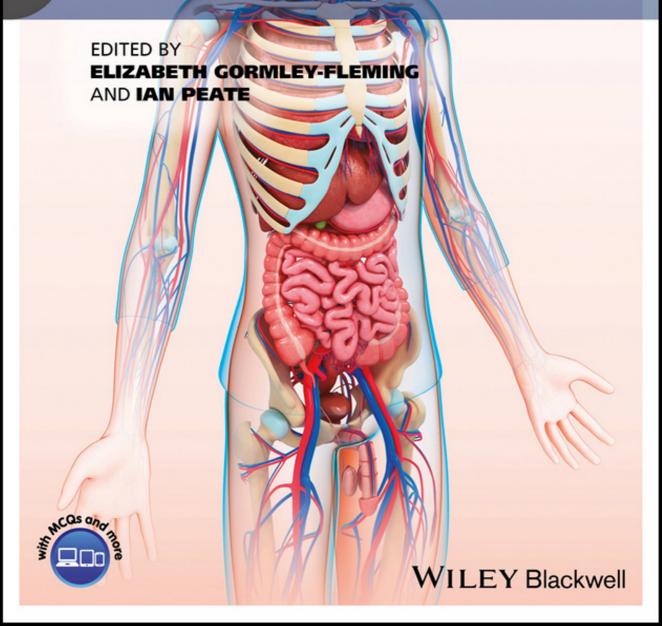
Fundamentals of

Children's Applied Pathophysiology

An Essential Guide for Nursing and Healthcare Students



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EDITED BY

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Contents

		vii xiii xv xvii xix
Chapter 1	The cell and body tissue Peter S. Vickers	1
Chapter 2	Genetics Peter S. Vickers	27
Chapter 3	Cancer Tanya Urquhart-Kelly	51
Chapter 4	Homeostasis <i>Mary Brady</i>	67
Chapter 5	Inflammation, immune response and healing Alison Mosenthal	83
Chapter 6	Shock Usha Chandran	115
Chapter 7	Pain and pain management Helen Monks, Kate Heaton-Morley and Sarah McDonald	133
Chapter 8	Disorders of the nervous system Petra Brown	159
Chapter 9	Disorders of the cardiac system Sheila Roberts	179
Chapter 10	Disorders of the respiratory system <i>Elizabeth Mills, Rosemary Court and Susan Fidment</i>	213
Chapter 11	Disorders of the endocrine system <i>Julia Petty</i>	233

Chapter 12	Disorders of the digestive system Ann L. Bevan	257
Chapter 13	Disorders of the renal system Cathy Poole	279
Chapter 14	Disorders of the reproductive systems <i>Michele O'Grady</i>	311
Chapter 15	Disorders of the musculoskeletal system Liz Gormley-Fleming	335
Chapter 16	Disorders of the skin Liz Gormley-Fleming	359
Index		383

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viii

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Liz commenced her nursing career in Ireland where she qualified as an RGN and RSCN. Initially working in paediatric oncology and bone marrow transplant, Liz moved to London and has held a variety of senior clinical nursing and leadership roles across a range of NHS Trusts both in the acute care setting and in the community. Liz has worked in education since 2001, initially as a clinical facilitator before moving into full-time higher education in 2003.

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Helen started her career in 1986 and gained her RGN qualification at Brad

Helen started her career in 1986 and gained her RGN qualification at Bradford and a few years later her RSCN qualification at Manchester. She has experience in nursing children and families within the fields of general surgery, plastic surgery and general medicine. During her practice, Helen has sought to empower both families and nurses via education and partnership approaches. Her career progressed to sister and ward manager where she became more interested in education, took up project nurse roles within Bradford's

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After a career break raising her family Alison returned to clinical nursing working as a clinical nurse specialist in paediatric immunology nursing at St George's Healthcare NHS Trust in 1996. She remains in clinical practice part-time and in 2010 returned to teaching in higher education at the University of Hertfordshire, where she currently works part-time as a senior lecturer in paediatric nursing.

