Children's Respiratory Nursing

EDITED BY **JANICE MIGHTEN**

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Foreword

I am delighted to write the foreword to this excellent book. It is a real honour for me as many of the authors work within the senior nursing and medical team at the Nottingham Children's Hospital. I know that nurses regularly working within this area of practice with children and young people will welcome this textbook which discusses all aspects of respiratory care. It will also be an *aide mémoire* for nurse specialists, doctors and allied health professionals, and will be a standard reference for nursing students. Particular attention has been paid to standards within national guidelines that can support practice.

The book has also been designed to adopt a systematic manner in which it is presented. My opinion is that the reader will greatly benefit from this, starting with Chapter 1 as a revision tool covering the anatomy and physiology of the respiratory system. Each chapter then increases the reader's knowledge, highlighting age-appropriate care. Transition to adult services is later covered in Chapter 14. Other professional nursing issues are also described in great detail which will improve the overall care for this patient group.

Children and young people are very different from adults. This is most evident to nurses when treating children diagnosed with illnesses similar to those of adults. For this reason, nurses need special skills when planning their care.

Congratulations first to the editor, Janice Mighten. Congratulations also to all the chapter authors, contributors and publisher.

The authors have given a wealth of knowledge that can only come from their extensive experience. Although this book is not meant to be exhaustive, it includes a wealth of information for all who are caring for children and young people with respiratory conditions.

This textbook will be a welcome addition to my collection.

Angela Horsley
Clinical Lead, Nottingham Children's Hospital

Preface

The ever-changing world of modern medicine has enabled healthcare professionals to provide good-quality care. The idea for this book was born out of experiences with a merger of children's respiratory services and the need to provide a unified service based on quality and high standards. It is beyond the scope of this book to discuss every aspect of paediatric respiratory medicine, although a variety of conditions will be covered alongside the role of the children's respiratory nurse specialist, working as an important member of the multidisciplinary team.

This book is aimed at qualified nurses caring for children with respiratory conditions. It may also be useful as a point of reference to nurse specialists and students from a variety of backgrounds such as nurses, doctors and allied health professionals. This is an essential text that will support your practice and is based on standards, including national guidelines. Also included are illustrations, images and case studies to aid learning.

How to use this book

The approach that has been adopted is straightforward to guide the reader through the book in a systematic manner.

The book is divided into three sections. Each chapter begins with learning objectives and some chapters will have case studies that share relevant experiences. Finally, most of the chapters conclude with questions and answers, to enable the reader to consolidate learning. A glossary is also included at the end of the book explaining some of the more complex medical terminology.

Section I begins with an introduction that provides an overview of the respiratory nursing role and how this has evolved over time. An insight is then given into the anatomy and physiology of the respiratory system for children in Chapter 1. The process of homeostasis and the effect on the respiratory system are outlined in Chapter 2. The significance of nursing assessment and history taking is discussed in Chapter 3. This concludes with consideration of collaborative working with professionals in primary and secondary care. Reference to this concept is also continued throughout the book.

Section II begins with an outline of investigations in Chapter 4; this provides an overview of the various investigations that aid diagnosis and treatment such as chest x-rays and bronchoscopy. The images allow the reader to gain an understanding of the exploratory nature of such investigations and the relevance to treatment options. The process of assessment continues in Chapter 5, providing an insight into the assessment of airflow, including the impact spirometry testing has on daily management of patients with respiratory conditions and the necessity of nursing support to facilitate the process. Chapter 6 takes the concept of airflow further in relation to oxygen therapy, assessment, monitoring and evaluation based on national guidelines. Chapter 7 pursues the theme of long-term ventilation for children. Although this focuses on physiological and practical aspects, consideration is also given to the ethical dilemmas associated with long-term ventilation for some children. The importance of adequate nursing support for the family is also a feature of this chapter.

