Sharma and Kapoor (2018), Edwards (2018),

Diagram 1



Diagram 2



Note to editing team these diagrams will need to be redrawn.

**Time out 1**

List the differences between the airways of the infant and the child. Why do you think that these distinctions might be important as you discuss a child’s health problems with a parent?

### Airway and Respiratory assessment

Regardless of the health care setting, an airway and respiratory assessment is a priority in understanding the physical status of the child. Some information can be gathered as you approach the child and it is important to keep the child calm. Whenever possible consent should be obtained.

There are a range of tools which can be used to support this assessment and most systems follow a logical sequence of observation and assessments. A score on its own will do nothing, however the use of a tool improves situational awareness and the use of a tool has been demonstrated to reduce incidents of emergency collapse. Scotland has had a range of 5 age appropriate early warning tools in place since 2017 (see links). The charts require the nurse to know the normal vital signs parameters, detect and record deviations and score the child accordingly. If used reliably it can alter the frequency of observations, bring more expertise to the bedside and  allow the care plan to be revised (NHS Scotland 2017). England is in the process of agreeing to agree to a National system which will be in place sometime during 2020.

## Time out 2

Find out what early warning tools are currently used in your local area. Are the same tools used in both the community and acute setting and how are nurses trained to use them?

### Airway

Assessment of the airway can be difficult when a child is anxious and distressed. An infant who can cry or vocalise is providing an indication that their airway is patent and clear. Challenges to airway patency, include symptoms which can lead to obstruction such as drooling, stridor, secretions, or snoring. Assessment of the airway is a continuous observation, if there are any concerns with regards to airway patency these must be addressed immediately.

### Breathing and respiratory assessment

The assessment should include an assessment of the child’s respiratory pattern and rate and the heart rate (see tables 2 and 3). These measurements should be taken over a full minute because irregular breathing can influence the count. The child should be unaware that the respiratory rate is being counted as knowing that this was happening could affect the rate. The respiratory rate in children is variable depending on age, size and mental or physical state.

### Table 3 Respiratory and heart rates by age

|  |  |  |
| --- | --- | --- |
| Age of child  | Respiratory rate (range) | Awake heart rate.Please note that an elevated heart rate is a strong indication of a stress state  |
| Newborn and neonate Infant under 1 year 1–2 years 2–5 years 5–12 years > 12 years  | < 60 breaths/min30–40 breaths/min25–35 breaths/min25–30 breaths/min20–25 breaths/min15–20 breaths/min | Term newborn 100-160100 - 150 100-150 95-140 80-120 60-100  |

Modified from Dixon and Crawford (2012) Paediatric Guidelines 2013-2014, Fleming, Thompson, Stevens et al (2011) O'Leary, Hayen, Lockie (2015) RCN (2017)

Having established the respiratory rate, any unusual characteristics of the respiratory pattern should be noted (see table 4).

### Table 4 Terminology and descriptors of abnormal respiratory rates and/or patterns. All changed patterns of breathing require medical assessment and regular monitoring, some children may require urgent transfer to high dependency care facilities\*.

|  |  |
| --- | --- |
| Term used  | Description, implications for the child and nursing actions. |
| Tachypnoea  | Breathing faster than the usual respiratory rate for the age of the child. The smaller younger infant can exhaust quickly and be unable to sustain such a rapid rate for long. Consequently, some of these infants / children are at risk of decompensation and collapse\*. |
| Bradypnoea  | Breathing slower than the usual rate for the age of the child. The fit young athlete may have this as a normal finding, however if present in infants/ children they are potentially at risk.  |
| Intermittent apnoea \* | Identified by periodic absence of breathing, this is common in premature infants but is outgrown and resolved before discharge. A reoccurrence may indicate additional underlying pathology and a risk of imminent collapse (Rocker and Bechtel 2018).  |
| Hyperpnoea  | Breathing which is faster and deeper than is usual for a child of their age. Is more common when a child is in pain, has a fever, or is distressed. There is a risk that the level of CO2 in the blood could fall and the child will experience dizziness and pins and needles. |
| Dyspnoea  | This is a marked degree of breathlessness and difficult, laboured respiration it may be associated with the child not being able to vocalise or finish a sentence.  |
| Orthopnoea  | Dyspnoea when lying supine due to the way this affects the respiratory function. Older children will find a position of comfort and be able to support their respiratory function using their accessory muscles, small infants may benefit from being turned prone with a 35 degree, head up tilt, but must never be left alone in this position.  |
| Hypoventilation \* | Slow, shallow breathing generating very small tidal volumes. These children require transfer to high dependency care facilities.  |
| Sighing breaths | Frequently interspersed deeper breaths. These children need to be assessed by medical personnel.  |
| Air trapping  | Increasing difficulty in getting air out during expiration this is usually associated with the presence of a wheeze. If the Asthmatic child has a care plan in place nebulizers can be offered and the child’s condition continually reviewed.  |
| Kussmaul respirations (also known as acidotic breathing) \* | Rapid, deep breaths with an additional effort at the end of expiration; this pattern is associated with the presence of a metabolic acidosis. These children require urgent transfer to high dependency care facilities . |