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Tanya graduated from Sheffield School of Nursing and Midwifery in March 1993 as an RGN/RSCN and has worked in a variety of roles within the field of paediatric oncology/haematology nursing since qualifying. Most recently she worked as a Macmillan clinical nurse specialist in paediatric and teenage and young adult late effects at Sheffield Children's NHS Foundation Trust. She has recently taken a substantive child field nursing lecturer post at Sheffield Hallam University. Her key areas of interest are teenagers and young adults with cancer, transition and survivorship; particularly the endocrine care for survivors of childhood cancer. She was awarded a distinction and the faculty prize for her contemporary Master's degree in the care of teenagers and young adults with cancer from Coventry University, and holds certificates in endocrine nursing and research studies. She is recognised in the international arena of late effects following cancer in childhood, and has presented at numerous international symposiums. She was the previous chair of the CAN UK Nurses group (Cancer Aftercare Nurses group) and an active member of the CCLG (Children's Cancer & Leukaemia Group) Late Effects group.

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Following several years as a schoolteacher, Peter began his nursing career in 1980 at York District Hospital, before specialising in paediatric nursing at The Hospital for Sick Children, Great Ormond Street. His nursing specialties were paediatric immunology and immunode-ficiency, infectious diseases, and genetics. In 1999, he was awarded his PhD following his study of children with severe combined immunodeficiency who had survived bone marrow transplants in the UK and Germany (which was later published as a book). Following award of his PhD, Peter entered nurse education as a senior lecturer in paediatric nursing at the University of Hertfordshire, where he first began writing, and has gone on to publish widely in nursing textbooks and journals. He has also undertaken research into adult hospice care and written computer programmes on immunology for distance learning. He has also presented at conferences in many European countries, as well as North Africa. In 2012, Peter was elected President of INGID (the international organisation for immunology nurses), and in 2014, upon stepping down as INGID President, he was presented with a life-time achievement award in immunology nursing by INGID.

Preface

In order to provide safe and effective care to children and families, it is essential that those who are providing that care are able to understand the pathophysiology that underpins the child's condition.

The overall aim of this text is to help make the sometimes complex subject of pathophysiology accessible and exciting, and to enable the reader to apply their knowledge to various contexts of care. The body has an extraordinary ability to respond to disease in a variety of physiological and psychological ways. It is able to compensate for the changes that come about as a result of the disease process – the pathophysiological processes. The text can assist you in advancing your critical thinking; it fosters innovation and creativity in relation to the health and wellbeing of those to whom you have the privilege to offer care.

The text adopts a user-friendly approach – inviting you to delve deeper, discover new facts, and to engender curiosity. There are many illustrations, which are used in such a way as to explain and assist in understanding and appreciating the complex disease patterns that are being discussed. Applying a fundamental approach will provide you with a crucial understanding of applied pathophysiology, while emphasising that at all times the child and the family must be at the centre of all that is done.

A series of activities are provided, which are intended to help you learn in an engaged way and support you as you apply your learning in the various care settings, wherever these may be. This text offers an up-to-date overview of pathophysiology and the key issues associated with care provision.

The need to constantly consider the wider context of care provision, supplementing a nursing focus and recognising the broadening of the professional base, is emphasised. In providing care that is contemporary, safe and effective, an integrated, multidisciplinary approach is a key requirement. Healthcare students are important members of any multidisciplinary care team. It should also be acknowledged that contemporary care provision is delivered in ever-changing environments to a range of children, families, communities and circumstances.

Most chapters provide case studies that are related to chapter content. The chapters will stimulate reflection and further thought. In all case studies the names used are pseudonyms, in order to maintain confidentiality. Nurses owe a duty of confidentiality to all those who are receiving care (NMC, 2015). The majority of case studies have been extended further and include data concerning the patient's vital signs and blood analysis. This can help you to relate important concepts to care, offering you further insight into the patient's condition and therefore their needs. A selection of case studies include a Paediatric Early Warning Score (PEWS).

In England, nearly every hospital uses a different PEWS chart and calculates PEWS in varied ways. The PEWS charts included in this text are only there to demonstrate how they may be used. It must be remembered that infants (0–11months), preschool children (1–4 years), school-age children (5–12 years), and teenagers (13–18 years) will all require a PEWS chart that is specific to their age. You should familiarise yourself with the PEWS chart used in the organisation where you work.

