Handbook of Clinical Anaesthesia

Fourth Edition

Edited by
Brian Pollard
Gareth Kitchen

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For Claire, Mum and Dad, your unwavering support makes endeavours like this possible.
To my children, Joseph and Amelia – follow your dreams; anything is possible.

GK

For all of our patients. May this book improve both your safety and your experience of anaesthesia.

BJP

Contents

Preface

Welcome to the fourth edition of the *Handbook of Clinical Anaesthesia*. We have retained the overall structure as in the first three editions. The book continues to be a collection of individual entries each covering a particular topic, condition or problem which may be encountered in clinical anaesthesia. The philosophy of the book has been retained in that all of the information is presented in a concise form without unnecessary information or 'padding'.

Over its lifespan between the first and the fourth editions, this book has undergone a significant evolution which we believe has served to improve it. The original idea was conceived by John Goldstone and Brian J Pollard in 1994. John unfortunately had to withdraw from the project at the second edition. For the fourth edition a second editor has been introduced again, Dr Gareth Kitchen. The choice of Gareth is clear. He is a young academic anaesthetist who has been able to instil new thoughts into the book and assist in driving it forwards and bringing on board a number of new names as experts in their fields.

In the first two editions, the authors of the various sections and monographs were drawn almost exclusively from the UK. In the third edition, the authorship was widened into a much more international field. In this fourth edition, we have returned to it being a UK-based field for the authors. Not only that but as we, the editors, are based in the Northwest, we have selected our authors principally from this area as there is a huge amount of expertise here.

Remember that this book is not an exhaustive treatise. It does not cover every eventuality; no book can do that. The *Handbook of Clinical Anaesthesia* is a distillation of facts and guidance and is intended to complement the major texts in the subject. Individual entries are referenced where appropriate but the references are limited to a small number of key sources and include up-to-date reviews wherever possible.

Over the years this book has proved popular with trainees preparing for examinations in the speciality. It has also proved very popular with established consultants and specialists who keep it beside the phone, on the office desk or in the operating theatre suite for straightforward advice on problems or situations encountered.

Finally, we would like to pay tribute to the many authors involved in the first three editions of this book. A significant proportion of their text and information has been retained where the advice has not materially changed. Many sections have nevertheless been rewritten as appropriate and updated as necessary. The authors involved in the first three editions are too numerous to mention but to each and every one we thank you for your input to the previous editions and hope that you approve of this new version and its updated information.

BJP and GK

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List of abbreviations

A&E	Accident & Emergency
AAGBI	Association of Anaesthetists of Great Britain and Ireland
ABG	Arterial blood gas
ACE	Angiotensin converting enzyme
Ach	Acetylcholine
AChE	Acetyl cholinesterase
ACS	Acute coronary syndrome
ACTH	Adrenocorticotrophic hormone
ADH	Antidiuretic hormone
AF	Atrial fibrillation
AHA	American Heart Association
AKI	Acute kidney injury
ALS	Advanced life support
AMP	Adenosine monophosphate
APTT	Activated partial thromboplastin time
ATP	Adenosine triphosphate
BIPAP	Bilevel positive airways pressure
BMI	Body mass index
BPM	Beats per minute
CABG	Coronary artery bypass graft
CAD	Coronary artery disease
CCF	Congestive cardiac failure
CNS	Central nervous system
CO	Cardiac output
CO_2	Carbon dioxide
COPD	Chronic obstructive pulmonary disease
CPAP	Continuous positive airways pressure
CPET	Cardiopulmonary exercise test
CRF	Chronic renal failure
CSF	Cerebrospinal fluid
CT	Computed tomography
CVA	Cerebrovascular accident
CVC	Central venous catheter
CVP	Central venous pressure
CVS	Cardiovascular system
CXR	Chest x-ray
DBP	Diastolic blood pressure
DVT	Deep venous thrombosis
ECG	Electrocardiogram
ECT	Electroconvulsive therapy
EDV	End diastolic volume
EEG	Electroencephalogram
EF	Ejection fraction
EMG	Electromyogram
ESA	European Society of Anaesthesiology
ESC	European Society of Cardiology
ETT	Endotracheal tube
FBC	Full blood count
FEV_1	Forced expiratory volume in 1 second
FiO_2	Inspired fraction of oxygen
FRC	Functional residual capacity
FVC	Forced vital capacity
GABA	Gamma hydroxybutyric acid
GCS	Glasgow comas scale
GFR	Glomerular filtration rate
GH	Growth hormone
GMP	Guanosine monophosphate
GP	General practitioner
GTN	Glyceryl trinitrate
Hb	Hemoglobin
HDL	High-density lipoprotein
HDU	High dependency unit
HIV	Human immunodeficiency virus
HOCM	Hypertrophic obstructive cardiomyopathy
I/E	Inspired : expired ratio
IABP	Intra-arterial blood pressure
ICP	Intracranial pressure
ICU	Intensive care unit
IG	Immunoglobulin
IHD	Ischemic heart disease
IM	Intramuscular
IPPV	Intermittent positive pressure ventilation
IQ	Intelligence quotient
IV	Intravenous
JVP	Jugular venous pressure
kPa	Kilopascal
LBBB	Left bundle branch block

LDL	Low-density lipoprotein
LFT	Liver function test
LM	Laryngeal mask
LMA	Laryngeal Mask Airway ™
LMN	Lower motor neuron
MAC	Minimum alveolar concentration
MAP	Mean arterial pressure
MSH	Melanocyte stimulation hormone
NICE	National Institute of Health and Care Excellence
NIV	Noninvasive ventilation
NMJ	Neuromuscular junction
NO	Nitric oxide
NREM	Nonrapid eye movement
NSAID	Nonsteroidal anti-inflammatory drug
NSTEMI	Non-ST-elevation myocardial infarction
NYHA	New York Heart Association
O_2	Oxygen
P_{50}	Partial pressure of oxygen at 50% saturation
PaO_2	Arterial partial pressure of oxygen
PAC	Pulmonary artery catheter
$PaCO_2$	Arterial partial pressure of carbon dioxide
PAP	Pulmonary artery pressure
PEEP	Positive end-expiratory pressure
PEFR	Peak expiratory flow rate
PET	Positron emission tomography
PFT	Pulmonary function test
PONV	Postoperative nausea and vomiting
PT	Prothrombin time
PTH	Parathyroid hormone
PTT	Partial thromboplastin time
PVR	Pulmonary vascular resistance
RBBB	Right bundle branch block
REM	Rapid eye movement
SaO_2	Arterial saturation of oxygen
SAP	Systolic arterial pressure
SLE	Systemic lupus erythematosus
STEMI	ST-elevation myocardial infarction
SVR	Systemic vascular resistance
TB	Tuberculosis
TIA	Transient ischemic attack
TIVA	Total intravenous anesthesia
TNF	Tumor necrosis factor
TNM	Tumor nodes metastases
TOE	Transesophageal echocardiogram
TSH	Thyroid stimulating hormone
TTE	Transthoracic echocardiogram
U&E	Urea and electrolytes
UK	United Kingdom
UMN	Upper motor neuron
USS	Ultrasound scan
V/Q	Ventilation : perfusion
VC	Vital capacity
VF	Ventricular fibrillation
VT	Tidal volume
WHO	World Health Organization

PART 1 PATIENT CONDITIONS

1 Respiratory system

MATTHEW STAGG

ASTHMA

A very common respiratory disorder characterized by recurrent attacks of paroxysmal dyspnoea with reversible variable airflow obstruction and increased bronchial hyper-responsiveness to a range of stimuli. Aetiology, pathology and clinical presentation are heterogenous, but an underlying inflammatory response is usually present. There is an immense range of clinical pathology from children with reversible bronchospasm through to elderly patients in whom bronchospasm is superimposed on chronic respiratory disease. The incidence of intraoperative bronchospasm is low and tends to occur in older asthmatics and those with active or poorly controlled asthma at the time of operation.

EPIDEMIOLOGY

Variable geographical distribution, affecting about 5% of the population as a whole but up to 10% of children.

MORBIDITY

Increased risk of postoperative respiratory complications, especially in the older patient with chronic airways disease in whom cardiac problems may also be present.

Mediator	Bronchospasm	Oedema	Mucus secretion
Histamine	+	+	+
Prostaglandin	+	+	
Leukotrienes C4, D4, E4	+	+	+
Thromboxane	+	+	
Platelet activating factor	+	+	

PATHOPHYSIOLOGY

Nonspecific airway hyper-responsiveness is common. There is an increased response to methacholine, exercise, histamine, cold-air challenge, hyperventilation or extreme emotional stimulus. Airway obstruction is due to constriction of airway smooth muscle, mucus secretion and oedema of the airway wall. Mechanisms include neural and cellular pathway activation. The neural pathway involves afferent irritant receptors in airways, causing reflex stimulation of postganglionic parasympathetic fibres, resulting in smooth muscle constriction and mucus secretion. C fibre stimulation releases local neuropeptides; substance P changes membrane permeability and mucus secretion; neurokinin A causes bronchoconstriction. Cellular pathway activation involves immunoglobin E mediated histamine release from mast cells. Eosinophils, neutrophils, macrophages and lymphocytes CD8 and Th1 may also release mediators including leukotrienes, LTB4 and the cysteinyl leukotrienes, CysLT. LTB4 is a pro-inflammatory mediator with potent neutrophil chemotaxic properties while CysLTs are potent bronchoconstrictors that increase vascular permeability, cause mucus secretion, mucociliary dysfunction, stimulate eosinophil recruitment and increase bronchial responsiveness. At a cellular level, smooth muscle tone is controlled by intracellular cyclic AMP and possibly cyclic GMP, with lower levels leading to bronchoconstriction. The effect on ventilatory function can be extreme, with increased work of breathing, air trapping, exhaustion, hypercapnia and potentially fatal V/Q mismatch resulting in life-threatening hypoxia. This may be sustained for several days.

PREOPERATIVE ASSESSMENT

Optimise treatment in consultation with a respiratory physician. Severity and frequency of attacks, hospital admissions, exercise tolerance, current medication and trigger factors are essential information. Frequency of inhaler use may inform about severity and stability of their asthma. Steroid use, time of last exacerbation, timing and duration of any hospital admission are important.

Factors that indicate increased propensity to bronchospasm include recent or current upper respiratory tract infection, steroid use and past history of respiratory complications related to surgery. Previous ICU admission, especially one requiring intubation and ventilation, should act as a 'Red Flag'. In non–asthmatics, a family history of atopy or of asthma should alert to the possibility of intra-operative bronchospasm.

Some patients with COPD may have a significant reversible component. The presence of wheezes might indicate inadequate control and suggest medication review. The presence of a respiratory tract infection is a relative contraindication to anaesthesia.

INVESTIGATIONS

Chest X-ray – Look for hyperinflation; chronic lung changes or concomitant cardiac problems in older patients; evidence of right ventricular predominance, suggesting long-standing major problems.

ECG – Look for evidence of long-standing right ventricular hypertrophy or cor pulmonale. Such patients constitute a very high-risk group.

Lung function tests – FEV_1 reduced more than FVC (FEV_1 normally 50 mL kg^{-1}, and 70%–80% FVC).

Blood gases – Useful in asthmatics with COPD to be used as a baseline to guide postoperative target goals.

MEDICATION

The range of agents that can maintain control of asthma is considerable (Table 1.1). Many are long acting. Patients should continue on their maintenance therapy throughout their hospital stay if possible.

Preoperative management strategies.

Table 1.1 Agents used to maintain control of asthma

Drug type	Example agents	Side effects
Stabilizing agents	Sodium chromoglycate	
Bronchodilators		
β_2 agonist short-acting 4–6 h	Salbutamol/levosalbutamol	Tremor anxiety tachycardia
	Terbutaline	Hypokalaemia/hypomagnesaemia
β_2 agonist long acting >12 h	Arformeterol	Less side effects
	Salmeterol	No antiinflammatory action
Phosphodiesterases	Aminophyline	Tachycardia/arrhythmias
Inhaled steroids	Becotide, flixotide, budenoside	
Inhaled anticholinergics	Ipratropium	
Leukotriene antagonists	Zileuton montelukast pranlukast	
IgE immunotherapy	Omalizumab	
Others	Ketamine	Sympathomimetic
	Magnesium	Smooth muscle relaxation
Gases	Volatile agents	Bronchodilation
	Heliox	Reduced airway resistance
		$FiO_2 < 1$

Clinical	Preoperative intervention
Asymptomatic No medications No recent asthma episodes No obstruction on spirometry	Nothing needed
Occasional bronchodilators	Probably nothing needed
No steroids	Dose prior to induction suggested
Inhaled steroids	Continue inhaled steroids Give bronchodilator prior to induction
Spirometry below baseline	Consider short course oral steroids
Oral steroids	Continue same dose preoperatively Consider extra dose at induction Probable hydrocortisone postop for several days

PREMEDICATION

Sedation is useful as anxiety may provoke an attack in some patients. Atropine or glycopyrrolate inhibits vagally mediated bronchospasm but produces tachycardia. Preoperative bronchodilators and steroids reduce the likelihood of postoperative complication, so consider an additional dose of bronchodilator by inhaler or nebulizer prior to induction. Patients on steroids should receive steroids and if on high doses (>1500 μg day^{-1} in adults; less in children) give peri- and postoperative replacement as adrenal suppression may be present. Neither wound healing nor infection problems are relevant with these short periods of increased steroid use.