Dixon and Crawford (2012) Springer and Corden (2016) Rocker and Bechtel (2018). To hear a range of breath sounds, see links.

The child’s posture, position and movement of the shoulders and chest may reveal the use of accessory muscles, as the child struggles to breathe and draw as much air into the lungs as possible. Infants may have nasal flare and be seen to head bob as the scalene and sternocleidomastoid muscles contract with the effort.

The altered intrathoracic pressures can cause a range of recession movements such as intercostal recession, which is indrawing of the chest wall, as infants and young children have a more cartilaginous thoracic cage and the chest wall is more compliant (see table 2). Where there has been trauma, the thoracic recessions may not be symmetrical and the nurse conducting the assessment should be alert for paradoxical (see-saw) chest wall movement. With a history of fall, crush or trauma diminished breath sounds over one side of the chest may mean a pneumothorax and the child needs emergency care (Crawford 2011b). Following a full systematic respiratory assessment, the nurse will progress to circulation.

### Circulation, colour and cough

The child in respiratory distress or under the pressure of a clinical examination is likely to have a higher than normal heart rate (see table 2) for normal values. The blood pressure (BP) may be elevated if the child is under stress but a lower than expected BP and poor perfusion may indicate the child is failing to maintain their circulatory homeostasis and have reached the limits of the compensatory mechanisms, they could become very ill, very quickly and in need of critical care (Crawford 2018b). An assessment of the capillary refill time must be performed and can be done quickly while making other observations and making decisions as to what support this child might need. The refill time should be less than 2 seconds (Thim, Krarup, Grove et al 2012).

Documenting the general colour of the child is important as peripheral or central cyanosis will mean that the child needs to be transferred to the appropriate health care setting as a matter of urgency. Cyanosis can be described as the presence of blue or purple colour of the skin, mucous membranes or nail beds due to the tissues being depleted of oxygen. In light-skinned patients, cyanosis presents as a dark bluish tint to the skin and mucous membranes (which reflects the bluish tint of unoxygenated haemoglobin). In dark-skinned patients, cyanosis may present as grey or whitish (not bluish) skin around the mouth, and the conjunctivae may appear grey or bluish. In patients with yellow/tanned skin tones, cyanosis may cause a greyish-greenish skin tone (Sommers 2011).

A cough is an important physiological protective reflex which prevents secretions, inhaled or aspirated material from entering the airways it is also a symptom of respiratory disease it may be acute or chronic of 8 weeks or longer duration (Halpel and Meyers 2018) and the characteristics of the cough should be documented. A cough may be moist, productive or dry. The nature of the secretions or sputum produced should be examined. Blood stained secretions whether fresh or rusty, liquid or clotted are always abnormal and a finding of haemoptysis should always result in an urgent referral. Haemoptysis may be indicative of tuberculosis (Herchline and Bronze 2018). Copious clear or white secretions may indicate a viral cause. Yellow or green secretions may indicate a bacterial infection (Healthline 2017). The texture of secretions should also be considered stringy mucous which the infants struggle to clear is common in bronchiolitis (NICE 2015). Thick mucopurulent sticky globules can indicate severe infections.