Where appropriate, significant information related to the chapter appears in boxed format to focus the reader, for example, red flags and medicines management. This can help you when you are offering care to children and families who may be vulnerable and scared.

xiv

A feature found in most chapters is the investigations box. One investigation has been chosen related to chapter content. This contains details about the test or investigation encouraging the reader to think about the pre-, peri- and post-procedural care that the child and family may require.

All chapters begin and end with questions, which are there to test your pre- and post-knowledge. A range of learning resources are included at the end of the chapters, such as word searches, 'fill in the blanks', crosswords, and label the diagram activities. A list of further resources that you may wish to access with the intention of increasing and advancing your learning is provided at the end of each chapter. Each chapter also has a glossary of terms.

Pathophysiology is concerned with the cellular and organ changes that take place when disease is present, and the effects that these changes have on a person's ability to function. When something happens that interrupts the normal physiological functioning of the body, for example, disease, it becomes a pathophysiological issue. It must always be acknowledged that normal health is not and cannot be exactly the same in any two children, and thus when the term 'normal' is used, it must be treated with caution. An understanding of pathophysiology 'normal' and 'abnormal' can assist the healthcare student in helping the child and family in a kind, sensitive, compassionate, caring, safe and holistic way.

This text is a foundation text providing support to the reader as you grow personally and professionally in relation to the provision of care. The text is primarily intended for nursing students who come into contact with children who may have a number of physically related healthcare problems, in the hospital and community setting. Illness and disease are discussed explicitly, highlighting the fact that children do become ill and they experience disease.

It is not imagined that you will read the text from cover to cover – we would encourage you to dip in and out of it. However, it may assist in your learning if you first read Chapter 1 (The cell and body tissue) and Chapter 2 (Genetics), as these provide a good starting point – they set the scene. The aim is to entice and encourage you, to whet your appetite, and inspire you to read further, and in so doing we hope to instill a sense of curiosity in you.

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Liz Gormley-Fleming

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Ian Peate

About the companion website

This book is accompanied by a companion website:

www.wileyfundamentalseries.com/childpathophysiology

The website includes:

- Multiple-choice questions
- Further resources
- Word-search exercises
- Glossaries
- Crosswords
- 'Fill in the blanks' exercises
- True or false questions

How to use your textbook

Features contained within your textbook

Learning outcome boxes give a summary of the topics covered in a chapter.

Learning outcomes

On completion of this chapter, the reader will be able to:

- Outline the structure and function of a human cell.
- · Name and describe the functions of the organelles.
- Explain the cellular transport system.
- Describe the structures and functions of the various tissues of the body, namely: epithelial, connective, muscle and nervous tissues.

Keyword boxes give a summary of the keywords covered in a chapter.

Keywords

- heart
- circulation
- congenital
- acquired
- disorders
- heart failure

Every chapter contains 'Test your prior knowledge' questions.

Test your prior knowledge

- 1. Name three different treatment approaches for childhood cancer.
- 2. Name the most common form of childhood cancer.
- 3. What percentage of children with cancer are now cured? (a) >60%, (b) >70%, or (c) >80%.
- What is the difference between a malignant tumour and a benign tumour?
 What are the differences between chemotherapy and radiotherapy?

Case studies give an up-close, in-depth, and detailed examination of a subject.

Case Study 1

Sophie is 11 and is admitted to hospital for the first time in her life with abdominal pain. Her Mum is with her and is understandably anxious to find out what is causing Sophie's pain. She cannot stay long as she has to get back to nursery to collect Sophie's younger brother, Danny. She will come back later after taking Danny to her estranged husband's flat.

- 1. What pain tools are appropriate to help to assess Sophie's pain?
- 2. What other factors would need to be considered?
- 3. What non-pharmacological methods could used to help Sophie?

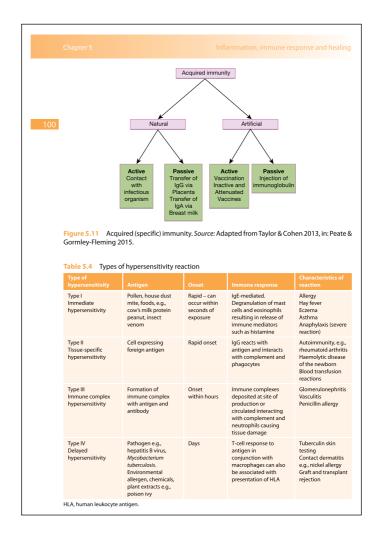
Red flags provide quick summaries of alert signs and symptoms.