Section III begins with Chapter 8 which introduces the topic of respiratory infection, providing an overview of the common infections in children such as bronchiolitis and pneumonia, again referring to national and local guidelines. Chapter 9 provides a brief overview of pharmacology and the drugs used in respiratory medicine. Chapter 10 provides current information surrounding the diagnosis, care and management of children with asthma, including reference to the most current version of the British Thoracic Society management guidelines. Some of the most common congenital abnormalities that affect children are the main focus of Chapter 11. The theme of long-term conditions continues in Chapter 12, with particular reference to genetically inherited diseases such as cystic fibrosis and primary ciliary dyskinesia. The concept of multidisciplinary working is illustrated explicitly and is also applied to many other disease processes throughout the book. Maximum treatment can eventually be ineffective for some children with chronic lung disease so lung transplantation needs to be considered as an option. This is covered in a comprehensive and systematic manner in Chapter 13, including all elements of nursing care.

Section IV moves away from the management of conditions and focuses on the pertinent issues that impact on practice for children's nurses. Chapter 14 provides an overview of transition from children's to adult services, with reference to youth work services that support transition. For all healthcare professionals, the legal context of practice and quality assurance is very much a necessity, and this is discussed in Chapter 15. Finally chapter 16 completes this theme with a very important overview of communication and the skills required for all professionals involved in caring for children and families, in particular with chronic and long-term conditions.

I hope that this book is a valuable read for all to enjoy.

Janice Mighten

Acknowledgements

There are so many people who have worked tremendously hard towards making this book happen. Firstly I would like to thank all the contributors who have given their time to make a fine contribution towards the development of this book, and the medical photography department at Nottingham University Hospital for their help with some of the images. The positive feedback and evaluation from the reviewers are also appreciated; many of their suggestions have been incorporated into the final text. To all friends, colleagues and family, thank you for your support throughout this long process, and thanks to the publishers who have facilitated the process throughout.

Introduction: the evolution of children's respiratory nursing

Janice Mighten

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The health service has progressed over the years largely due to advancements in technology, which define how we treat many diseases. The changes that have occurred in nursing have been responses not only to technology but also to political influences and standards outlined within quality assurance frameworks. Other developments within the National Health Service (NHS) have also emerged, such as the concept of regional centres, generating high costs and resources. Consequently, nurses with specialist knowledge and skills were required, to meet the demand.

Many models of practice originated from North America and had some impact on elements of nursing care within the United Kingdom. This included specialist areas of nursing practice, which were recognised as early as 1979, within the Merrison Report, which also made reference to the concept of clinical nurse specialists (Middleton 2005).

Project 2000, introduced in the 1990s, changed nursing education and the concept of specialist areas (Holland et al. 2008). This provided specific areas of nurse training, such as the children's branch, and also set the standard for changes within nurse education. This change has continued further with the important move towards nursing becoming an all-degree profession, with emphasis on quality and standards, as suggested by the Prime Minister's Commission (Department of Health 2010). Basford and Slevin (2003) allude to such changes in nurse education and suggest that they have lead to the emergence of practitioners with qualities that include competency, safety and effective communication , which the modern health service demands.

Within the realms of paediatric respiratory medicine, we have witnessed the development of many nursing positions. Specialist areas such as paediatric respiratory nursing have emerged through the interest of individuals practising within the field of general respiratory medicine. This began with long-term conditions, such as asthma, and lead on to many more health conditions.

Wooler (2001) outlines the importance of the children's respiratory nurse specialist in the management of children with asthma in both primary and secondary care. Wooler also highlights the opportunity that such a role provides for children's respiratory nurse specialists to broaden their skills within respiratory medicine.

A general medical placement provides the learner with the opportunity to gain experience when caring for children with a variety of respiratory conditions. A qualified nurse with a special interest in paediatric respiratory medicine can be presented with opportunities within this field. Such positions are very varied, from clinical nurse specialists and advanced nurse practitioners to nurse consultants.

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Currently, there are very few nurses at consultant level within paediatric respiratory medicine. This suggests that the time is right to promote and encourage professional development for those who have the desire, passion and drive to reach such heights, even in these times of austerity. The ultimate aim will be to provide positive role models for the nurse specialists of the future.