CHOICE OF ANAESTHESIA

Regional anaesthesia is recommended but anxiety can trigger bronchospasm so patient acceptance is important. If general anaesthesia is necessary, avoid stimulation of the respiratory tract and drugs known to cause bronchospasm.

INDUCTION

Avoid agents that may release histamine. Thiopentone is safe although it can cause histamine release and does not block airway reflexes. Propofol has bronchodilator properties and suppresses airway reflexes. Etomidate is safe. Ketamine is suitable for induction and maintenance by infusion in the asthmatic patient with bronchospasm requiring emergency anaesthesia,

although it may produce tachycardia and increased secretions. It may also be used to treat status asthmaticus. Sevoflurane is widely used and well tolerated.

INTUBATION

Spraying the larynx with lidocaine, prior to intubation, may help although it can itself stimulate bronchospasm (not histamine mediated). The use of a laryngeal mask avoids airway stimulation and the need for muscle relaxants. If control of hypercapnia, airway protection or emergency ventilation is required, an endotracheal tube is the only option.

MAINTENANCE

Halothane, enflurane, isoflurane and sevoflurane are all potent bronchodilators. They have been used in the treatment of refractory asthma and are ideal for maintaining anaesthesia.

MUSCLE RELAXANTS

Suxamethonium is a potent histamine releaser, so avoid if possible. Atracurium and mivacurium are associated with bronchospasm from histamine release. Pancuronium, vecuronium and cisatracurium appear safe while rocuronium has been associated with some severe reactions although in high does may be used as an alternative to suxamethonium. Reversal with anticholinesterases can trigger bronchospasm although atropine or glycopyrrolate given concurrently reduces the severity of this.

ANALGESIA

Local and regional techniques are recommended, but are not always feasible. Morphine and diamorphine release histamine and should be avoided. Pethidine has been widely used although may have some histamine-releasing potential. Fentanyl and alfentanil are safe. Exercise caution with NSAIDs unless previous exposure has not yielded problems.

POSTOPERATIVE MANAGEMENT

Problems in older asthmatics usually relate to underlying chronic lung disease. Effective analgesia assists physiotherapy and coughing so preventing the development of atelectasis and concurrent infection. Warm, humidified air and bronchodilators minimize the impact of mucus retention and plugging.

THE EMERGENCY CASE WITH CURRENT SYMPTOMATIC BRONCHOSPASM

A potentially disastrous situation, but fortunately rare. Surgery must be absolutely life or limb threatening to warrant proceeding. If possible, use a regional technique. Treat bronchospasm aggressively with IV steroids, magnesium sulphate 2 gm IV and/or aminophylline IV. The induction agent of choice is ketamine, followed by ketamine infusion, although other induction agents are often used effectively and safely. Suxamethonium may release histamine but its use may be difficult to avoid, unless high dose rocuronium is an option. Fentanyl is recommended for analgesia. Inhalational agents (sevoflurane or halothane) are effective in treating bronchospasm. Once deep on these agents, the patient may be better controlled than prior to induction. Continued bronchospasm with high airway pressure may necessitate IV beta agonists or even epinephrine (nebulizer or intravenously) in extreme circumstances.

Ventilation may pose problems, as airway pressures are likely to be high. Manipulate tidal volume, rate and I/E ratio to minimize peak airway pressure but maintain adequate minute ventilation. Permissive hypercapnia is reasonably tolerated. The possibility of a pneumothorax must be continuously considered. Postoperative management should be in ICU.

DEVELOPMENT OF INTRAOPERATIVE ASTHMA

Not all wheezing is asthma. Tube contact with the carina or a main bronchus can produce wheezing. Airway obstruction may result from tube blockage, secretions or blood. Aspiration, tension pneumothorax, anaphylactic or anaphylactoid reaction may all produce bronchospasm.

Salbutamol (2–5 μg kg^{-1}) or aminophylline (5 mg kg^{-1}) slow IV may be given. Steroids (e.g.

hydrocortisone) will not have an immediate effect but may assist in gaining control. Airway pressures may have been very high so beware of a pneumothorax.

The end of the case is a critical time when bronchospasm may appear in an awakening patient. Extubation deep may reduce the likelihood of bronchospasm but in many cases is inappropriate. Reversal with neostigmine can provoke bronchospasm but atropine or glycopyrrolate reduce the risk. Avoiding all reversal agents is ideal. Sugammadex may be a viable alternative.

DRUGS TO AVOID

Tubocurarine
Morphine, diamorphine and other histamine-releasing opiates
Beta blockers
Aspirin and other NSAIDs which are prostaglandin mediated

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CROSS-REFERENCES

Intraoperative bronchospasm, Chapter 30
Ventilators, Chapter 27

BRONCHIECTASIS

Bronchiectasis is characterized by long-standing abnormal dilatation of bronchi with chronic inflammation. This chronic inflammatory process results in patients being extremely productive of sputum with a predisposition to either chronic infection or colonisation with intermittent acute episodes of infection.

Historically bronchiectasis was a consequence of chronic recurrent infection. Pneumonias, measles, whooping cough, TB and fungal infections were the main causes. Now with antibiotics, vaccination and better nutrition it is far less common. Cystic fibrosis and smoking are now the main causes. Sometimes patients will present for surgical treatment of their bronchiectasis. There are some specific associated syndromes including Kartageners (the combination of situs inversus, sinusitis and bronchiectasis).

Diagnosis is by high-resolution CT scan and anaesthesia for bronchography has been relegated to history.

PATHOPHYSIOLOGY

Following childhood pneumonia or recurrent adult infections.

Congenital:
- – Cystic fibrosis
- – Bronchial cartilage deficiency
- – Abnormal ciliary motility (Kartageners)
- – Hypogammaglobulinaemia

Distal to bronchial obstruction:
- – Inhaled foreign body
- – Tumour

Clinical features are variable. In severe bronchiectasis there is up to 500 mL of purulent sputum per day, which gets dramatically worse during an acute exacerbation. Other features include haemoptysis from areas of severe inflammation with altered local circulation arising from bronchial and intercostal arteries. In long-standing disease pulmonary hypertension and cor pulmonale may develop. Metastatic abscess formation can occur. Amyloidosis is a rare complication.

MANAGEMENT

Chest physiotherapy with percussion and postural drainage is key but early intervention with antibiotics may prevent acute exacerbations. These patients are often chronically colonised with resistant organisms due to frequent antibiotic exposure. *Pseudomonas aeruginosa* and *Haemophilus influenzae* are particularly common.

PREOPERATIVE ASSESSMENT

Exercise tolerance (compared with their usual state), sputum production and frequency of acute exacerbations predict the severity. Information about colonising organisms and antibiotic history are important.

INVESTIGATIONS

Blood gases – To determine present baseline, and to guide postoperative target goals.

Chest X-ray – Probably not of benefit. A recent CT scan is helpful.

Pulminary function tests – Generally not very helpful.

ECG – Look for signs of right ventricular strain or cor pulmonale.

Echocardiogram – Helpful in assessing right ventricular hypertrophy, myocardial function and raised pulmonary pressures.

PREOPERATIVE MANAGEMENT

The patient will need extensive physiotherapy and be exacerbation free prior to surgery. Discussion with chest physician and microbiologist should determine the appropriate antibiotic to use preoperatively.

ANAESTHETIC MANAGEMENT

The surgery will determine the most appropriate form of anaesthesia. If possible, use regional techniques. Use routine monitoring commensurate with the anaesthetic and surgery. Have a very low threshold for an arterial line.

There are no particular agents that are contraindicated. Try to keep the oxygen saturations high (>90%) to maintain a safety margin. End tidal CO_2 is likely to be different from the arterial value but should provide trend measurements.

Sputum retention is likely to be a problem and will predispose to secondary infection. Humidify all gases and persist with regular tracheal suction. It may be necessary to use a bronchoscope to remove inspissated secretions and sputum. In cases with very severe localised bronchiectasis, it may be feasible to try to isolate that part of the lung with a bronchial blocker.

Proper attention to sterile technique is important, particularly in those with Kartageners syndrome as they also have a defect in neutrophil chemotaxis. Nasal tubes should be avoided in view of the accompanying sinusitis.

POSTOPERATIVE CARE

Arrange early postoperative physiotherapy in advance. In cases of cystic fibrosis, HDU care may be helpful to ensure mobilization and physiotherapy. Good analgesia is essential and patient-controlled devices, epidural analgesia or NSAIDs are all useful. Entonox may be helpful. Avoid postoperative ventilation wherever possible.

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CROSS-REFERENCES

Preoperative assessment – specific medical problems, Chapter 25
Cardiopulmonary exercise testing, Chapter 25

BRONCHOGENIC CARCINOMA

Lung cancer is the most common cause of cancer mortality worldwide for men and women, causing approximately 1.2 million deaths per year (Table 1.2). The most common symptoms are unexplained persistent cough, haemoptysis, shortness of breath, chest pain, bone pain and weight loss. They may develop from airways or parenchyma.

The main types are *non-small cell lung carcinoma* (NSCLC) and *small cell lung carcinoma* (SCLC). Early stage (stage 1–2) NSCLC is treated with surgery, while SCLC is treated by chemotherapy and radiation. Other tumours including large cell, neuroendocrine (carcinoid), bronchioloalveolar and rarer forms can all present as lung malignancies. The most common cause is long-term exposure to tobacco smoke. Lung cancer in non-smokers (15% of cases) is often attributed to a combination of genetic factors, radon gas, asbestos, air pollution and passive exposure to cigarette smoke.

Derived from the epithelium, squamous cell carcinomas are the most common NSCLC. They are usually centrally located at the carina or in the 1–3rd generation bronchi. Adenocarcinoma is less common with peak incidence in men in their fifties.

Presentation includes airway obstruction, lung collapse, and distal infection or through spread via the peribronchial tissues with subsequent invasion of the mediastinum. It spreads by both lymphatic and haematological routes and distal metastasis is common in liver, adrenals, bone and brain.

All forms of treatment can be associated with notable toxicity. Patients with significant impairment due to their lung cancer or comorbid conditions may not be fit to undergo resection or even aggressive chemoradiotherapy. Performance status can be assessed by a variety of methods including the Karnofsky Performance Status (KPS) or the World Health Organisation (WHO) status.

Anaesthetic involvement is mainly for lung resection (e.g. lobectomy, pneumonectomy). However, newer indications for palliative interventional bronchoscopic procedures are increasing. Debulking/disobliteration of central symptomatic obstructive lesions followed if necessary by tracheobronchial stents can ameliorate some symptoms of advancing disease. This may be done by rigid or flexible bronchoscopy, using a number of different modalities such as electrocautery, laser, cryotherapy/cryoextraction, argon plasma coagulation or mechanical debulking.

PREOPERATIVE ASSESSMENT

Patients may be asymptomatic or may present with a range of symptoms and signs including:

Local – Chest pain, cough, dyspnoea, haemoptysis, hoarseness, pleural effusion.
Distal – Metastases with associated problems.
Other – Ectopic hormonal activity from paraneoplastic tumours (e.g. ACTH, PTH, ADH, insulin and glucagon). Some manifestations of Cushing's syndrome can occur with hypokalaemia although the full clinical features of Cushing's syndrome are rarely seen as they do not have time to develop. Lambert–Eaton syndrome has been reported. Serotonin secreting adenomas may present as episodic sweating, wheeze and

Table 1.2 Lung cancer and its incidence

Characteristic	Squamous cell (epidermoid)	Adenocarcinoma	Large cell	Small cell
Approximate incidence	25%–30%	30%–35%	15%–20%	20%–25%
5-year survival	25%	12%	13%	1%
Operability	43%–50%	35%	35%–43%	Rare
Potential for metastasis	Low to moderate	Moderate	Moderate	High
Response rate to systemic treatment	Low	Low	Low	Moderate

breathlessness. These patients are usually smokers and COPD is a common concomitant problem.

INVESTIGATIONS

- *Chest X-ray* – May not reveal the tumour but may show signs of concomitant problems such as COPD. A pleural effusion or pericardial effusion would suggest mediastinal invasion.
- *ECG* – Thoracic surgery can result in rhythm disturbance, especially atrial fibrillation. Smokers have a high incidence of asymptomatic heart disease.
- *Electrolytes* – May indicate ectopic ADH secretion with low sodium which will eventually produce clinical signs of confusion and weakness. Ectopic ACTH secretion can result in hypokalaemia or hyperkalaemia with or without hypernatraemia. PTH produces hypercalcaemia but so do widespread bony metastases with elevated alkaline phosphatase. Glucose values can be adversely influenced by ectopic insulin or glucagon.
- *Lung function tests* – Important if any significant lung resection is planned. FEV_1 and FVC are most useful, whilst low gas transfer (below ~30%) may have implications for risk of post-operative respiratory failure. CPET may be helpful and baseline arterial blood gases on air should be taken.