Nursing considerations

Because this is a potentially fatal condition nurses need to be alert to older children presenting with a more chronic picture of diarrhoea, anorexia, weight loss, periodic pain and vomiting.

Your textbook is full of illustrations and tables.



Chapter 1

The cell and body tissue

Peter S. Vickers

Aim

The aim of this chapter is to introduce the reader to the various cells and tissues of the body in order to develop their insight and understanding.

Learning outcomes

On completion of this chapter, the reader will be able to:

- Outline the structure and function of a human cell.
- Name and describe the functions of the organelles.
- Explain the cellular transport system.
- Describe the structures and functions of the various tissues of the body, namely: epithelial, connective, muscle and nervous tissues.

Keywords

- cytoplasm
- plasma membrane
- organelles
- nucleus
- passive transport
- active transport
- epithelial tissue
- muscle tissue
- connective tissue
- mitochondria

Test your prior knowledge

- 1. What are the characteristics of human cells?
- 2. Describe the ways in which substances can pass through the cell membrane.
- 3. What is the role of the cell nucleus?
- 4. What are the four main roles of connective tissue?
- 5. How many different types of muscle tissue are there?
- 6. Where is epithelial tissue to be found within the body?

CELLS Introduction

What is a cell? Put simply, a cell is a building block for the formation of all life and, particularly in this case, for the formation and development of the human body. There are many different types of cells and they play different roles in both the structure and functioning of the body. For example, certain cells come together to form skin (a tissue), which acts as a cover and protector for our internal organs (tissues). Other cells combine to form bone (tissue) and hence our skeleton. Then there are other different cells which combine to make up the brain and neurological tissue (nerves). Outside the cells that form our structure are the cells that help to keep us functioning, for example, the cardiac cells, which combine to make the heart (tissue), which in turn keeps blood (cells and a tissue) flowing around our body carrying nutrients to all our cells and tissues and removing waste products from them. Some cells are involved in protecting us from infectious organisms, whilst others form muscles (tissues) which allow us to work and move. So, it can be seen that cells are the basic building blocks of our bodies – indeed, our very 'being'.

All these different types of cells are actually produced from just two cells – ovum and sperm – which fuse together at the moment of conception. Within those two cells are all the plans and schemata for producing the number and diversity of cells that make a human body – truly a miracle! Once they fuse together at conception, they begin to multiply and divide into the different types of cells. This manufacture and diversification of cells is dictated by the genes carried in all of our cells (see Chapter 2, Genetics).

This chapter will give a brief overview of the structure of cells and their roles within the body. In addition, it will discuss some of the problems that can occur and how these can affect the working and health of the body, commencing with the common characteristics of cells (Fig. 1.1).

Characteristics of cells

- Cells are active carrying out specific functions.
- Cells require nutrition to survive and function. They use a system known as endocytosis
 in order to catch and consume nutrients they surround and absorb organisms such as
 bacteria and then absorb their nutrients. These nutrients are used for the storage and
 release of energy, as well as for growth and for repairing any damage to themselves.
- Cells can reproduce themselves by means of asexual reproduction in which they first develop double the number of organelles (the organs of a cell) and then divide, with the same number and types of organelle and structure present in each half. This is known as simple fission.
- Cells excrete waste products.

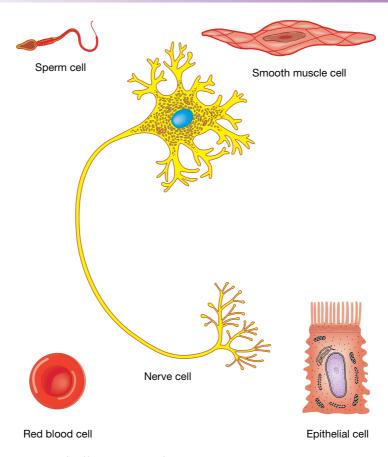


Figure 1.1 Examples of different types of cells in the body. *Source*: Tortora & Derrickson 2009, in: Peate & Gormley-Fleming 2015. Reproduced with permission of Wiley.