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Section I

The fundamental principles of respiratory nursing

Chapter 1

Anatomy and physiology of the respiratory system

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Learning objectives

After studying this chapter, the reader will have an understanding of:

- the anatomy of the upper and lower respiratory tract
- stages of lung development
- the development of the respiratory system
- physiology of the respiratory system.

Introduction

A solid understanding of the anatomy and physiology of the respiratory system is an essential part of children's respiratory nursing. Furthermore, some knowledge of the embryological origins of those respiratory structures allows understanding of the development of congenital pathology.

The function of the respiratory system is simple: to provide oxygenation to the blood and removal of carbon dioxide. In disease, the mechanisms allowing such gaseous exchange are impaired. Therefore knowledge of the physiology of the upper and lower respiratory structures allows an understanding of why impairment of ventilation and perfusion occurs in various disease states.

Anatomy of the upper respiratory tract

The respiratory tract begins at the tips of the nostrils (alae nasi), which are kept open by soft cartilage. Around the nostrils are the alar nasalis muscles which cause the nostrils to flare open during states of respiratory distress, and can reduce nasal airway resistance by up to 25% (Carlo et al. 1983). The nasal cartilage encloses the anterior nasal cavity called the nasal vestibule. The cells of the nasal vestibule are the same as skin and contain small hairs, vibrissae, which can help stop debris such as dust from entering. There is a large vascular capillary network in the anterior vestibule, commonly called Little's area, which is a common site of nosebleeds in children.

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A midline nasal septum divides the nasal cavity into two. On the lateral walls lie three curved turbinate bones called conchae, which direct airflow. Air passing through the nasal cavity is warmed and humidified and prevents the airways from drying out. Ventilator humidifiers do the same thing when the nose is bypassed by an orotracheal tube. This is the beginning of the nasopharynx, the site where nasopharyngeal aspirates are taken. The cells in this area are ciliated respiratory epithelial cells, rather than squamous cells, and move any particulate matter towards the oropharynx where it can be swallowed.

It is notable that the lacrimal ducts drain into the nasal conchae and the eustachian tube, that equalizes pressure in the middle ear. The adenoids are located near this region of the nasopharynx, and during viral upper respiratory tract infections adenoidal hypertrophy can block the eustachian tube in some infants and children which can lead to otitis media with effusion (Wright et al. 1998).

The naso- and oropharynx lead to the pharnyx where the epiglottis protects the laryngeal opening from the tracheal aspiration of food and liquids. During swallowing, the epiglottis moves down