Patients will have likely presented through a lung multidisciplinary team. A chest CT and or CT-PET scan, and tissue sampling by bronchoscopy, trans-bronchial needle aspiration, mediastinoscopy or interventional radiology will have staged the disease enabling appropriate management.

INOPERABILITY

The TNM staging system of the international union against lung cancer (Table 1.3) will determine which primary lung cancers are theoretically operable. In general, stage 1 and 2 disease is operable. Some classical indicators of inoperability exist which indicate stage 3 or 4 advanced disease. These include SVC obstruction or other great vessel involvement, nerve palsies including left recurrent laryngeal and phrenic nerve damage, carinal or tracheal involvement, oesophageal invasion, vertebral involvement and Pancoast's syndrome.

Pancoast's syndrome is an apical carcinoma invading the eighth cervical and first thoracic nerves. Severe pain and wasting in the upper limbs occur with stellate ganglion involvement. The patient often has Horner's syndrome (ptosis, enophthalmos, miosis, impaired sweating on face).

Very often these patients have palliative stents placed for debulked endobronchial disease or symptomatic compressive extrinsic disease. They can be silicon or metallic-nitinol alloy (placed via rigid bronchoscopy or interventional radiology) requiring general anaesthesia. Nitinol bronchial stents can be placed via flexible bronchoscopy under general anaesthesia or conscious sedation, or through endobronchial tubes. Complications include migration, misplacement, infection, biofouling and stent fractures (in older generation stents). These procedures usually offer immediate relief of symptoms and at least short-term benefit in the acute setting. They have even been attributed to liberation from mechanical ventilation after acute respiratory failure.

PREOPERATIVE PREPARATION

Optimize respiratory function – beta 2-adrenergic agonists, anticholinergics, active physiotherapy and steroids as indicated.

Any sizeable effusions should be drained. Electrolytes and haemoglobin should be corrected. While a restrictive approach to transfusion should be adopted, these patients are at risk of ischaemic heart disease so aim for Hb > 10 g/dL.

In patients having debulking techniques or stenting, careful consideration of anatomical placement of the stent should be discussed with the operator prior to anaesthesia. Modern imaging provides useful information that often correlates with functionality. Patients will often be dyspnoeic and may have partially collapsed lung segments. They are usually dramatically improved by the procedure but if the collapse has been long-standing it may be

Table 1.3 TNM staging system for lung cancer (7th edition)

Primary tumour (T)			
T1	Tumour ≤3 cm diameter, surrounded by lung or visceral pleura, without invasion more proximal than lobar bronchus		
T1a	Tumour ≤2 cm in diameter		
T1b	Tumour >2 cm but ≤3 cm in diameter		
T2	Tumour >3 cm but ≤7 cm, or tumour with any of the following features: Involves main bronchus, ≥2 cm distal to carina Invades visceral pleura Associated with atelectasis or obstructive pneumonitis that extends to the hilar region but does not involve the entire lung		
T2a	Tumour >3 cm but ≤5 cm		
T2b	Tumour >5 cm but ≤7 cm		
T3	Tumour >7 cm or any of the following: Directly invades any of the following: chest wall, diaphragm, phrenic nerve, mediastinal pleura, parietal pericardium, main bronchus <2 cm from carina (without involvement of carina) Atelectasis or obstructive pneumonitis of the entire lung Separate tumour nodules in the same lobe		
T4	Tumour of any size that invades the mediastinum, heart, great vessels, trachea, recurrent laryngeal nerve, oesophagus, vertebral body, carina, or with separate tumour nodules in a different ipsilateral lobe		
Regional lymph nodes (N)			
N0	No regional lymph node metastases		
N1	Metastasis in ipsilateral peribronchial and/or ipsilateral hilar lymph nodes and intrapulmonary nodes, including involvement by direct extension		
N2	Metastasis in ipsilateral mediastinal and/or subcarinal lymph node(s)		
N3	Metastasis in contralateral mediastinal, contralateral hilar, ipsilateral or contralateral scalene, or supraclavicular lymph node(s).		
Distant metastasis (M)			
M0	No distant metastasis		
M1	Distant metastasis		
M1a	Separate tumour nodule(s) in a contralateral lobe; tumour with pleural nodules or malignant pleural or pericardial effusion		
M1b	Distant metastasis		
Stage groupings			
Stage IA	T1a–T1b	N0	M0
Stage IB	T2a	N0	M0
Stage IIA	T1a,T1b,T2a	N1	M0
	T2b	N0	M0
Stage IIB	T2b	N1	M0
	T3	N0	M0
Stage IIIA	T1a,T1b,T2a,T2b	N2	M0
	T3	N1,N2	M0
	T4	N0,N1	M0
Stage IIIB	T4	N2	M0
	Any T	N3	M0
Stage IV	Any T	Any N	M1a or M1b

irrecoverable and predispose to infection. Careful planning is required.

PREMEDICATION

Minimise stress to the patient with an anxiolytic if necessary. A drying agent may help.

ANAESTHETIC TECHNIQUE

In patients with tracheal or bronchial compromise, coughing may become problematic and threaten airway patency. Inhalational techniques are likely to precipitate problems. General anaesthesia with muscular relaxation and mechanical ventilation is usually required. Almost any induction technique is suitable. Short- to medium-acting relaxants which do not accumulate are ideal with neuromuscular monitoring. Volatile agents are bronchodilators. Some advocate the use of heliox during induction if there is significant airway narrowing. Remifentanil has been recommended.

For lung resection, a double lumen endotracheal tube allows single lung ventilation and optimises surgical field. Alternatively an endobronchial blocking balloon may be placed under bronchoscopic vision.

Partial or complete central airway obstruction or symptomatic trachea-broncho-oesophageal fistulae can sometimes be palliated by debulking and/or stenting, respectively. Stents require appropriate and careful planning regarding position, size and type. Bronchial stents may be deployed awake or under general anaesthesia. Rigid bronchoscopy with a Sanders injector is a well-established technique, as is the suspension laryngoscope. Remember that the Sanders injector can result in high pressure air trapping if there is partial obstruction. Adequate neuromuscular reversal is vital prior to extubation. At the end of the case ensure there is a good cough reflex.

PATIENTS WITH PREEXISTING STENTS NEEDING ANAESTHESIA

Ensure the stent position is known, image if possible, seek an opinion from whoever placed the stent and ideally view the stent bronchoscopically prior to intubation The aim is to avoid dislodging the stent. In an emergency, try to visualise it before intubation if possible.

ACUTE POSTOPERATIVE CENTRAL AIRWAY OBSTRUCTION

It may be difficult to reestablish spontaneous breathing. The appearance has been likened to inadequate neuromuscular reversal with an ineffective breathing pattern that is largely abdominal. Desaturation ensues often associated with a deteriorating level of consciousness which may be in part due to hypercarbia. Blood gases show hypercarbia and hypoxia. Assume airway obstruction. Control the airway and with the aid of a surgeon go to rigid bronchoscopy as secretions at the carina or in the trachea are the most likely cause. The differential diagnosis is tension pneumothorax after airway instrumentation but that is very rare from stent placement in experienced hands.

POSTOPERATIVE CARE

This depends on the nature of the surgery, requirement for ventilation, preoperative respiratory function and other co-morbidities. Even without ventilation, these patients will often require specialist postoperative care.

Epidurals, oral opioids and PCA are most commonly used for analgesia. Pain will impair chest movement so good analgesia is key to recovery.

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Lobectomy, Chapter 15
Pneumonectomy, Chapter 15
One lung anaesthesia, Chapter 28
Intraoperative bronchospasm, Chapter 30
Preoperative assessment of pulmonary risk, Chapter 25
Cushings syndrome, Chapter 6
Myasthenia, Chapter 3

CHRONIC OBSTRUCTIVE PULMONARY DISEASE (COPD)

A common chronic inflammatory disease of the lungs, there is a spectrum associated with expiratory airflow obstruction including emphysema and chronic bronchitis. It has pulmonary and systemic manifestations. Management guidelines are well established with evidence-based recommendations for chronic disease and acute exacerbations.

In 1990, COPD was ranked 12th as a burden of disease by the WHO; by 2020 it is projected to rank 5th. Cigarette smoking is its primary cause and up to 25% of smokers are likely to develop COPD.

Respiratory disease accounts for more than 25% of acute hospital admissions, of which more than half are acute exacerbations of COPD. Hospitalization carries up to 26% mortality, rising to 66% within 2 years.

The prevalence of COPD is 5%–10% among general surgical patients, 10%–12% in cardiac surgery and 40% in thoracic surgery as compared to 5% in the general population.

The pulmonary component of COPD is characterised by expiratory airflow limitation that is not fully reversible. The diagnosis, severity assessment and monitoring rely heavily but not exclusively on spirometry. In smokers, lung function decline is accelerated beyond the natural 20–30 mL annual loss.

Airflow limitation is usually progressive and associated with an abnormal inflammatory response of the lung. Respiratory failure in COPD may be type 1 (predominantly hypoxic) or type 2 (associated hypercapnia). Chronic respiratory failure is often related to chronic hypoventilation, and may lead to cor pulmonale if untreated.

Patients with COPD, particularly severe disease, are at significant risk of postoperative complications. Preoperative recognition and optimisation can reduce these risks.

PATHOPHYSIOLOGY

Inflammatory small airways disease, destruction of alveolar units, inflammatory bronchiolitis and excess mucus production lead to airflow obstruction. Airways are no longer held open due to reduced elasticity and tone of the parenchyma. The combination of airway collapse prior to full emptying, bronchospasm and secretions produces expiratory airflow limitation and gas trapping. Loss of alveolar units decreases gas transfer.

An early manifestation is an increase in residual volume. The natural history is progressive gas trapping with decreasing vital capacity (VC). This results in a decline in forced expiratory volume in 1 second (FEV_1), further exacerbated by rapid shallow breathing, which leads to dynamic hyperinflation (Figure 1.1).

This increase in work of breathing is in part responsible for dyspnoea and exercise limitation. Due to differential transluminal pressures within the small conducting airways and at the alveolar level, lung units have different time constants for emptying. Across the whole lung, this results in retained intrathoracic pressure and so called 'intrinsic PEEP'. In the longer term, an increase in residual volume and chronic hyperinflation with reduced efficiency of resting diaphragm position and function results. In those with chronic carbon dioxide retention, there is a predisposition to developing pulmonary hypertension and right heart failure (cor pulmonale). Associated conditions are malnutrition, musculoskeletal disorders, cardiovascular disease,

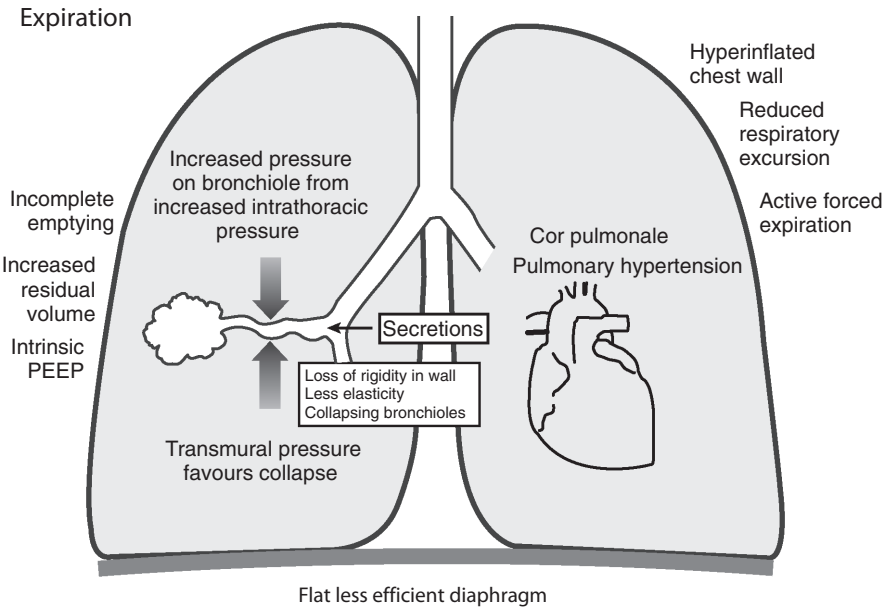

Figure 1.1 Respiratory effects of COPD during expiration.

diabetes and depression. Reduced weight, peripheral muscle strength and chronic sputum production portend a worse prognosis.

Ultimately, the final common pathway of decreased gas transfer, alveolar hypoventilation and respiratory muscle disadvantage produces ventilation/perfusion mismatch (V/Q) resulting in hypoxaemia and/or hypercarbia.

There is no clear correlation between lung function and blood gas features. Patients with hyperinflation, and resting tachypnoea, often have low arterial oxygen tensions and low gas transfer, but do not retain carbon dioxide on increasing oxygen therapy – these tend to be the emphysema spectrum patients. Conversely, it is the chronic bronchitic patients who tend to hypoventilate, are comfortable at rest, have chronic ventilatory failure with stable hypercapnia, but better spirometry, less gas trapping and preserved gas transfer that are at risk of hypercapnic narcosis with high inspired oxygen levels.

PREOPERATIVE ASSESSMENT

The risk of postoperative pulmonary complications and postoperative respiratory failure is high while lesser complications such as atelectasis or infection are common.