 Cells react to things that irritate or stimulate them – for example, in response to threats from chemicals and viruses.

The structure of the cell

There are four main compartments of the cell:

- cell membrane
- cytoplasm
- nucleus
- nucleoplasm.

Within these compartments are many organelles (or small organs). These organelles perform numerous roles to keep cells alive and functioning.

The cell membrane

As can be seen in Fig. 1.2, the various structures of the cell are contained within a cell membrane (also known as the plasma membrane). This cell membrane is a semi-permeable biological membrane separating the interior of the cell from the outside environment, and protecting the cell from its surrounding environment. It is semi-permeable because it allows only certain substances to pass through it for the benefit of the cell itself. For example,

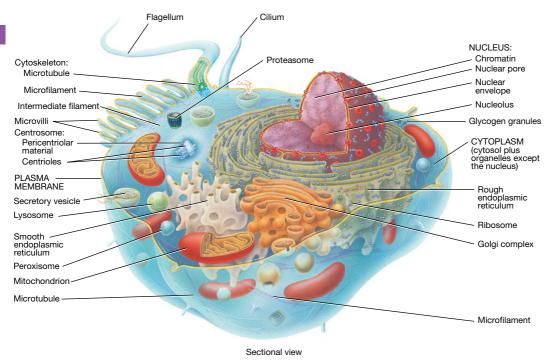


Figure 1.2 Structure of the cell. *Source*: Tortora & Derrickson 2009, in: Peate & Gormley-Fleming 2015. Reproduced with permission of Wiley.

it is selectively permeable to certain ions and molecules (Alberts *et al.*, 2014). Inside the cells are the cytoplasm and the organelles, which include, for example, the lysosomes, mitochondria, and the nucleus of the cell.

The cell membrane, which can vary in thickness from 7.5 nm (nanometres) to 10 nm (Vickers, 2009) is made up of a self-sealing double layer (bilayer) of phospholipid molecules with protein molecules interspersed amongst them (Fig. 1.3). A phospholipid molecule consists of a polar 'head', which is hydrophilic (mixes with water), and a tail that is made up of non-polar fatty acids, which are hydrophobic (repel water). In the bilayer of the cell membrane, all the heads of each phospholipid molecule are situated on the outer and inner surfaces of the cell facing outwards, whilst the tails point into the cell membrane; it is this central part of the cell membrane consisting of hydrophobic tails that makes the cell impermeable to water-soluble molecules (Marieb, 2014). In addition to the phospholipid molecules, the cell membrane contains a variety of molecules, mainly proteins and lipids, and these are involved in many different cellular functions, such as communication and transport. The proteins inserted within the cell membrane are known as plasma member proteins (PMPs), which can be either integral or peripheral. Integral PMPs are embedded amongst the phospholipid tails whilst others completely penetrate the cell membrane. Some of these integral PMPs form channels for the transportation of materials into and out of the cell, others bind to carbohydrates and form receptor sites (e.g., attaching bacteria to the cell so they can be destroyed). Other examples of integral PMPs include those that transfer potassium ions in and out of cells, receptors for insulin, and types of neurotransmitters (Vickers, 2015). On the other hand, peripheral PMPs bind loosely to the membrane surface, and so can be easily separated from it. The reversible attachment of proteins to cell membranes has been shown to regulate cell signalling, as well as acting as enzymes to catalyse cellular reactions through a variety of mechanisms (Cafiso, 2005).

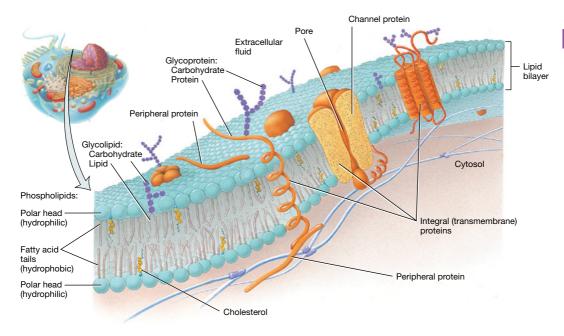


Figure 1.3 Cell membrane. *Source:* Tortora & Derrickson 2009, in: Peate & Gormley-Fleming 2015. Reproduced with permission of Wiley.