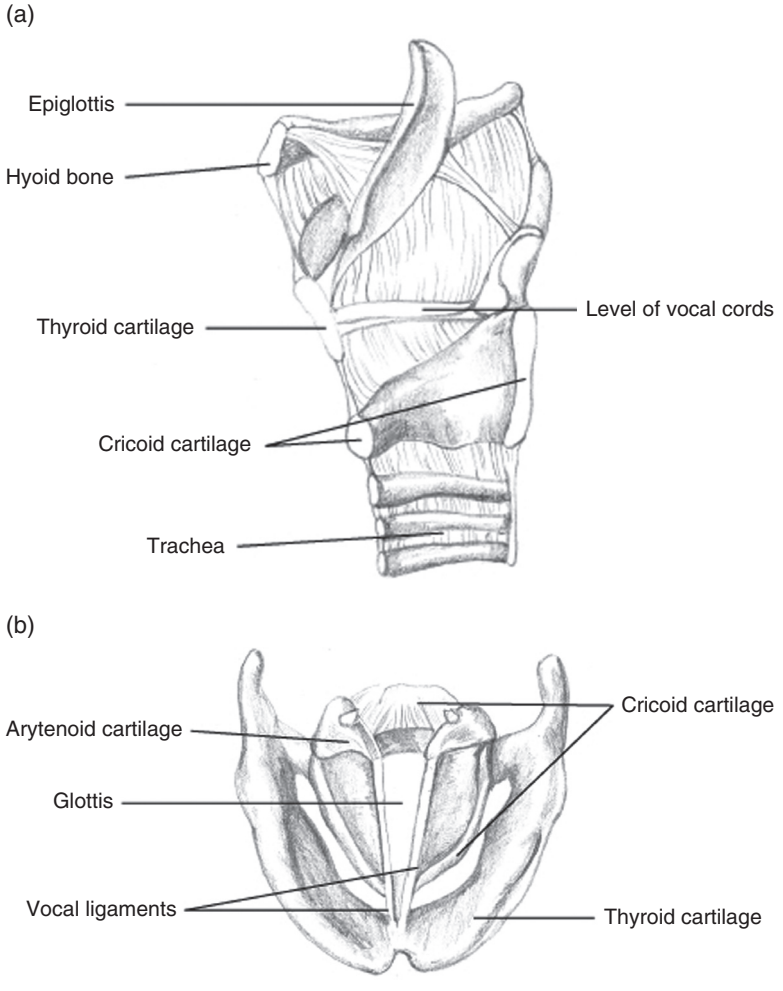

Figure 1.1 (a) Larynx showing cricoid and thyroid cartilages and level of vocal cords. (b) Vocal cords. Courtesy of Dr Phoebe Sneddon.

to close off the larynx. In epiglottitis, the epiglottis becomes very red and inflamed, swallowing becomes too painful, and the child drools.

The larynx is a complex structure that contains 'C'-shaped rings of cartilage and the vocal cords and muscles (Figure 1.1). The vocal cords and the space between them are commonly referred to as the glottis. Any abnormalities in this area will cause a variety of sounds, the most common being stridor. Below the vocal cords are the windpipe or trachea, which is part of the lower respiratory tract.

Anatomy of the lower respiratory tract

The trachea bifurcates at the carina to become the right and left main bronchi. The angles are slightly different, with the left main bronchi coming off at a more acute angle. Thus any inhaled foreign bodies tend to go down the right main bronchus. These bronchi then divide repeatedly into secondary and tertiary bronchi until finally dividing into the terminal bronchioles, respiratory bronchioles and finally alveoli.

The lung itself is covered by a pleural membrane which consists of the visceral and parietal pleura, with a small fluid-filled space in between. The visceral pleura covers the lung itself, while the parietal pleura is attached to the inner walls of the thorax. Infection and/or inflammation within the lung tissue can lead to accumulation of fluid or pus in this pleural space, respectively called a pleural effusion and empyema.

The work of breathing is done by the diaphragm and the intercostal muscles, located between the ribs. In poorly controlled respiratory conditions such as asthma, the diaphragm works much harder than usual and can deform the chest wall, as the muscle fibres attach to the lower part of the rib cage. This chronic deformity of the chest wall is called Harrison's sulci.

Surface anatomical landmarks

The ability to describe surface locations on the chest is important, and is usually described in terms of ribs or intercostal spaces and vertical lines drawn from anatomical landmarks. The second rib is located first by feeling for the sternal angle, then moving laterally. Other ribs can then be identified by counting downwards. The important vertical lines are the midclavicular and midaxillary. The midclavicular line passes straight down from the middle of the clavicle and the midaxillary line passes straight down from the axilla, when looking at the patient side on. In pneumothorax, needle thoracocentesis is performed by inserting a butterfly needle or venflon into the second intercostal space in the midclavicular line. Emergency chest drains are inserted into the fifth intercostal space in the midaxillary line.