HISTORY

Enquire about:

- Exercise tolerance, breathlessness, orthopnoea, sputum productivity.
- Exacerbations requiring noninvasive ventilation, oral steroids, hospital and ICU admissions.
- Symptoms of sleep disordered breathing (excessive daytime sleepiness or tiredness, snoring and witnessed apnoeas).

EXAMINATION

Look for:

- Hyperinflation
- Right heart dysfunction
- Incipient infection in the oropharynx
- Wheezes or rhonchi (correlate with complications so consider bronchodilators or steroids to eliminate wheezing)
- Ischaemic heart disease

There is a significant risk of sudden death in uncontrolled heart disease. Heart failure, in particular, is a prognostic indicator with a 30% five-year survival rate. The exclusion and treatment of reversible ischaemia is paramount. The use of beta-blockers is controversial because of the risks of bronchospasm. A discussion between cardiologist and respiratory physician should determine the benefit risk ratio. A cardioselective beta blocker combined with inhaled steroid/bronchodilator may be indicated. Optimise statin and anti-platelet treatment.

INVESTIGATIONS

- Blood tests according to local guidelines.
- FBC to look for polycythaemia.
- Respiratory function tests. Compare current values with pre-existing results to identify any deterioration. Peak flow, FEV_1 and mid-expiratory flow rates are useful as a marker of the severity of limitation when considered with exercise tolerance. The residual volume to total lung capacity ratio is a useful indicator of gas trapping and potential surrogate of dynamic hyperinflation when approaching 50%. Reduced gas transfer, particularly kCO, may be indicative of emphysema.
- Blood gases will give the normal values for the individual, and the likelihood of chronic carbon dioxide retention, with high bicarbonate levels.
- ECG is essential. Include exercise testing to identify reversible ischaemia, and echocardiography if concerns of secondary pulmonary hypertension associated right heart dysfunction or cor pulmonale exist.
- Sleep studies should be considered if an element of sleep apnoea is suspected.

PREOPERATIVE OPTIMISATION

1. *Timing*

   Unless surgery is urgent, time is helpful in improving the preoperative state. Involve a chest physician and investigate cardiovascular disease.

2. *Stop smoking*

   Current smokers are at greater risk of complications. Smoking should be stopped at least eight weeks before surgery. There is some evidence to suggest that cessation or reduction <8 weeks before surgery increases the risk of complications.

3. *Optimise drug treatment*

   Most patients have some reversibility of lung function, or functional improvement with bronchodilators – refer to NICE guidance. The use of short-acting beta agonists, with long-acting muscurinic agonists is now routine – it reduces exacerbations and improves quality of life. When FEV_1 is less than 60% predicted and associated with two or more exacerbations per year, then an inhaled steroid/long acting beta agonist combination is recommended. Oral mucolytics are used as adjuncts in chronic deteriorating disease, whilst oral methylxanthines do not have a favourable evidence base and are no longer advised in acute or chronic settings, mainly due to increased risk of arrythmias.

   Oral steroids, for a minimum of a week, are known to reduce the duration of exacerbations, reduce reattendance rates after hospital admissions, and prevent admission at the sign of infection, when combined with antibiotic in those with severe disease. Surgery should be delayed if possible.

   A few days before surgery, in those with severe disease, a short course of oral steroids can be considered, if there are no objections related to surgical wound healing. Special care with diabetic patients is advised. An alternative is IV hydrocortisone at induction. Nebulised bronchodilator therapy should be given perioperatively. The role of the nebulised mucolytic N acetylcysteine in the perioperative or postoperative setting is not established. It is considered as an adjunct to physiotherapy in those with retained mucus and limited expectorating ability. It should be used with bronchodilators because of a risk of bronchoconstriction.

4. *Preoperative physiotherapy*

   Important for airway clearance. Continue postoperatively to reduce retained sputum and segmental collapse. In severe disease, noninvasive ventilation may be considered in the

postextubation period together with breaks for airway clearance.

5. *Pulmonary rehabilitation*

   Exercise tolerance and lung function are improved up to 6 months after completion. There is also emerging data to suggest improvements with pulmonary rehabilitation after exacerbations. Its value in the shorter term is not clear.

6. *Thromboprophylaxis*

   These patients have an increased risk of venous thromboembolism, so appropriate thromboprophylaxis is important, as well as early mobilisation and appropriate hydration.

REGIONAL ANAESTHESIA

Regional anaesthesia circumvents many problems. The patient must be able to tolerate lying relatively flat. Position, procedure and duration are important. These patients are often dependent on abdominal excursion and have prolonged active expiration when breathing normally and so regional techniques that extend as high as T8 may be problematic. Exercise care when using interscalene blocks as the potential for phrenic nerve and diaphragm palsy exist.

GENERAL ANAESTHESIA

Indications include major or prolonged procedures where regional or other techniques are inadequate or inappropriate, the need for muscle relaxation and when the patient's condition necessitates ventilation. Laryngeal mask ventilation is being increasingly used to preserve laryngeal reflexes, particularly relevant to the need for effective postoperative airway clearance.

At induction attempts to modify the bronchoconstrictive effects of intubation include the local application of lidocaine or the use of beta sympathomimetics. Use drugs unlikely to cause histamine release or exacerbate bronchospasm – e.g. propofol, thiopentone ketamine and etomidate. The muscle relaxant used should also be chosen carefully. Morphine may release histamine and also may have long-acting sedative properties. Fentanyl or the use of regional or central analgesic blocks, with or without infusion catheters, may be preferred. In severe COPD, doses of all drugs should be tempered by the predisposition of these patients to cardiovascular instability. Inhalational agents have good bronchodilating properties except desflurane which may provoke coughing, bronchospasm and tachycardia. The beneficial effects may, however, be offset by a delayed recovery. TIVA may be considered. As optimal mobility and coughing is important, a technique that results in a rapidly awake alert and comfortable patient has advantages.

MONITORING

ROUTINE MONITORING TO AAGBI STANDARDS

Additional monitoring depends on magnitude and type of surgery. An arterial line is recommended both for pressure monitoring and for repeated blood gases.

IPPV may cause air trapping and increase intrathoracic volume. Careful monitoring of ventilator parameters is therefore important. The increase is unpredictable as is the consequent increase in intrinsic PEEP. This may impede venous return and hence cardiac output. Raised pulmonary hypertension associated right heart dysfunction is a theoretical risk, and may manifest as rhythm changes, as a result of left ventricular impairment. BIPAP, reduced frequency rates and long expiratory times, with 'permissive hypercapnia' may be considered to minimise dynamic hyperinflation and its cardiovascular impact.

Capnography is essential as it will clearly show if the CO_2 trace does not reach a plateau. This indicates ongoing incomplete emptying of alveoli. There will be a large difference between end tidal and arterial CO_2. If air trapping is occurring, there will be some degree of intrinsic PEEP. Ventilators that can measure intrinsic PEEP are useful.

The use of extrinsic PEEP is controversial. It may increase air trapping. Alternatively it may splint airways open reducing trapping and reducing the inspiratory effort to reopen collapsed bronchioles.

The use of bronchodilators should be considered intra-operatively if difficulties in ventilation arise.

Lung volume reduction surgery in this population has specialist anaesthetic implications beyond the remit of this chapter.

POSTOPERATIVE CARE

These patients pose difficult postoperative management problems and pulmonary complications are common. The risk of postoperative respiratory failure (>48 h mechanical ventilation) is 3%–3.5%. The presence of severe COPD increases the risk 1.5 times, whilst lesser complications such as atelectasis or infection are more common. These are more likely in thoracic or head and neck procedures than abdominal. Tissue trauma, fluid shifts and blood transfusion are risk factors. Population factors for adverse outcome include age >70 years, ASA ≥ 3, smoking and congestive cardiac failure. The 30-day mortality rate after postoperative respiratory failure is 26%.

ANALGESIA AND PHYSIOTHERAPY

Postoperative needs include good deep breathing, coughing and early mobility but too much sedation will impair these activities and may be detrimental. An epidural may be useful for abdominal or thoracic surgery but high epidurals may embarrass breathing. There are also risks from co-morbidities such as arrhythmias; atrial fibrillation is common. On the second and third days postoperatively there are often recurrent hypoxic episodes that have been attributed to pharmacologically disturbed sleep patterns. Later complications include ileus and pseudo ileus; the resultant splinting of the diaphragm from a distended abdomen can be dangerous.

Nasogastric decompression of the stomach is important, particularly if NIV or CPAP are used, and if thoracic or abdominal surgery has been performed.

Incentive spirometry or intermittent positive pressure breathing are important adjuncts to physiotherapy.

POSTOPERATIVE MONITORING

The main problems are iatrogenic respiratory depression, sputum retention and respiratory failure. While some problems are immediate many occur in the days following surgery. Knowledge of normal and abnormal respiratory patterns (in particular, either fast and shallow or very slow) is crucial as they are early warning signs. Oxygenation is important and easily tracked with pulse oximetry but CO_2 is probably more important and hence an arterial line is helpful. Body temperature should be maintained as hypothermia may induce ischaemia.

OXYGEN THERAPY

Most patients are not hypoxic drive dependent and hypoxia is a greater threat than hypercarbia. In patients with chronically elevated CO_2 who are hypoxic drive dependent, too much oxygen may result in hypercarbia and narcosis. Recent work suggests that saturations of 90% or just above are likely to be safe. It is prudent to pursue a safe target rather than limit oxygen and risk hypoxia. Patients with chronically elevated CO_2 and high bicarbonate tend to be at more risk of hypercarbic narcosis. They appear comfortable at rest, not hyperinflated, with relatively preserved spirometry, gas transfer and less gas trapping.

NONINVASIVE VENTILATION AND CPAP

The aims are lung volume recruitment and maintenance of that state while normal spontaneous ventilatory function and airway clearance mechanisms are restored in the postoperative period.

Noninvasive positive pressure ventilation is the treatment of choice for AECOPD associated with hypercapnic respiratory failure not requiring emergency intubation. It reduces the risks of deteriorating respiratory failure, mechanical ventilation, infectious complications, length of hospital stay, and death, and is health economical. It is also an important weaning tool in mechanically ventilated COPD patients on ICU. Whilst its role in the postoperative period is not yet defined, it is intuitive and common to have it available for use immediately after extubation in the anesthetised patient with COPD who is at risk of postextubation compromise.

In COPD patients with associated obstructive sleep apnoea, access to their home device perioperatively is advisable. In those with suspected but undiagnosed OSA-COPD overlap, postoperative bilevel NIV with higher levels of expiratory positive airway

pressure (EPAP), which is akin to CPAP, should be used. Both treatments aid breathing and improve oxygenation, as there is an increased risk of profound postoperative desaturations in these patients, as a result of the residual effects of sedation and sleep deprivation on hypoxic arousal mechanisms.

OUTCOMES

Complications, especially pulmonary, are common. The hypercapnic group has significantly impaired function as they cannot easily clear CO_2 and may have altered drive. It is not always the disease state but comorbidities and the nature of the surgery that define outcome.

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CROSS-REFERENCES

Polycythaemia, Chapter 7
Intraoperative bronchospasm, Chapter 30
Preoperative assessment – specific medical problems, Chapter 25

CYSTIC FIBROSIS (CF)

Cystic fibrosis is the most common genetic Caucasian disease with an incidence in northern Europeans of about 1 in 3000 births. The gene involved encodes CF trans-membrane conductance regulator protein (CFTR). It functions as a chloride channel on the apical border of epithelial cells lining most exocrine glands and affects many transport systems including sodium, ATP channels, intracellular vesicle transport and bicarbonate-chloride exchange which is critical to mucin structure and activities. There have been at least 1500 mutations identified that affect CFTR function in a variety of ways, but the genotype is a poor predictor of disease severity and outcome.

Diagnosis is usually made in infancy and the sweat test is easy and reliable. A chloride concentration greater than 60 mmol/L is diagnostic. With improved intensive management of affected individuals, the median age of survival is now 38 years.

There are often some very clear 'red flags' for the diagnosis, although clinical presentation can be very varied and non-specific so a high index of suspicion should always occur. Table 1.4 identifies the common symptoms. In infancy and childhood, gastrointestinal problems are common such as meconium ileus, intussusception and pancreatic insufficiency. Respiratory problems are slightly later and infections commence during childhood. Later in childhood and adulthood the full panoply of gastrointestinal, respiratory and renal manifestations may be seen. The respiratory problems (as summarised in Table 1.5) are chronic infection, with recurrent acute exacerbations leading to bronchiectasis, and chronic colonisation often with resistant organisms. Pseudomonas is particularly likely to develop in the uncleared plaques of mucus, especially with impairment of the normal mechanisms that inhibit bacterial binding to epithelium combined with faulty immunological responses to the bacteria, which then goes on to form resistant biofilms. Airway inflammation is a notable finding. An allergic response to aspergillus fumigatus occurs in some patients.

Diabetes is a common endocrine problem, associated with many pancreatic exocrine functions.