Functions of the cell membrane

Briefly, the two major physiological functions of the cell membrane are endocytosis and exocytosis. These are both concerned with the transport of fluids and other essential particulates and waste matter into and out of the cell.

- **Endocytosis** is the passing of fluids and small particles into the cell. There are three types of endocytosis, namely:
 - **Phagocytosis** the ingestion of large particulates, such as microbial cells
 - **Pinocytosis** the ingestion of small particulates and fluids
 - **Receptor-mediated** involving large particulates, such as protein. It is also highly selective as to which particulates are taken up.

Endocytosis involves part of the cell membrane being drawn into the cell interior, along with particulates or fluid, in order to facilitate their ingestion. This part of the membrane is then 'pinched off' to form a vesicle within the cell. At the same time, the cell membrane reseals itself. Once inside the cell, the fate of this vesicle depends upon the type of endocytosis involved and the material that is contained within the cell membrane surrounding it. In some cases, the vesicle may ultimately fuse with a lysosome (an organelle), following which the ingested material can be processed. Endocytosis is also the means by which many simple organisms – such as amoeba – obtain their nutrients.

The cell membrane and transport

Selective permeability, as mentioned in the previous section, is very important to the process of transporting materials into and out of the cell, allowing certain materials to pass through the membrane, whilst preventing others that could harm the cell. This process depends upon the hydrophobicity of some of its molecules (as mentioned earlier). Because the phospholipid molecule tails are composed of hydrophobic fatty acid chains, it is difficult for hydrophilic (water-soluble) molecules to penetrate the membrane. Hence it forms

an effective barrier for these types of molecules, which can only be penetrated by means of specific transport systems that control what can enter or leave the cell. For example, the membrane controls the process of metabolism by restricting the flow of glucose and other water-soluble metabolites into and out of cells – as well as between subcellular compartments. In addition, the cell stores energy in the form of transmembrane ion gradients by allowing high concentrations of particular ions to accumulate on one side of the membrane. lons can pass through the membrane from inside the cell to the outside – or vice versa – so that there are more supplies of these ions just outside the cell or inside it. The membrane controls the speed/rate at which these ions pass through the membrane. The controlled release of such ions on the gradients can be used for:

- extracting nutrients from the fluids around the cells
- passing electrical messages (nerve excitability)
- controlling the volume of the cell.

Cell membrane permeability

There are four factors involved in the degree of permeability of a cell membrane, namely:

- 1. The size of molecules large molecules cannot pass through the integral membrane proteins, whilst small molecules (e.g., water, amino acids) can.
- 2. Solubility in lipids (fats) substances that easily dissolve in lipids (e.g., oxygen, carbon dioxide, steroid hormones) can pass through the membrane more easily than non-lipid soluble substances can.
- **3.** If an ion has an electrical charge that is the opposite of that found in the membrane, then it is attracted to the membrane and so can more easily pass through it.
- **4.** Carrier integral proteins can bind to substances and carry them across the membrane, regardless of the three processes above, i.e., size, ability to dissolve in lipids, or membrane electrical charge.

Movement of substances across the membrane

There are two ways for this to occur, namely, passive and active.

Passive processes

A passive process is one in which the substances move under their own volition down a concentration gradient from an area of high concentration to an area of lower concentration. In this process, the cell expends little energy on the process (like rolling down a hill).

There are four types of passive transport processes, namely:

- diffusion
- facilitated diffusion
- osmosis
- filtration.

Diffusion is the most common form of passive transport. A substance in an area of higher concentration moves to an area of lower concentration (Colbert *et al.*, 2012). The difference seen between areas of different concentrations is known as the concentration gradient. This particular passive transport process is essential for respiration. It is through diffusion that oxygen is transported from the lungs to the blood and carbon dioxide from the blood into the lungs.