The frequency of the occurrence of cough and the duration of the spasm is important, a paroxysmal exhausting bout of coughing ending in a whoop as a frantic inhaled breath is made, is typical of pertussis (Bocka and Steele 2018). Any obvious triggers to the cough is important as this can help pin down allergy or sensitivity. The child or the parents should also be asked if the cough disturbs sleep.

The management of some types of cough has recently come under scrutiny with (NICE 2019) setting out an antimicrobial prescribing strategy for acute cough associated with an upper respiratory tract infection or acute bronchitis in adults, young people and children. It aims to limit antibiotic use and reduce antibiotic resistance (NICE 2019b).

### Additional considerations when assessing a neonate

Discoloration such as cyanosis of hands and feet (acrocyanosis) is common in the newborn and may persist for several days with altitude or in a cool environment, it is still not well understood (Luo 2018) is thought to be caused by the constriction of small blood vessels that decrease the flow of oxygenated blood to the extremities. There is controversy as to whether infants are obligatory nasal breathers or not, and the narrow airways and rapid respiratory rate can result in infants snoring and appearing to breath heavily (Gill 2018) this can be a great distress to parents. Nasal flaring without other signs of respiratory distress can also be a common finding and is of no clinical significance. However, infants who have been born prematurely are more likely to have a greater incidence of breathing irregularity when compromised, any infant with a history of apnoea will need transferring urgently to an appropriate care setting. Coughing in infants is rare and usually indicative of a more serious problem (Bocka and Steele 2019), whereas sneezing is frequent and can occur without underlying pathology. Hiccups in new-borns are common, usually they are silent and associated with feeds, frequent non-feed-associated hiccups may be suggestive of additional pathology (Sheth and Nelson 2019).

### Other observations of note

Finger clubbing is the swelling of the distal portion of the digits, which may be bilateral or unilateral, or occasionally may involve a single digit (Schwartz and James 2018). It can be caused by a range of pathologies but is often indicative of chronic hypoxaemia (Schwartz and James 2018) and the child will most likely have a long-standing condition such as cystic fibrosis. The size and shape of the child’s chest the presence of any skeletal abnormalities such as a scoliosis may affect the child’s respiratory performance (Mehlman and Goldstein 2018) also the presence of any scars might suggest a previous surgical intervention such as a thoracotomy or the use of chest drains.

Other factors to note while assessing children is the general nutritional status of the child. This is important as obese children may get breathless more easily on activity and children with large amounts of fatty tissue adds weight to the thorax and could compromise the child’s ease of respiration if in a recumbent position. Obese children are more likely to suffer asthma and sleep apnoea (Dixon and Crawford 2012, Childhood Obesity Foundation 2015).

Additional respiratory assessment techniques that should be applied more selectively.

## Palpation Percussion and Auscultation.

Palpation is a useful tool; it is a method of feeling with the fingers or hands during a physical examination (Lone and Mosenifar 2019). Often nurses can do this discretely when they pick up a child and they can feel the secretions rattling within the chest cavity. Palpation by a stranger could be stressful to an older child and make the respiratory distress worse, see table 5.

### Table 5 using Palpation Percussion and Auscultation

|  |  |
| --- | --- |
| Technique  | Findings  |
| Palpation | Normal palpation should reveal bilateral symmetry of movement of the chest wall, with a degree of elasticity of the rib cage. The sternum should be relatively inflexible; however considerable care should be given to applying any pressure to the chest wall of a child. Crepitus is a crackly/crinkly sensation which can be both felt and heard. This is indicative of air in the subcutaneous tissues, possibly from an air leak (Lone and Mosenifar 2019). A pleural friction rub is a palpable, coarse, grating vibration, felt on both inspiration and expiration, which may be compared to the feeling of rubbing suede leather. It may be caused by inflammation of the pleural surfaces. Other findings can include abnormal pulsations, areas of pain or tenderness. The presence of bulges and depressions. |
| Percussion | Often underused during a nursing assessment, it can detect abnormal sounds and assess whether underlying tissue is air-filled, fluid-filled or solid (Lone and Mosenifar 2019). The skill requires careful interpretation as the round shape of the infant’s chest causes a hyper-resonant percussive note, which is abnormal in the older child. By 6 years normal percussive sounds should have a resonant pitch; therefore, hyper-resonance in this age group is indicative of underlying pathology such as pneumothorax.  |
| Auscultation | Involves listening to sounds in the chest which are produced by the movement of gas or liquid within the chest cavity, to aid diagnosis (Lone and Mosenifar 2019). When listening to a child’s chest sometimes the sounds are loud and obvious such as stridor or wheeze. Others such as fine crackles can only be heard using a stethoscope (Jevon and Cunnington 2006). |