Development of the respiratory system

Congenital defects of the upper airway originate from abnormalities of the embryological pharyngeal arches. Six arches are formed in the ventral surface of the hindbrain during the fourth to fifth weeks of embryological development, and give the embryo a characteristic appearance. These arches are derived from mesenchymal cells. The first pharyngeal arch mainly forms the lower jaw and anterior tongue, and defects can present as the Pierre Robin sequence with micrognathia, cleft palate and glossoptosis. The second pharyngeal arch gives rise to the root of the tongue, as well as other structures in the neck. Such a difference in embryological origin explains why the different

Table 1.1 Stages of human lung development

Stage	Age (weeks)	Structures
Embryonic	3–6	Trachea, lung buds, right and left main bronchi, lobar and segmental bronchi
Pseudoglandular	6–16	Bronchial tree to terminal bronchioles, pneumocyte precursors
Canicular	16–26	Terminal bronchioles, acini, type I and II pneumocytes
Saccular	26–36	Respiratory bronchioles, smooth-walled sacculi
Alveolar	36–maturity	Alveoli

parts of the tongue are innervated by different cranial nerves. The epiglottis forms from the fourth arch. The larynx opens in the 10th week of gestation. Incomplete opening at this point can lead to a laryngeal web, which can present in infancy as stridor.

The trachea and lower respiratory tract develop in the fourth week from the outpouchings of the embryological foregut, and thus are derivatives of endoderm. Incomplete separation from the gut leads to the condition of tracheo-oesophageal fistula. The lung buds divide in the fifth week, with three main divisions in the right bud and two in the left. These will eventually correspond to the three lobes of the right lung and the two of the left. By the end of the fifth week the embryonic stage of lung development is finished.

The diaphragm forms in the 6th week and failure of fusion can result in herniation of abdominal contents into the thorax – congenital diaphragmatic hernia. The left side is most commonly affected.

Following on from the embryonic stage are stages of lung development (Table 1.1) (Scarpelli 1990). Before 24 weeks' gestation, the lungs simply cannot function, even with exogenous surfactant. This stage of gestation is commonly seen as the limit of viability.

Changes in anatomy with age

In infancy, the narrowest point of the upper airway is the cricoid ring, rather than the vocal cords as in older children. Endotracheal intubation requires placing a suitably sized tube so as not to damage the vocal cords, whilst ensuring that any air leak is minimal. In younger children and infants the cricoid ring provides a seal, whereas in older children an endotracheal tube with an inflatable cuff is used. When the endotracheal tube passes through the vocal cords and is in the correct position, the cuff is inflated which creates a seal against the trachea and prevents air leak.

An important consideration in airway resistance is the change that occurs when the diameter is reduced due to mucus or inflammation. Poiseuille's law states that airway resistance is inversely proportional to the fourth power of the airway radius (Figure 1.2). Thus a 1 mm change in airway diameter in an older child will have little effect on resistance compared to that of a newborn or infant (Balfour-Lynn and Davies 2006).

Physiology of the respiratory system

The function of the lung is to oxygenate the blood and remove carbon dioxide. Air at sea level contains 21% oxygen, with inert nitrogen making up the remainder. In order for the oxygen to be delivered to the blood, flow of air into the lung must occur. To accomplish this, a pressure gradient

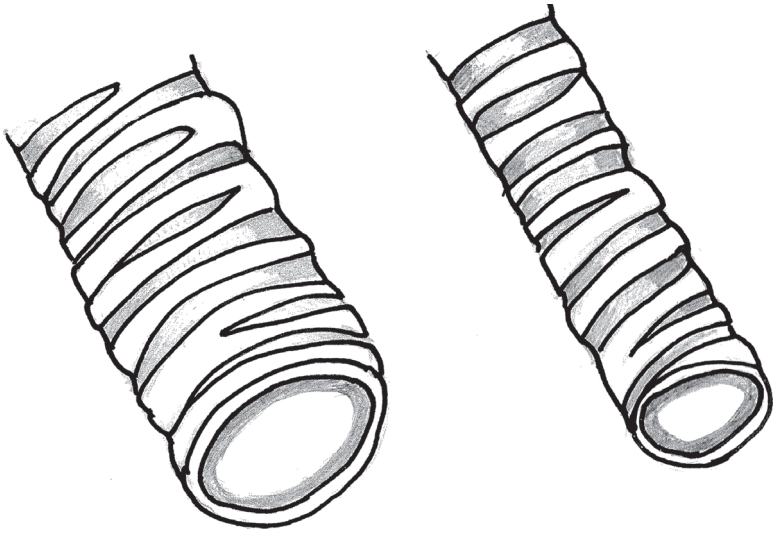

Figure 1.2 A similar amount of airway narrowing causes a much larger increase in airway resistance in smaller airways. Courtesy of Dr Phoebe Sneddon.

must be created between the terminal respiratory unit and the outside air. By contraction mainly of the diaphragm, against a thoracic cavity held rigid by the rib cage, a negative intrathoracic pressure is generated and flow of air occurs.