Although similar to diffusion, facilitated diffusion differs from it by the use of a substance (a facilitator) to help in the process (see Fig. 1.4). As an example, glucose is moved

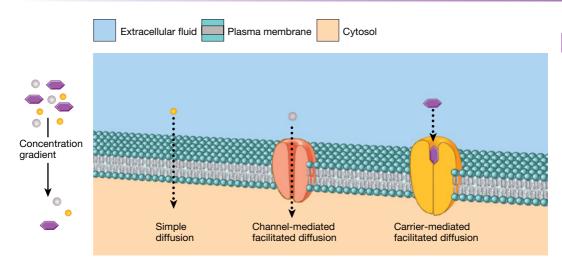


Figure 1.4 Facilitated diffusion. *Source*: Tortora & Derrickson 2009, in: Peate & Gormley-Fleming 2015. Reproduced with permission of Wiley.

using this process. To be able to pass through a membrane, glucose needs to attach itself to a carrier/transport protein (McCance & Huether, 2014).

Osmosis is the process by which water travels through a selectively permeable membrane so that concentrations of a solute (a substance that is soluble in water) are equal on both sides of the membrane. This gives rise to osmotic pressure. The higher the concentration of the solute on one side of the membrane, the higher the osmotic pressure available for the movement of water (Colbert *et al.*, 2012).

If osmotic pressure rises too much, then it can cause damage to the cell membrane, so the body attempts to ensure that there is always a reasonable constant pressure between the cell's internal and external environments. We can see the possible damage if, for example, a red blood cell is placed in a low concentrated solute, then it will undergo haemolysis. On the other hand, if it is placed in a highly concentrated solute, the result will be a crenulated cell. If the red blood cell is placed in a solution with a relatively constant osmotic pressure, it will not be affected because the net movement of water in and out of the red blood cell is minimal.

Filtration is similar to osmosis, with the exception that physical pressure is used in order to push water and solutes across a cell membrane. This is seen in renal filtration, where the heart beating exerts pressure as it pushes blood into the kidneys, where filtration of the blood can then take place to remove any impurities (Colbert *et al.*, 2012).

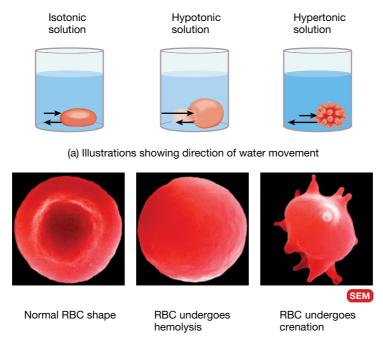
Active processes

Active processes are:

- active transport pumps
- endocytosis
- exocytosis.

An active process is one in which substances move against a concentration gradient from an area of lower to higher concentration. In order for this to happen, the cell must expend energy, which is released by the splitting of adenosine triphosphate (ATP) into adenosine diphosphate (ADP) and phosphate.

ATP is a compound of a base, a sugar and three phosphate groups (triphosphate), and is held together by phosphate bonds, which release a high level of energy when they are



(b) Scanning electron micrographs (all 15,000x)

Figure 1.5 Effect of solute concentration on a red blood cell. *Source:* Tortora & Derrickson 2009, in: Peate & Gormley-Fleming 2015. Reproduced with permission of Wiley.

broken. Once one of the phosphate bonds is broken and phosphate has been released, that compound then becomes ADP. The 'spare' phosphate will then join another ADP group, so forming ATP (with energy stored in the phosphate bond). This process is continually recurring within the body.

Active transport pumps need energy to be able to function. This energy occurs as a result of the reaction mentioned earlier. It is necessary when the body is attempting to move an area that already has a high concentration of that substance. The higher the concentration already present, the more energy is required to move further molecules of that substance into that area. Fig. 1.5 demonstrates the effect of solute concentration on a red blood cell.

The organelles

These are rather like small 'organs' within the cells. The following sections give a brief overview of the many cell organelles and their functions.

Cytoplasm

Although, not strictly speaking, an organelle, the cytoplasm is a very important and integral part of the cell interior. Cytoplasm is ground substance (a 'matrix') in which various cellular components are found. It forms part of the protoplasm of the cell (protoplasm is the collective name for everything within a cell). Cytoplasm is a thick, semi-transparent, elastic fluid containing suspended particles along with the cytoskeleton (the cell framework). The cytoskeleton provides support and shape to the cell and is involved in the movement of structures within the cytoplasm – for example, phagocytic cells. Chemically, cytoplasm is made up of 75–90% water along with solid compounds, particularly carbohydrates, lipids and inorganic substances.