A Chest X-ray (CXR) can identify a range of conditions such as infection, inhaled foreign body, neoplasms or tuberculosis but interpretation can be difficult (Morgan, Pettet, Reed et al 2018) and time should not be wasted trying to get the best quality image or try to distinguish between pathologies (Morgan, Pettet, Reed et al 2018) if the child is sick or have issues which raise concern on an X ray, they need more expert assessment and they should be referred on.

## Respiratory conditions

Respiratory conditions can be considered under structural anomalies which affect the pulmonary mechanics, obstructive conditions which impacts on the flow of gas and conditions which impair /impede gas exchange, although there may be some overlap. For example, Cystic fibrosis an inherited condition causes changes in the cellular function and alters the characteristics of the secretions they produce, the resulting build-up of secretions can impede pulmonary and digestive functions. Another example is Asthma where there are genetic influences but triggers such as atmospheric pollutants causes airway reactivity and impacts on gas flow and exchange.

### Congenital conditions

Include conditions where there are structural anomalies which result in pulmonary mechanical problems. Invariably, they will require a surgical referral and transfer for correction. They include Tracheoesophageal Fistula (TOF) and Diaphragmatic Hernia (DH).

### Common conditions of the premature and the young infant

Pre-term birth is the biggest cause of neonatal mortality and morbidity. Increasingly survival of the extremely premature infant <26 weeks (gestation) contributes more significantly to the infant morbidity rather than the mortality rate. It can be expected that in the UK rates of survival will increase owing to the confidence and willingness to treat infants born at lower gestational ages.

One of the consequences of prematurity is Chronic Lung Disease (CLD) Bronchopulmonary Dysplasia (British Lung Foundation 2019, NICE 2019). This can be defined as continued oxygen dependency at 36 weeks gestation. It is characterised by the need for prolonged respiratory support, the infants displays compromised lung function and the suffer recurrent respiratory infections during their first year of life. Infants who have CLD may also have an associated risk of neurodevelopmental impairment. Owing to the continued potential for lung development until the age of 7 or 8 years these children are comprehensively and aggressively managed in the expectation that their lung function can improve. However sometimes impairment of the lung function persists into young adulthood, with impaired exercise tolerance reduced vital capacity and a degree of airway hyper-reactivity.

### Bronchiolitis

 Bronchiolitis is the result of viral inflammation of the bronchioles, with some groups such as ex pre-term infants being more at risk than others. In children of less than 2 years of age it is characterised by tachypnoea, sternal and rib retraction and wheezing (NICE 2015). Respiratory syncytial virus (RSV) infection is the most important cause of bronchiolitis and it is one of the major causes of hospital admissions in infants under 1 year of age. The history of the infection commonly involves a few days of being unwell with rhinorrhoea, a cough, and a low-grade fever. The condition does not improve, and the mild symptoms are followed by a deterioration made worse by poor feeding and vomiting. As there is no medication which can eliminate the virus treatment is supportive, whether the child is in the home or hospital. If at home small feeds can be offered frequently, to keep the child hydrated, paracetamol can be given to manage fever and the infant kept comfortable and nursed upright (NHS 2018) most cases can get better without the need for a hospital admission but as it can last two – three weeks the parents are usually very anxious and need a lot of community support.