The anatomical 'dead space' consists of the terminal bronchioles, bronchi, trachea and upper airway. Although air passes through this dead space, no gas exchange occurs. Similarly, the tubes from a ventilator to the patient, including the endotracheal tube, extend this dead space. In neonatal ventilation, endotracheal tubes are kept as short as safely possible to reduce dead space.

During inspiration, the negative pressure exerts a force against the extrathoracic trachea and larynx, which instead of the rib cage relies on the cartilaginous rings to prevent collapse. During times of upper airway obstruction such as croup, increased effort to create flow will create further narrowing in the upper airway which is why inspiratory stridor occurs before expiratory stridor. In laryngomalacia the cartilage is not fully formed and stridor occurs as the larynx partially collapses with inspiration.

Involuntary breathing is controlled by centres in the brainstem which receive signals from chemoreceptors located in the medulla, carotid and aortic bodies. These chemoreceptors mainly respond to changes in acid–base balance which correspond to changes in blood carbon dioxide levels. Higher centres in the cortex can over-ride brainstem signals, allowing voluntary control of ventilation.

During exhalation, the diaphragm relaxes and the elastic recoil of the lungs creates a relative positive pressure within the airways to create flow of air out of the lungs. *Resistance* is the obstruction to airflow and is increased in conditions such as acute bronchiolitis and asthma. *Compliance* is the extent of lung inflation at a given inflation pressure. It is dependent on the production of surfactant by type II pneumocytes, which reduces the surface tension on the alveoli and prevents atelectasis. Low compliance is commonly referred to as a stiff lung.

The alveoli provide an enormous surface area for the diffusion of oxygen into the pulmonary blood and the removal of carbon dioxide. This assumes that the areas of the lung that are ventilated are also being perfused with pulmonary blood. In conditions such as asthma, in which

mucous plugging occurs, areas of lung are not ventilated or perfused by blood. This is called *ventilation/perfusion* or *V/Q mismatching*.

Oxygen then transfers across the alveolar capillary membrane, binds to haemoglobin and is carried to the tissues, where it is made available for aerobic metabolism.

Conclusion

This chapter has provided an overview of the development of the respiratory system. This should enable readers to fully appreciate how ill health and congenital abnormalities can affect the function of the respiratory system.

Questions

1. What is the function of the alar nasalis muscle?
2. What is the function of the conchae?
3. The main reason why an inhaled foreign body would go down the right main bronchus much more easily than the left is?
4. What defects are present in Pierre Robin sequence?
5. An infant with a laryngeal web would present with what?
6. Why does inspiratory stridor occur before expiratory stridor?

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Chapter 2

Homeostasis and the respiratory system

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Learning objectives

After studying this chapter, the reader will have an understanding of:

- the principles of homeostasis
- the respiratory rate, carbon dioxide and pH
- negative feedback mechanism
- how ill health disrupts homeostasis.

Introduction

We live in an ever-changing environment but despite this, the body needs to maintain its internal environment within strict limits. The process by which the body maintains internal consistency (or internal equilibrium) is termed *homeostasis* (Chiras 2002). Respiration is one of the many body systems which are regulated by homeostatic processes. This chapter describes this process and gives an example of how it can be affected by ill health.

Respiratory rate, carbon dioxide and pH

Blood pH needs to be held within a neutral range of approximately 7.35–7.45. A lower pH is too acid and a higher pH too alkaline. The body's metabolism naturally produces acids, most of which are ultimately excreted by the kidneys. Carbon dioxide is produced by all cells as they make energy, and is also acidic. However, carbon dioxide is an acidic gas and so it is removed from the bloodstream by the lungs through breathing. The rate of carbon dioxide removal from the body is proportional to the volume of each breath (bigger breaths remove more carbon dioxide) and the respiratory rate (faster breathing removes more carbon dioxide).