Despite this plethora of problems, modern treatment is continually improving. Nebulised hypertonic saline, macrolide antibiotics, beta agonists and ibuprofen are useful in disease management. Hypertonic saline helps by pulling fluid into the airways and helps hydrate the peri-ciliary layer and improve mucociliary clearance.

PREOPERATIVE ASSESSMENT

Patients are always under the care of a specialist unit and always have insight and are well informed about their disease state. Ask about the normal level of function and exercise capacity, whether there are any current infective problems, cough, sputum quality and quantity or wheezing. It is important to be cognisant of pancreatic and bowel dysfunction but also any endocrine problems such as diabetes.

Key features are

- Current chest status of the CF
- Exercise tolerance
- Recent hospitalisation
- Current or recent antibiotics, including any intravenous antibiotics

Table 1.4 Clinical manifestations and surgical presentation

Infancy	Childhood	Adulthood
Infection	Sinusitis	Haemoptysis
	Polyposis; polypectomy	Pneumothorax
	Allergic aspergillosis	Infection
	Intravenous access difficulties	Sinusitis and nasal polyposis
		Allergic aspergillosis
		Need for lung transplant
Meconium ileus/peritonitis; intestinal atresia	Distal intestinal obstruction	Biliary fibrosis; obstructive jaundice – need for cholecystectomy
	Intussusception	
Pancreatic insufficiency	Biliary fibrosis	Cirrhosis; varices, coagulopathy
Rectal prolapse	Hepatic steatosis	Distal intestinal obstruction
	Malabsorption	Adenocarcinoma bowel
	Diabetes	Diabetes
Hyponatraemic hypochloraemic alkalosis	Renal calculi	Renal calculi
	Hyponatraemic hypochloraemic alkalosis	Renal failure
Dehydration		Hyponatraemic hypochloraemic alkalosis
		Vasculitis
		Hypertrophic pulmonary osteoarthritis
		Osteoporosis, fractures

Table 1.5 Respiratory pathophysiology

Reduced mucociliary clearance	Physiotherapy, multidisciplinary team care.
Mucus plugging	Mucolytics
Atelectasis	
Colonisation; *Pseudomonas*, *Staphylococcus*, *Haemophilus*, *Stenotrophomonas*, *Burkholderia cepacia* and *Aspergillus*	Aerosolised antibiotics, such as tobramycin, colistin. Targeted treatment of acute infection
Obstructive airway pattern reduced FEV_1, reduced peak flow and increased residual volumes	Beta adrenergic agents. Oral or inhaled steroids
Bronchiectasis, emphysema, fibrosis	
Apical blebs – pneumothorax	
Pulmonary hypertension	Oxygen therapy
Cor pulmonale	NIV/CPAP/BiPAP

INVESTIGATIONS

Chest X-ray looking for hyperinflation, extent of bronchovascular markings and evidence of cysts or bronchiectasis. A CT scan may be more informative.

Lung function tests may show an obstructive pattern.

Blood gases if indicated. As the disease progresses, chronic hypoxia and hypercapnia predispose to raised pulmonary artery pressures and vascular resistance which leads to right ventricular strain and cor pulmonale. These patients may require home oxygen or may be on NIV. This needs to be known so that access to their devices postoperatively is possible.

Renal and liver function should both be checked as these may deteriorate insidiously. In advanced disease, there may be abnormal clotting.

PREOPTIMISATION

Engage physiotherapists who will have a plan to ensure the patient is as good as they can be – they and the patient will know. Request physiotherapy immediately prior to going to theatre. Bronchodilators, steroids as required and hydration are all important. Current antibiotics or those recommended by microbiology for the surgery.

Bowel preparation to avoid constipation. H_2 antagonists or similar as reflux is common.

Plan the anaesthetic technique to suit the surgery. Use regional anaesthesia where possible either as the entire technique (difficult in children), or as an adjunct to general anaesthesia so emergence is rapid and pain free at the end of surgery. Try to have minimal impact on respiratory function and also plan to be able to commence physiotherapy immediately postoperatively if possible. Use humidified gases and care should be taken with any nasal tubes as most patients have hypertrophic sino-nasal mucosa with or without polyps.

Monitoring should be appropriate to fit the surgery and the patient. If there is evidence of pulmonary hypertension or impaired myocardial function, invasive monitoring may be appropriate. Watch the airway pressure as it may be an indicator of plugging or collapse. End tidal CO_2 and oximetry are both useful but may need to be supplemented by arterial blood gases and, if diabetic, the blood sugar should be monitored. In neonates, transcutaneous monitors can be used.

INDUCTION

If the patient has reflux a rapid sequence technique should be used. Pre-oxygenation followed by propofol as it wears off rapidly. Some anaesthetists will avoid nitrous oxide as there is a small risk of pneumothorax, and a high FiO_2 is often required. A volatile agent that is non-irritant is ideal. Sevoflurane has the advantage of bronchodilation and will also facilitate intubation where a minimal but adequate dose of a non-histamine releasing relaxant such as vecuronium or cisatracurium can be used.

Positive pressure ventilation is usually not a problem unless there is very severe disease. Suctioning

may be necessary to clear the secretions perioperatively. Intraoperative physiotherapy may be useful on occasions. Only extubate when the patient will breathe well and be able to cough as avoiding atelectasis is important. Prior to extubation, instillation of saline may be helpful for the physiotherapy following extubation.

POSTOPERATIVE MANAGEMENT

Rapid emergence and good analgesia, with a combination of opioids, NSAIDs and local anaesthesia where appropriate will enable early physiotherapy and mobilisation. These patients are at high risk of postoperative complications particularly pulmonary complications from sputum retention, plugging and consequent atelectasis. An enhanced recovery area is ideal unless more intensive monitoring and care is needed. If necessary, CPAP and noninvasive ventilation may be required. Positive pressure ventilation can produce significant problems in these patients with barotrauma and a tendency to air trapping, detrimental changes in V/Q and increasing dead space so it is best avoided if possible.

Proper hydration and opiate-sparing techniques may avoid the complication of distal intestinal obstruction.

PREGNANCY

The normal physiological changes of pregnancy, such as increased minute ventilation and oxygen requirements, may stress respiratory function while fluid shifts may exacerbate problems with right ventricular strain. Prognostic factors include weight gain <4.5 kg, FVC <50%, colonisation with *B. cepacia*, frequent respiratory infections and hospitalisations, diabetes and pancreatic insufficiency. If there is evidence of cor pulmonale, this is likely to get much worse with pregnancy and there is a recognised mortality.

Regional techniques are clearly preferable, in particular combined techniques where a good block can provide postoperative analgesia. There are circumstances in patients with severe disease where this may not be feasible but the decision to use general anaesthesia should not be taken lightly in particular if it may exacerbate the incipient respiratory infection and failure.

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CROSS-REFERENCES

Infants and children, Chapter 24
Medical problems in obstetric anaesthesia, Chapter 12

RESTRICTIVE LUNG DISEASE

A range of conditions produces a restrictive picture on lung function with reduced total lung capacity, reduced resting volume yet often with normal airways resistance and airflow.

Restrictive lung diseases may be classified as intrinsic or extrinsic. Intrinsic restriction is characteristic of a group of over 200 diverse conditions affecting the pulmonary interstitium (i.e. the space bounded by the alveolar epithelium and the pulmonary capillary bed and including the perivascular and perilymphatic tissues) and encompassed

by the term diffuse parenchymal lung disease (DPLD) (Figure 1.2). These are usually characterised by impaired gas transfer factor and reduced gas transfer coefficient (Kco), as a result of impaired exchange between alveolar–capillary units within the interstitium.

The most commonly encountered DPLDs in clinical practice are the so-called idiopathic interstitial pneumonias (IIP), which separate into idiopathic pulmonary fibrosis (IPF) (previously known as cryptogenic fibrosing alveolitis), and the non–IPF diseases, which generally have a better prognosis. Other DPLDs are subclassified as granulomatous, exposure related (organic or inorganic), drug and radiation induced, associated with collagen vascular or rheumatological diseases, pulmonary-renal and vasculitides, and rare orphan diseases (e.g. histiocytosis X, lymphangioleiomyomatosis)

Extrinsic restriction of lung function is usually associated with reduced gas transfer but normal or increased gas transfer coefficient corrected for lung volume (Kco). Essentially, the reduced lung volumes are due to limited excursion of the chest wall, pleura or neuromuscular impairment of the respiratory system. Note that left ventricular dysfunction is also a cause. Table 1.6 summarises the main causes of intrinsic and extrinsic restrictive lung diseases.

PATHOPHYSIOLOGY

The volume of the functional residual capacity (FRC) is determined by the balance of inward elastic recoil of the lungs and outward elastic recoil of the chest wall. Impairment of either will restrict movement and result in a lower FRC. Total thoracic compliance is the combined compliance of lung and chest wall which is reduced. Particularly in advanced DPLD, there is V/Q mismatch and oxygen transfer reduction, leading to hypoxaemia. This is often apparent earlier following exercise.

The restrictive nature of the system means that smaller tidal volumes necessitate a higher respiratory rate to maintain effective minute ventilation and acid base homeostasis. This is generally true in intrinsic disease but extrinsic restrictive conditions such as neuromuscular disease or obesity have a propensity to respiratory muscle fatigue and alveolar hypoventilation, which may over time lead to type 2 or hypercapnic respiratory failure and pulmonary hypertension. The efficiency of ventilation is reduced

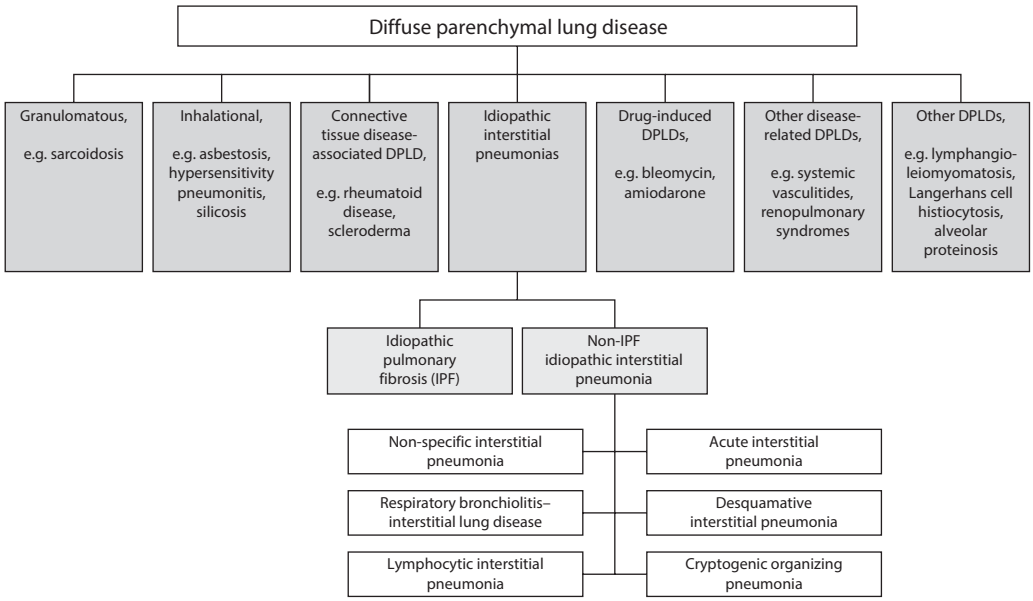

Figure 1.2 A classification scheme for diffuse parenchymal lung disease (DPLD).

Table 1.6 Restrictive lung diseases

Type	Mechanism	Condition
Intrinsic	DPLD	See Figure 1.2
Extrinsic	Limitation of chest wall excursion	Kyphoscoliosis, ankylosing spondylitis, thoracoplasty, pleural effusion, obesity
	Respiratory muscles/neuromuscular	Polio, Guillain–Barre, muscular dystrophy
	Pleural thickening	

by the smaller volumes as the effective dead space rises in relation to the tidal volume. The underlying disease process will further add to lung dysfunction.

ANAESTHESIA

The problems posed are the restricted lung volumes which reduce the ability of the lung to respond to stress. There is limitation of gas transfer and a predisposition to infection. Compliance is reasonable over a limited range of lung volumes above which it reduces dramatically so ventilation must remain within these limited volumes.

PREOPERATIVE PREPARATION

It is important to elicit the underlying cause of the restrictive picture.

HISTORY

With likely DPLD, exertional breathlessness, cough and reduced exercise tolerance may be apparent depending on the type and severity of disease. The history should elucidate the exercise tolerance and the degree of dyspnoea at rest and on exercise. Features of pulmonary hypertension and right ventricular failure such as ankle oedema may be present. Viral prodromal-like respiratory illnesses often characterise the clinical history and may be difficult to distinguish from respiratory tract infections. The past medical history should identify disorders associated with DPLD (e.g. rheumatoid arthritis and connective tissue disease). Radiotherapy for breast or thoracic malignancy can result in pulmonary fibrosis. Patients with a past history of granulomatous disease, e.g. ulcerative colitis, are at increased risk of developing sarcoidosis.

Chemotherapy such as bleomycin and other drugs such as amiodarone, methotrexate, gold, and homeopathic or complementary medications can cause DPLD.

Occupational history of exposures (organic and inorganic), and systemic features that may indicate connective tissue, vasculitis or rheumatological disease should be determined.