If admitted to hospital the infant may be isolated or nursed in a cohort with other bronchiolitis patients to contain infection. Some may benefit from intravenous fluids to re-hydrate, tube feeding and the provision of humidified oxygen. This would depend on the infant’s oxygen saturations and blood gas results. Intensive care admissions for bronchiolitis are high and some will need the support of Continuous Positive Airways Pressure (CPAP) this is a type of non-invasive ventilation. Others may go on to require invasive mechanical ventilation. There are occasional fatalities with the infection, usually in infants who have co-existing conditions such as Chronic Lung Disease or a congenital cardiac lesion.

#### Croup / epiglottitis

This can be an extremely serious disease (Defendi and Steele 2018). Croup is usually caused by a virus infection, most often a parainfluenza virus; initially it may resemble a common cold with symptoms of fever, sore throat and rhinorrhoea. The infants and children who are most at risk are between 6 months and 3 years of age. The peak incidence is around two years old. Most cases of croup are mild however some cases can involve extensive swelling of the airway structures of the epiglottis, cricoid and upper trachea which can then narrow to such an extent as to interfere with breathing. The signs and symptoms reflect this.

Hoarse cough and husky voice

Secretions

Stridor

Tachypnoea

Dyspnoea

Mild Croup can be managed at home with fluids, paracetamol comfort and rest. Children admitted to hospital may be managed with corticosteroids, nebulised adrenaline and oxygen (Defendi and Steele 2018). A few cases may need to be electively intubated and ventilated to sustain the airway and support the breathing. Croup is a self-limiting disease recovery is usually rapid.

Epiglottitis is an inflammation of structures above the glottis usually caused by bacterial infection. Before widespread Haemophilus influenzae type b (Hib) vaccination, H influenzae caused almost all paediatric cases of epiglottitis. The epiglottis becomes swollen to such an extent that the airway can become compromised. The peak age is between 2-4 years of age. It can be a serious medical emergency (Udeani and Steele 2016) because in a few cases swallowing becomes so difficult, they drool rather than swallow, drooling occurs in about 80% of cases (Udeani and Steele 2016). Some children with epiglottitis may appear to be shocked and if epiglottis is suspected the main strategy of management is securing the airway as soon as possible and an experienced clinician will intubate, if necessary, a percutaneous transtracheal incision may be performed while preparations for a tracheostomy are made (Udeani and Steele 2016). These children will need to be ventilated and transferred to intensive care.

#### Asthma

Asthma is a chronic inflammation of the airways associated with widespread, variable narrowing of the airways and severe airflow obstruction during an attack, see diagram 4.

Diagram 4

 

This will need to be redrawn would suggest the title be asthma and the impact on the airway.

Asthma is now the most common respiratory disorder of childhood (Pinfield, Gaskin Bentley et al 2015, Tidy, 2014). Asthma is more common in boys than girls until adolescence after that the incidence of asthma in young women increases while asthma with symptoms requiring anti-asthma medication in young men declines (Pinfield et al 2015).

Asthma is a complex condition and the links with genetics are as yet unclear, there are a range of genetic research projects underway such as the 100,000 genomes project (https://www.genomicsengland.co.uk/about-genomics-england/the-100000-genomes-project) which will eventually result in better understanding, treatments and perhaps for some a cure. The triggers that cause Asthma are multifactorial they include environmental pollutants, such as cigarette smoke, nitrous oxides, nitrogen dioxide, sulphur dioxide, black carbon and particulate matter, can produce the airway changes that lead to hypersensitivity of the airways and this would appear more likely in the first three years of life (Pinfield et al 2015, Clark, Demes, Karr et al 2010). Infection would also appear to influence the risk of developing asthma with some arguing that early exposure to inflections and animals reduce the risk of developing the condition (Pinfield et al 2015, Global Asthma Network 2014 and Lenney 2009). Asthma could be regarded as an immune inflammatory response condition where the normally protective and beneficial inflammatory reactions start to occur in the airways when there is no need for them to react (Crawford 2011) and in common with other immune response mechanisms some element of inherited transmission is implicated. Many genes have been associated with asthma or asthma-related traits such as allergy and high concentrations of immunoglobulin E (IgE) in serum.  The pathology and the management of asthma beyond the scope of this CPD but the reader could see BTS/SIGN (2019), NICE (2017b) for supporting information.