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Carbon dioxide dissolved in the blood regulates the respiratory rate

The brain regulates the amount of carbon dioxide in the blood by altering the respiratory rate and depth (also termed the *tidal volume*). Chemical sensors termed chemoreceptors in the medulla of the brain can determine if carbon dioxide levels have increased by detecting the decreased blood pH caused by the increased carbon dioxide (Chiras 2002). A drop in blood pH is detected by the medulla which then stimulates nerves to the diaphragm and intercostal muscles, increasing the respiratory rate and tidal volume (West 2004). This leads to an increase in the rate of removal of carbon dioxide from the body, and the blood levels of carbon dioxide fall back to normal. This in turn returns the blood pH to its normal level, removing the stimulus which previously increased the respiratory rate and tidal volume, and the tidal volume and respiratory rate settle at this new level (Figure 2.1).

Control of respiratory rate is an example of a negative feedback mechanism

In a negative feedback mechanism, a stimulus causes a response which removes the original stimulus, thus 'turning off' the response. You will notice that raised carbon dioxide triggers an

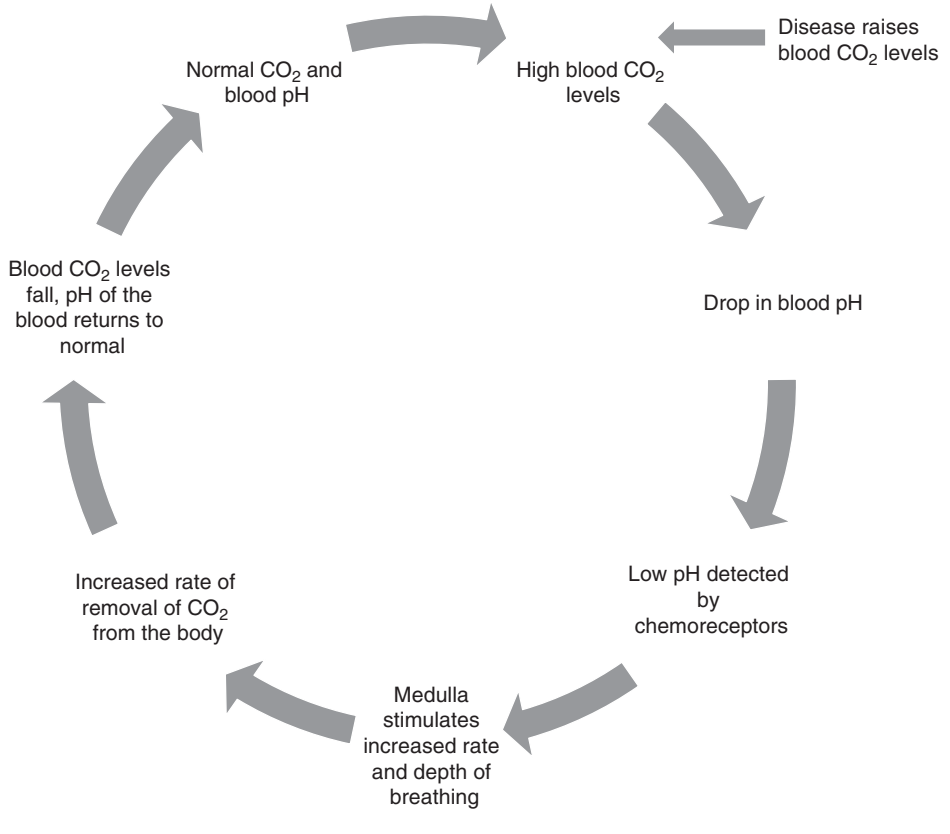

Figure 2.1 Carbon dioxide and respiratory control operate as a negative feedback loop.