With suspected extrinsic diseases, weight-related problems, sleep-disordered breathing, left ventricular failure and neuromuscular weakness should be asked about.

EXAMINATION

Assess the degree of dyspnoea, look for cyanosis and evidence of finger clubbing (indicative of idiopathic pulmonary fibrosis). Look for features of systemic disease such as Raynaud's or polyarthropathy. There may be fine bilateral 'velcro-like' crackles heard on auscultation. Evidence of pulmonary hypertension, right heart dysfunction (i.e. loud pulmonary second heart sound, tricuspid regurgitation, raised jugular venous pressure [JVP] and ankle swelling), oropharyngeal indicators of sleep apnoea and left ventricular dysfunction should be excluded.

INVESTIGATIONS

Chest X-ray often shows a reticulonodular appearance, with characteristically small lung fields. The distribution of changes is indicative of the aetiology. Upper zones are associated with granulomatous or acute exposure-related DPLD. Lower zone predominance is usually related to the idiopathic interstitial pneumonias. Honeycombing and loss of clarity of the heart borders is generally a sign of advanced disease.

High-resolution CT scan is the diagnostic investigation of choice in suspected DPLD. The patterns of distribution of ground glass, interstitial thickening, traction bronchiectasis, and consolidative conglomerates are often sufficient to allow diagnosis without need for lung biopsy. However, this is usually in the context of secure clinical features and ultimately the profile of longitudinal functional behaviour.

Respiratory function tests show decreased vital capacity and FEV_1 so the ratio remains normal. FRC is reduced. The carbon monoxide diffusing capacity (DLco) is reduced in intrinsic lung disease as is the gas transfer coefficient Kco. Progressive decline in DLco (<40% predicted) is an independent predictor of poor prognosis in idiopathic interstitial pneumonia. Preserved or high Kco associated with low DLco is evidence of extrinsic disease. In neuromuscular disease, maximum inspiratory pressures (both volitional 'sniff' and nonvolitional diaphragm studies) are dramatically reduced. The vital capacity (VC) is a helpful serial measure of progression of DPLD (especially if >10% change). In neuromuscular weakness, a serial fall in VC may warrant a discussion about assisted ventilation in the acute or postoperative setting.

In patients with coexistent emphysema, lung volumes may be preserved. A mixed obstructive/restrictive defect may sometimes be seen in sarcoidosis, lymphangioleiomyomatosis (LAM), respiratory bronchiolitis interstitial lung disease (RB-ILD) and hypersensitivity pneumonitis.

Arterial blood gases may show hypoxaemia. CO_2 rises in extrinsic disease and sometimes with advanced DPLD.

Exercise tests such as the 6 minute walk test are useful in IPF. Desaturation to 88% or 200 m portend a poor prognosis. Exercise tolerance is reduced so exercise testing with oximetry will indicate oxygen requirement and can be used to follow disease progression.

PREOPERATIVE OPTIMISATION

Reverse any airflow limitation with bronchodilators; steroids may be needed. Treat cardiac failure appropriately well in advance of surgery. Treat any possibility of infection. If there are limiting factors such as pleural effusions, then drainage may be very helpful. Involve the physiotherapists. A cardiological opinion is essential. If pulmonary hypertension may be present, perform echocardiography.

THE ANAESTHETIC

Plan the anaesthetic in terms of the procedure and the limitations of the patient. Consider if the procedure is amenable to regional technique. If a regional technique is used, then beware the height of the block may impair ventilatory muscle function, both chest and abdomen, so not above a level of T10 depending on the patient.

GENERAL ANAESTHESIA

Some advocate anticholinergic agents. Monitoring should encompass oximetry, capnography and the ability to do blood gas sampling. Cardiovascular monitoring will be defined by the cardiovascular stability of the patient and the nature of the procedure. In patients with kyphoscoliosis or ankylosing spondylitis, difficult intubation should be anticipated. A further problem in these patients with chest wall abnormalities is surgical positioning.

Ventilation may be difficult. Small tidal volumes will be necessary and if exceeded can result in very high airway pressures and a risk of pneumothorax. Oxygenation may also be problematic despite ventilation so high inspired oxygen may be necessary.

POSTOPERATIVE MANAGEMENT

As with other severe lung diseases, the postoperative period is a potential source of problems. Sputum retention and basal atelectasis will both contribute to the restrictive picture and may have significant effects on the already poor lung function. Extubate when compliant and awake so that coughing, mobilisation and physiotherapy are possible early. Adequate analgesia is essential with the usual difficult balance between analgesia and sedation. A high dependency area is ideal postoperatively.

Beware of hypoxia and of insidious hypercapnia. Noninvasive ventilation, either CPAP or bilevel, can be used to facilitate postoperative lung volume recruitment and relieve the work of breathing as necessary.

The physiotherapists are key to the management for several days postoperatively until the patient is fully mobile. Adjuncts like incentive spirometry, intermittent CPAP or intermittent positive pressure breathing may be helpful as bridges to recovery.

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SARCOIDOSIS

Sarcoidosis is a systemic, granulomatous disease of unknown aetiology. It seems likely that the granulomas form through an interaction between antigens, as yet unknown, and T cells. It has geographical variation. Slightly more common in women, its peak onset is in the twenties and thirties. Presentation is variable but 90% of patients have lung involvement, often with bilateral hilar lymphadenopathy or pulmonary infiltrates. Skin, lymph node, eye and liver are the next most affected organs in that order. Cardiac involvement is less common but potentially fatal. There may be radiological appearances, particularly involving the small bones of the hand and feet, or symmetrical arthritis of large joints. Occasionally there is neurological involvement.

While imaging and a plethora of tests can imply sarcoid, such as elevated ACE levels, a raised calcium, raised immunoglobulins and 'gallium lit' lesions, the only real way to diagnose the condition is by biopsy which will show noncaseating granulomas. TB and fungal infection are often the differential diagnosis.

The implications to the anaesthetist mainly relate to the cardiac and pulmonary involvement which may involve fibrotic lung changes and a restrictive pattern usually with reduction of diffusing capacity. Most patients will have an abnormal chest X-ray at some stage in the disease and usually hilar lymphadenopathy. Occasionally there may be obstructive lesions in the airways themselves.

There may be nasopharyngeal and laryngeal involvement affecting the arytenoids and supraglottic area and patients occasionally present with dysphonia, then stridor and dyspnoea which may necessitate emergency tracheostomy.

Cardiovascular involvement is an uncommon manifestation in clinical practice at 2%, but 25% of postmortem examinations of known cases of sarcoid have cardiac involvement. Preferential granulomatous involvement of the conduction system is manifested as a variety of dysrhythmias, including complete heart block. Congestive cardiac failure with features of a dilated cardiomyopathy may also be present.

Renal involvement is uncommon at less than 2%. It may occur through hypercalcaemia or nephrocalcinosis or both. It may also cause either interstitial or membranous nephritis.

Hepatic and pancreatic involvement have been reported.

The neurological system may be affected in 5%–15% of patients with sarcoid although, again, the postmortem evidence suggests far more. Most common are cranial nerve palsies, which account for 65% of the neurological manifestations. Headache is also common but fitting is uncommon. Rarely, mono- or polyneuropathies can develop which may cause sensory or motor deficit, or a combination of both. Cerebellar symptoms can also occur.

Neuropsychological disturbance is also uncommon. Spinal involvement is rare but may present with various forms of paresis including cauda equina syndrome.

PREOPERATIVE ASSESSMENT

In such a protean disease it is hard to suggest a universal approach. A clear history of the range of problems that are known should be elicited but awareness of occult cardiology and neurology should be borne in mind.

The prevalence of respiratory system involvement indicates careful respiratory assessment. Specific attention is focused on a history of stridor (suggesting laryngeal involvement), swallowing difficulties (hinting at neurological problems), or any breathlessness indicating the more common interstitial-type lung disease. A chest X-ray and blood gases will be useful to identify any overt respiratory issues. Pulmonary function tests may help clarify degrees of restrictive lung injury defects. The covert nature of cardiac involvement mandates taking a history of any palpitations or fainting episodes, an ECG to look for any signs of actual or potential heart block, and echocardiogram to assess cardiac function. Pacing may be indicated.

Renal function is relatively easy to assess as is the measurement of calcium looking for hypercalcaemia. Neurological involvement needs to be elicited especially if it is intended to use a regional technique. Table 1.7 summaries the most likely pre-operative findings.

Table 1.7 Investigations and results

Investigations	Results (if system involved)
Chest X-ray	Bilateral hilar lymphadenopathy, reticulonodular shadowing, pleural effusions, cardiomegaly, atelectasis
Electrocardiogram	Conduction defects, ventricular hypertrophy
Arterial blood gases	Reduced Po2 on room air
Lung function tests	Restrictive/obstructive defects
Electrolytes	Raised calcium/potassium
Echocardiography	Ventricular hypokinesis, mitral valve involvement, septal thickening and bright echoes (consistent with fibrogranulomatous infiltration)

PERIOPERATIVE ANAESTHETIC MANAGEMENT

Given the massive range of potential problems that may be an issue, it is vital to tailor the anaesthetic to the patient. If feasible, the use of a regional technique may be advantageous in the presence of significant respiratory disease but may be difficult if there is neurological involvement. Laryngeal involvement with stridor is a special case that needs careful planning. The most common problem generally is the respiratory system. As with other respiratory conditions, caution with sedation is advised. Avoiding and preventing hypoxia is the aim and the liberal use of supplemental oxygen is recommended. These patients may already be on steroids, but if not, steroids may be of benefit.

POSTOPERATIVE MANAGEMENT

If the predominant area of risk is respiratory, the focus should be on good analgesia, mobilisation and physiotherapy. Renal issues may need attention to avoiding prerenal insults. With such a myriad of presentations and potential problems, there is no specific area in which sarcoid is different from its component parts. The role of steroids and other agents needs careful attention as these may need to be continued and probably increased in the postoperative phase.

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CROSS-REFERENCE

Preoperative assessment of pulmonary risk, Chapter 25

ANAESTHESIA AND SLEEP APNOEA SYNDROME (SAS)

In obstructive sleep apnoea (OSA), breathing during sleep is periodically interrupted by closure of the upper airway for 10–45 second intervals. This partial obstruction results in periods of reduced ventilation. Most individuals with OSA have a combination of apnoeas and hypopnoeas and respiratory effort-related arousals. The result is fragmented sleep which leads to excessive daytime sleepiness, fatigue, or poor concentration. Partners comment on apnoeas, snoring, restlessness and resuscitative snorts.

Grading of severity is based on the frequency of apnoeas and hypopnoeas per hour (apnoea/hypopnoea index [AHI]) and consequent symptoms. Alternatively, the frequency of dips in oxygenation during sleep is used (oxygen dip rate). AHI 5–14 correlates with mild symptoms, AHI 15–30 moderate and AHI > 30 severe.

Sleep apnoea can also be defined as obstructive (cessation of flow in the presence of respiratory effort), central (no flow and no effort) or mixed (a combination of the two).

Risk factors for OSA include obesity, craniofacial abnormalities and upper airway soft tissue abnormalities. Potential risk factors include heredity, smoking, nasal congestion and diabetes mellitus.

SAS affects 2%–4% of middle aged males and 1%–2% of adult females. Only 20%–30% of affected individuals have currently been diagnosed in the UK and the prevalence is increasing with increasing obesity.

Patients are at increased risk for organ system dysfunction and impaired neurocognitive performance due to chronic nocturnal hypoxaemia and repeated arousals over months and years.

An increased incidence of cardiovascular and cerebrovascular events and an emerging association with endocrine abnormalities including diabetes and sex hormone dysfunction exists due to chronic hypoxaemia-related microvascular dysfunction. Patients with SAS have a 7–10 times increased chance of road traffic accidents compared with other drivers.

Patients with OSA have an increased risk of peri- and postoperative complications. Prolonged apnoeic events may follow reduced consciousness due to iatrogenic loss of protective arousal mechanisms that overcome upper airway obstruction. Severe respiratory complications and unexpected deaths have occurred. The possibility of REM rebound-associated nocturnal desaturations, perhaps after hospital discharge, increases further the risk of unrecognised or untreated OSA. Preoperative diagnosis and optimisation are important. Three scenarios are identifiable:

- Known SAS on treatment
- Known SAS non-compliant with treatment
- Undiagnosed SAS

This last group, particularly who may not have typical features of SAS, can present with difficult airways, difficult intubation, or hypoxaemia postoperatively. Patients with clinical features suggestive of SAS and those with unexplained hypoxaemia, polycythaemia or pulmonary hypertension warrant diagnostic sleep testing. It is likely that the majority of patients with SAS are undiagnosed or untreated. The prevalence of SAS is thought to be very high in the morbidly obese, perhaps up to 70%. However, factors predicting this have poor specificity and not all patients in this category have SAS.

The most effective treatment for symptomatic SAS is CPAP which improves symptoms and reduces

cardiovascular comorbidity in those using it effectively. However, adherence remains a challenge in more than 30%–40% of patients. They should be managed as part of a specialist sleep clinic.