#### Cystic Fibrosis.

Cystic fibrosis (CF) is an autosomal recessive genetic condition affecting more than 10,400 people in the UK. The CF gene codes for a protein transmembrane conductance regulator (CFTR) that functions as a chloride channel and is regulated by cyclic adenosine monophosphate (cAMP). Mutations in the CFTR gene result in abnormalities of cAMP-regulated chloride transport across epithelial cells on mucosal surfaces. The defective regulation of chloride and the increased reabsorption of sodium and water across epithelial cells decreases the hydration of mucus and results in mucus that is sticky and can harbour bacteria (Sharma and Haver 2018). These sticky secretions in the respiratory tract, pancreas, GI tract, sweat glands and other exocrine tissues have increased viscosity, which are difficult to clear, and the bacteria promotes infection and inflammation. Consequently, CF is a disease affecting the exocrine glands function and involves many organ systems (Sharma and Haver 2018).

In the UK, most cases of cystic fibrosis are picked up at birth using the newborn screening heel prick test (NHS 2018). Although there is no cure for cystic fibrosis, there are an improving and evolving range of treatments which help control the symptoms, prevent or reduce complications, and make the condition easier to live with (NHS 2018, NICE 2018). Typical treatments include physiotherapy, antibiotics, mucous thinners and enzyme replacements (Cystic Fibrosis Trust see links). Lung transplantation is now an accepted treatment for end stage management when other management strategies have been exhausted (Kourliouros, Hogg, Mehew et al 2018) however the demand for lung transplantation has vastly exceeded the availability of donor organs. This has translated into long waiting times and high waiting list mortality (Kourliouros et al 2018). Although this might improve with the recent legislation which supports opt out consent to donation rather than opt in and hopefully this will further improved CF life expectancy. Using the most recent report, the median predicted survival, using the 2013-2017 data, is 47 years old. There is a gender difference as the median predicted survival for females is 43.1 years and this is 6.5 years lower than males at 49.6 years (Cystic Fibrosis Trust, 2017 report - see link). The pathology and the management of CF is beyond the scope of this CPD but the reader could see NICE (2017c, 2018)

## The care pathway of a child with a respiratory health condition.

Children with respiratory conditions have greatly benefited from a change in the culture of care away from hospital-based care to community and home-based care and management. In the past it was not unusual for infants who had been born prematurely and who were oxygen dependant to have their first birthday in hospital and it was common for children living with cystic fibrosis to spend long periods of time on wards having nothing more than IV antibiotics. In part this is owing to an influential charity which champions a manifesto for change to ensure that all children and young people who are living with a serious illness or who have complex care needs to have the support they need to enable them to make the journey home from hospital as quickly and as efficiently as possible (WellChild see link). This has enhanced the quality of life for children and their family. At the same time there has been an evolution with a shift from medically dominated are to care lead by specialist nurses (Coombes 2008) allowing for a more holistic approach involving the wider MDT rather than compartmentalised care. This all makes the future of respiratory care attractive and exciting. Arguably this approach towards advanced nursing could be expediated to the benefit of children and their families much quicker if there was more commitment from the nursing regulator (King, Tod and Sanders 2017). However, all this optimism for the future has to be guarded and balanced when set alongside worldwide microbial resistance (NICE 2016b) which could make TB, chest infections and cystic fibrosis much more difficult to manage in the future.

## Conclusions

This CPD has considered the embryology, development and basic anatomy of the respiratory system. The article has reviewed some of the respiratory signs and symptoms which may be seen, and which should form part of a respiratory assessment. A few of the more common conditions that the children’s nurse could encounter in the community or in the hospital have been provided as clinical examples and the change in the respiratory care pathway has been offered for reflection and debate.

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Cystic Fibrosis Trust <https://www.cysticfibrosis.org.uk/what-is-cystic-fibrosis> accessed April 2019

Embryological development of the respiratory system. Learn more about embryology <https://embryology.med.unsw.edu.au/embryology/index.php/Respiratory_System_Development>

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Embryology. http://www.embryology.ch/indexen.html

European Lung (the white book) http://www.erswhitebook.org/chapters/

Easy Auscultation Lung sounds <https://www.easyauscultation.com/lung-sounds> accessed April 2019.

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