OBSTRUCTIVE SLEEP APNOEA

Breathing is normally a function of generating a negative pressure in the thorax and entraining air. Narrowing of the airway associated with a tendency for tissues to collapse inwards are the key features for obstruction. The three areas in the pharynx subject to collapse are the retropalatal pharynx, the retroglossal pharynx and the retroepiglottic pharynx. Narrowing may be due to excess soft tissues in obesity and OSA correlates with neck circumference.

Narrowing of the airway produces increased turbulence and local tissue vibration while increased velocity through narrow passages generates a Bernoulli effect pulling tissues inwards (see Figure 1.3). When conscious, muscle tone prevents this but a reduction in muscle tone, through sleep or sedatives, may be all that is required for loss of a patent airway. In those predisposed to OSA, normal nocturnal tone and function of the dilator muscles is impaired. This may be due to both turbulent flow and a deficiency of orexins (neurohormones that govern wakefulness).

REM sleep is associated with decreased muscle tone which recovers as the patient wakes and then returns as they fall asleep again. There are usually four periods of NREM sleep which culminate in an episode of REM with its marked slowing of the EEG. Drugs that influence muscle tone either through peripheral or central actions may increase obstruction. Predisposing factors to the development of SAS are summarised in Table 1.8.

CENTRAL SLEEP APNOEA

This is a control effect and so is associated with conditions affecting ventilatory drive, e.g. neuromuscular disorders or neurological damage of the respiratory centre through stroke or head injury. Obesity may also affect the chest wall and reduce lung volume. Sedatives or opiates, which inhibit central control, will worsen these effects.

The sequence of physiological events that follows an obstructive apnoea is

- A fall in oxygen tension.
- A rise in carbon dioxide tension.

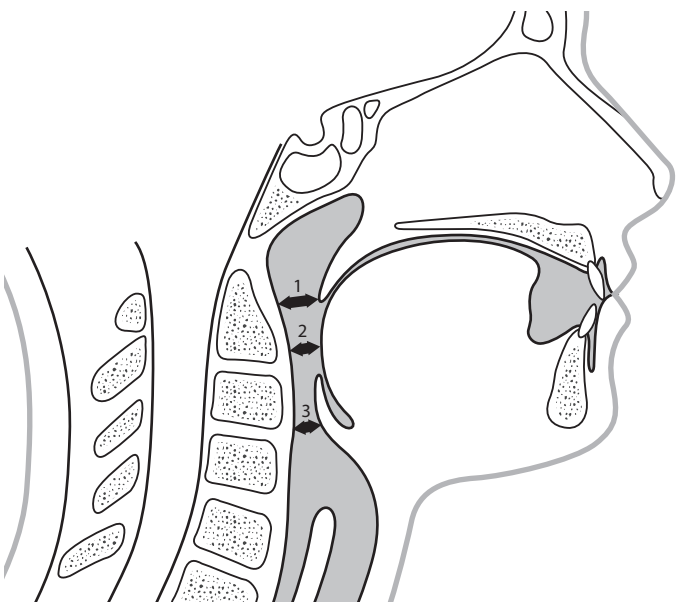

Figure 1.3 Airway obstruction in sleep apnoea. 1, Nasopharynx – tensor palatine; 2, oropharynx – tongue enlargement and posterior tissues; 3, laryngopharynx – tissues around epiglottis and base of tongue.

Table 1.8 Predisposing conditions for SAS

Condition	Mechanism
Obesity; acquired; genetic (Prader–Willi syndrome)	Soft tissues, anatomical
Smoking	Pharyngeal inflammation and oedema
Nasal obstruction	Polyps, septal deviation, nasal congestion
Laryngeal tracheal obstruction	Laryngo/tracheo malacia
Neuromuscular; cerebral palsy, stroke, head injury	
Endocrine; thyroid Cushings acromegaly	Soft tissue
Connective tissue; Marfans syndrome	Upper airway tissues
Mucopolysaccharidoses	
Craniofacial; Pierre Robin, Treacher-Collins, Downs	

- Increasing ventilatory effort.
- Increasing negative inspiratory airway pressure.

All four mechanisms trigger 'reflex' activity, increase EEG activity (seen as twitching, movement, etc.) and cause arousal.

Immediate effects during an apnoea include:

- Low PaO_2 associated with tachycardia or bradvcardia.
- Associated nocturnal angina, and myocardial infarction.
- Diurnal pulmonary and systemic hypertension.

Consequences of SAS include excessive daytime sleepiness, impaired concentration, mood changes, morning headache, waking with a choking sensation and dry mouth. Signs include snoring, excessive daytime sleepiness, nocturnal sweating and witnessed apnoea. Secondary effects include polycythaemia, pulmonary hypertension and right heart failure. In patients with excess weight there are likely to be significant comorbidities. Other nonspecific effects include gastro–oesophageal reflux, hypertension, ischaemic heart disease and in diabetics increased instability. There is an increased incidence of sudden death in untreated patients with SAS, compared with age-matched controls.

SLEEP, ANAESTHESIA AND APNOEA

REM sleep is the time with the most influence on sleep apnoea. In neonates it accounts for up to 50% of sleep while by middle age it is about 20%. It diminishes with age or with medications such as antidepressants.

THE PERIOPERATIVE IMPACT OF SLEEP APNOEA

Repetitive episodes of upper airway obstruction during sleep, with sleep disruption, hypoxaemia and autonomic arousals, contribute to cardiovascular risk. Anatomic narrowing in the pharynx due to excess tissue, tonsillar hypertrophy or craniofacial variations can lead to airway difficulties. Desaturations of sufficient intensity may precipitate arrhythmias or acute coronary syndrome in susceptible individuals. The CNS depressant effects of sedatives, analgesics and anaesthetics suppress the natural arousal mechanism induced by hypoxaemia or hypercapnia in patients with SAS leading to prolongation of postoperative apnoeic episodes. Another potential concern is the impact of restoration of sleep after a period of perioperative sleep deprivation. The phenomenon of rebound REM sleep, with its associated profound desaturations may be under-recognised in the postoperative period.

Although no prospective randomised trials of anaesthetic risk in patients with SAS exist, there are reports of postoperative cardiac arrhythmias, myocardial infarction, cerebrovascular events and hypoxaemia-induced organ dysfunction. There have been sporadic reports of fatalities in patients with OSA in the postoperative period.

THE EFFECT OF SEDATION AND ANAESTHESIA ON PATIENTS WITH SLEEP APNOEA

The effects of sedatives and sedating analgesics (e.g. opioids) mimic those on the upper and lower

respiratory tracts in sleep. There is a reduction in FRC and atelectasis. This has potential implications for preoxygenation. In patients with OSA associated with obesity, this reduced pharyngeal anatomical space, together with the functional disturbance of the dilator muscles (particularly genioglossus), is accompanied by a reduction in lung volumes as a result of fat distribution around the diaphragm in central obesity. This may reduce the traction on the pharynx exerted by the trachea. The usual neural mechanisms in wakefulness, to compensate for these anatomical imbalances, are lost during sleep. The pharynx is more susceptible to closure in these patients, potentially exacerbating the upper airway risk.

Sedatives reduce the phasic activity of pharyngeal muscles just prior to inspiration, mimicking the response to REM in patients with OSA. Thus, during general anaesthesia, there is a loss of the protection against upper airway collapse (caused by the lower respiratory tract muscles generating negative pressure on the airway). Moreover, sedatives depress the compensatory arousal responses to hypoxia, hypercapnia and upper airway collapse that characterise the repeated sleep/wake cycle in OSA (Figure 1.4). The risk of prolonged apnoeas and desaturation then increases, as has been noted in many patients with OSA undergoing sedation.

In the postoperative period, residual central depressant effects of these agents may cause prolonged apnoeas and desaturation when reduced monitoring is present. Disruption of sleep has also been documented in postoperative periods following surgery. Thus, reduced total sleep time with less REM and non-REM slow wave sleep are reported, which may take several days to return to normal.

ASSESSMENT

History

SAS is still predominantly under-recognised. Look for a history of snoring, daytime sleepiness or lethargy and witnessed apnoeas. Identify other features such as depression, neurocognitive or functional decline. Seek a corroborative history from a partner,

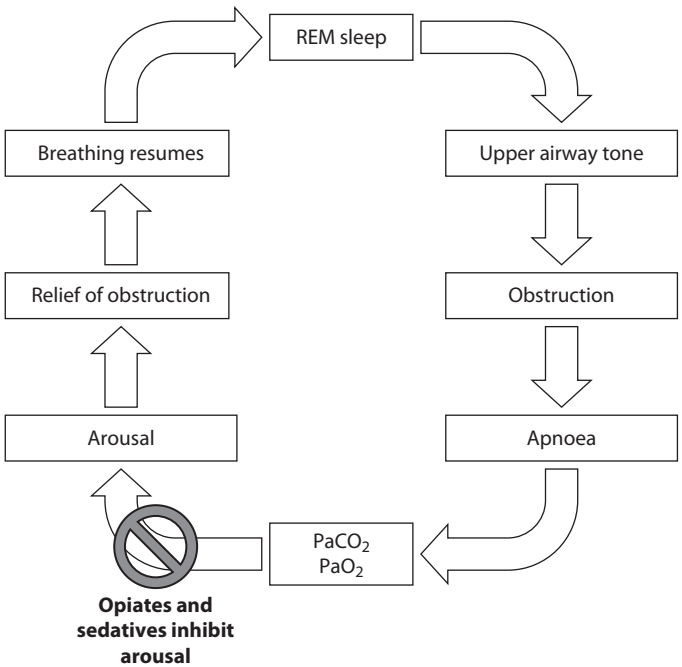

Figure 1.4 The pathophysiology of obstructive sleep apnoea and how sedatives can suppress the natural arousal responses of hypoxaemia and hypercapnia. REM, rapid eye movement.

as patients will often deny symptoms due to insidious onset or being unaware of their own sleep disturbance. Identify comorbidities (e.g. hypertension, diabetes mellitus) and risks (skilled mechanical work, HGV driving, pilots, etc.). The Epworth sleepiness scale is a self-reported indicator of sleepiness, when other confounders to adequate sleep are excluded (e.g. prostate problems, symptomatic nocturnal acid reflux, noise or light disturbance). It has 8 questions of different situations each with a weighted scale of 0–3 of likelihood of dozing off. A score >10/24 indicates a pathological reason for excessive sleepiness. However, it does not correlate well with severity of SAS. Other questionnaires have been validated in different settings such as the Berlin and STOPBANG scoring models.

Physical examination

Look for signs of excess soft tissue, retrognathia, short distance between hyoid and mandible, crowded oropharynx, kissing tonsils, or thick uvula. Look for signs of undiagnosed or undertreated hypothyroidism, pulmonary hypertension, polycythaemia due to chronic hypoxaemia or right heart failure.

Investigations

If surgery is not urgent, then investigation of severity and associated comorbidity should be undertaken. Referral to a sleep clinic requires overnight polysomnography with sleep staging. However, a limited respiratory multichannel study without sleep staging can be performed at home, and is diagnostic when the clinical probability of SAS is high. Information it provides includes the number, duration and severity of obstructive events and levels of desaturation, average, nadir and duration. Heart rate, ECG, actigraphy (movement as a surrogate for wakefulness) and position are also available. In the absence of other confounders such as underlying lung disease, the characteristic episodic desaturation pattern of overnight oximetry is sufficient for a diagnosis of OSA.

If OSA is diagnosed some advocate using CPAP preoperatively to 'train' the patient for its use postoperatively. There is also some evidence from dynamic imaging to suggest a remodelling of the upper airway after 8–12 weeks of effective use of CPAP. In long-term CPAP use, they should bring their own devices into the hospital for use pre- and postoperatively.

The risk of postoperative problems in patients with SAS having peripheral surgery is twice that of normal patients.

ANAESTHESIA

SAS is a risk factor for a difficult airway and difficult intubation. Large tongue, limited mouth opening, large tonsils and short neck indicate caution. Prepare for a difficult intubation. Intraoperative end tidal CO_2 monitoring may act as a guide to potential use of postextubation assisted ventilator requirements (i.e. CPAP or BIPAP).

PREMEDICATION

Avoid premedication if possible. Benzodiazepines contribute to loss of muscle tone and predispose to sleepiness postoperatively. If essential, it should be given where the patient can be on the CPAP machine or at least closely observed and where the agent can be reversed. More important is discussion with the patient of what is to be done and good preoxygenation. Most patients will be able to be intubated and difficulty is fortunately uncommon.

INDUCTION

Use a local or regional technique if possible. All sedative agents depress the respiratory reflexes and reduce muscle tone. The only exception may be dexmedetomidine.

If a general anaesthetic is required, then use controlled ventilation. Patients can breathe spontaneously but need close monitoring, will need CPAP to prevent atelectasis and with most techniques will remain drowsy postoperatively. Use short-acting agents.

SURGICAL ISSUES

Thoracic or abdominal surgery compromises the chest. Some procedures such as uvulopalatopharyngoplasty and tonsillectomy result in upper airway problems postoperatively. There may be physical narrowing through haematoma, oedema or

bleeding. Analgesics and opiates increase respiratory depression. Nasal surgery with packs may impair the airway dramatically and pose problems from apnoea and from the packs. Occasionally it will be sensible to leave the patient intubated until they can be extubated while awake. Rarely the safe option will be elective tracheostomy.

Extubate the patient semi-upright especially in the obese patient with OSA. Complete reversal of neuromuscular block is important. Obstruction postextubation is more common and there is a risk of negative pressure pulmonary oedema.

Pharyngeal oedema has been reported. The cause is uncertain but a small amount of swelling may be critical in a narrowed airway. Haemodynamic instability may need controlling with beta blockers and other antihypertensives.

POSTOPERATIVE MANAGEMENT

Supplemental oxygen is important. The balance between analgesia and good breathing is important. These patients need careful monitoring in the postoperative phase until their analgesic requirements are minimal and they are mobilising. The use of effective CPAP will allow adequate analgesia. In patients with SAS and obesity hypoventilation, type 2 respiratory failure can ensue. They are also more prone to greater desaturations, even on CPAP. These patients should have been identified, and provision for noninvasive ventilation planned.

Early complications include airway obstruction, bleeding into the airway, vomiting and aspiration and respiratory depression with airway obstruction. Later complications include insidious respiratory depression with hypercapnia, increased episodes of hypoventilation or even episodes of apnoea and hypoxaemia. Later effects include those of basal atelectasis with late secondary chest infection.

Patients with home CPAP should continue its use. Some patients may benefit from preoperative training. In the early phase postextubation it may impair coughing, suctioning, communicating and give a false sense of reassurance. It may be counterproductive and the real benefit comes in later in the recovering patient, particularly for sleeping at night.

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CROSS-REFERENCES

Preoperative assessment – specific medical problems, Chapter 25

Preoperative assessment of risk, Chapter 25

Effect of general anaesthesia on the upper airway and alimentary canal, Chapter 26

SMOKING AND ANAESTHESIA

Recognition of the problems of smoking has taken a long time and is only just beginning to alter behaviour; therefore, a significant proportion of patients will be smokers or have a long, but recently stopped, smoking history.

PATHOPHYSIOLOGY

The effects of smoking on the respiratory system include:

- Airway hyper-reactivity, especially small airways
- Reduced mucociliary clearance
- Increased mucus secretion
- A change in epithelial permeability
- Altered surfactant and hence compliance
- Small airway narrowing
- V/Q mismatch

The effects of smoking on the cardiovascular system include:

- Hypertension (due to chronic nicotine exposure and atherosclerotic change).
- Increased catecholamine levels (15–50 ng/L).
- Reduced oxygen uptake and shift in the oxygen dissociation curve to the left caused by carboxyhaemoglobin.
- Increased haemoglobin values in long-standing smokers secondary to relative hypoxaemia and carboxyhaemoglobin.
- A predisposition to thrombosis due to the increased haemoglobin together with damage to vascular endothelium. Endothelin is released resulting in a negative effect on nitric oxide dynamics and altering superoxide production. It acutely affects clot dynamics and thrombin structure and thus is thrombogenic. Curiously, cigarette smoke may have a synergistic effect with clopidogrel reducing platelet aggregation but in general should be considered thrombogenic.

The immunological effects of smoking are diverse but include reduced phagocytic and cytotoxic T-cell activity. There is some evidence for impaired immune defences.

Smoking is associated with enzyme induction so there may be an altered response to some drugs although the clinical relevance of these effects is questionable.

In the postoperative phase, smokers are more prone to hypoxaemia, have slightly higher PCO_2, have more change in pulmonary function tests, with a reduction in FEV_1/FVC ratio suggesting greater small airway obstruction. Pulmonary complications were doubled in one series and in major surgery time to extubation, ICU stay and hospital stay were all increased.

PREOPERATIVE ASSESSMENT

- Ideally, a patient should have stopped some weeks previously.
- Examine for chronic respiratory disease.
- Examine for cardiovascular disease, especially hypertension and ischaemic heart disease.
- There is an association with excess alcohol intake.
- In heavy smokers with airway disease, nutritional state may be affected.

INVESTIGATIONS

Routine investigations according to local policy (Table 1.9). Depending on the clinical picture, also consider:

- ECG with echocardiography if indicated
- Pulmonary function tests
- Arterial blood gases
- CPET
- Look for signs of infection

PREMEDICATION

An anticholinergic may be helpful as these patients will probably have irritable airways due to hypersecretion.

In those who have given up, there may be signs of nicotine withdrawal with agitation so anxiolytics may be considered.

H_2 antagonists or antacids should be considered.

Table 1.9 Effects of heavy or long-term smoking on preoperative investigations

Investigation	Result
Full blood count	Increased haematocrit
ECG	Signs of ischaemic heart disease
Chest X-ray	Chronic airways limitation Infection Malignancy
Arterial blood gases	Hypoxaemia, hypercarbia
Lung function tests	Decreased FEV_1, FVC, PEFR

PERIOPERATIVE MANAGEMENT

MONITORING ACCORDING TO AAGBI GUIDELINES

Oxygen saturation monitor may overestimate SaO_2 if there is significant carboxyhaemoglobin content. Smoking greater than 20 cigarettes a day is associated with a carboxyhaemoglobin <4% and this will fall fairly rapidly after stopping smoking, so the P_{50} will be returning to normal at 12 hours, or faster if receiving an increased inspired oxygen concentration. Table 1.10 highlights several benefits of smoking cessation.

If there is evidence of impaired respiratory function or heavy smoking right up to the point of anaesthesia, then increased FiO_2 is recommended.

CHOICE OF ANAESTHETIC TECHNIQUE APPROPRIATE TO THE PATIENT AND THE PROCEDURE

In the presence of airway problems associated with smoking, a regional technique may be preferred where feasible and may allow more effective physiotherapy postoperatively.

GENERAL ANAESTHESIA

Main considerations are the irritable airways and there may be a nicotine-mediated exaggerated pressor response to intubation. This may be obtunded with lidocaine applied locally. Spontaneous breathing, unless deep, may be problematic with coughing.

Table 1.10 Benefits of stopping smoking in the perioperative period

Time before surgery	Benefit
2 h	Nicotine blood levels fall
12 h	Carbon monoxide blood levels fall
Days	Sputum volume reduced; Haematocrit falls
Weeks	Ciliary activity restored towards normal Epithelial permeability returns towards normal
Months	Immune system recovery Drug metabolism restored towards normal

POSTOPERATIVE MANAGEMENT

This should reflect the comorbidities. Early active physiotherapy should be instigated if there are chronic lung problems. The risk of secondary infection is increased as is the likelihood of postoperative complications generally. It is wise to continue enhanced oxygen by mask for 24 hours minimum. The incidence of postoperative nausea and vomiting is reduced amongst smokers.

STOPPING SMOKING AND POSTOPERATIVE OUTCOME

The issue of smoking and complications is more contentious than it appears. Most studies show current smokers to have higher complication rates than non-smokers or previous smokers. The complication rate in some series amongst smokers is doubled. Wound healing is also impaired. Not all studies show this; for lung resection for carcinoma, the incidence of complications was not different between those who stopped smoking and those that did not. This was seen as a reason not to delay surgery. It is reasonable to assume smoking is associated with more postoperative complications and impaired wound healing.

More difficult is the issue of advice as to when to stop smoking (Table 1.10). Stopping 6–8 weeks prior to surgery is beneficial in terms of reduced

complication rate (52%–18%), but stopping for less time might be detrimental. Benefit has also been demonstrated for 3–4 weeks. One study has shown no benefit but no detriment for cessation at 1–3 weeks. Current advice should be that postoperative complications are reduced and wound healing improved by stopping smoking even for as little as 3 weeks. Shorter intervals may not be helpful but have no detriment. This was not seen in a recent paper in patients undergoing thoracotomy.

In 1944, Morton reported a sixfold increase in the incidence of postoperative respiratory morbidity in smokers over non-smokers. These findings have been confirmed in several other studies more recently. Every opportunity should be taken to discourage smoking in the perioperative period.

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CROSS-REFERENCES

COPD and anaesthesia, Chapter 1

Ischaemic heart disease, Chapter 2

Preoperative assessment of cardiac risk, Chapter 25

Preoperative assessment of pulmonary risk, Chapter 25

2 Cardiovascular system

REDMOND P TULLY AND ROBERT TURNER

AORTIC VALVE DISEASE

AORTIC STENOSIS

Aortic stenosis (AS) remains the most common valvular lesion worldwide. Prevalence is estimated at between 9.2% and 16% in people over 75 years of age and severe AS is estimated at 3.4%. Approximately 75% of patients with severe AS are symptomatic, with 40% requiring surgical intervention leaving a significant proportion either unrecognized or untreated.

AETIOLOGY

Calcific aortic valve disease is the most common cause, ranging from aortic sclerosis, with thickening of the leaflets, to aortic stenosis where obstruction of the left ventricular outflow occurs. Calcification can occur with a structurally normal aortic valve, although progression is accelerated in the presence of congenital valvular abnormalities, e.g. bicuspid aortic valve (in approximately 2% of cases). Infective endocarditis and rheumatic heart disease account for the remaining cases.

PATHOPHYSIOLOGY

In systole, the aortic valve offers little resistance to outflow with near identical pressures in the aorta and LV. Thickening of the valvular leaflets and calcification lead to progressive narrowing of the open valve area. Once this has reached 50% of normal size (<2.0 cm^2), a pressure gradient develops resulting in pressure overload of the left ventricle. To compensate for the increased pressure required to maintain stroke volume, concentric left ventricular hypertrophy (LVH) occurs.

The hypertrophied left ventricle becomes impaired or 'stiff' with delayed isovolumetric relaxation, and consequent shortening of filling time – diastolic dysfunction. In turn, this increases filling pressures required for a given volume (Figure 2.1) meaning there is a greater reliance upon atrial contraction for adequate LV filling. Consequently, these patients are particularly susceptible to reductions in preload and atrial dysthymias, e.g. AF. These diastolic changes can also result in a 'fixed cardiac output state' where the cardiac output (CO) cannot be increased in response to systemic vasodilation.

Angina and an increased risk of myocardial infarction are present in a significant proportion of patients with AS despite up to a third having normal coronary arteries. This is largely a consequence of increased myocardial mass and hence oxygen demand, and the shifted balance between myocardial oxygen debt (systole) and myocardial oxygen repayment (diastole).

As the disease progresses, the pressure required to overcome the pressure gradient of the valve will no longer be balanced by the LVH resulting in increased wall stress, and eventually dilatation of the left ventricle cavity. These changes result in congestive cardiac failure.

PREOPERATIVE MANAGEMENT

Increased perioperative cardiac risk occurs in the presence of moderate or severe AS. This is increased further in the presence of symptoms. Hence, symptomatic patients for all elective noncardiac surgery and asymptomatic patients in the presence of severe or critical AS undergoing high-risk surgery, or the presence of concurrent CAD or coexisting moderate-severe MR, should be considered for valve replacement preoperatively. In patients unsuitable for surgical intervention, transcutaneous approaches may be considered (TAVR) or balloon valvuloplasty. If considered ineligible for the above, the patient should be counselled about the risks and benefits of proceeding.

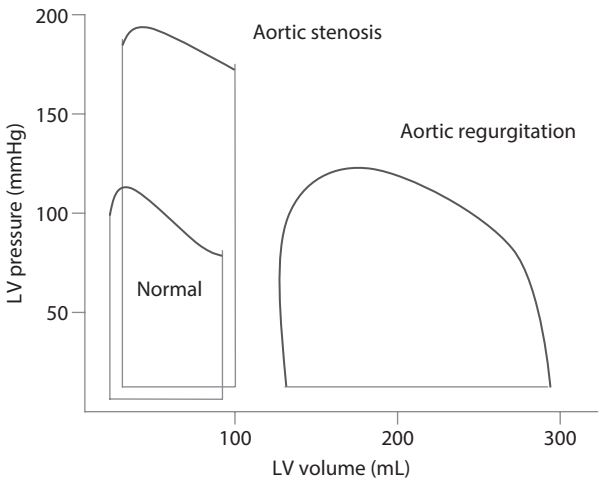

Figure 2.1 Aortic valve disease pressure volume loops.

HISTORY

- Look particulary for symptoms of AS or cardiac failure. If a new diagnosis of AS is made, or the presence of new symptoms or poor symptom control elicited, obtain a cardiologist's opinion.
- Review all medications.
- Obtain a history of comorbid diseases.
- Review recent cardiovascular investigations, in particular echo and exercise tolerance tests.
- Syncope, angina and dyspnoea (SAD) are the classical triad of symptoms. The presence of symptoms does not correlate well with severity of AS. The presenting symptom does correlate with mortality: CCF 2 years, syncope 3 years, angina 5 years.

EXAMINATION

Look for

- Slow rising low volume pulse with narrow pulse pressure.
- Sustained heaving apical impulse in the presence of LVH, or displaced apex in the presence of CCF.
- Ejection systolic murmur heard throughout the precordium, loudest in the aortic area and radiating to the neck. As the stenosis worsens, S2 will become singular and then reverse splitting occurs and S2 can be obliterated by the murmur. S4 may also be heard as atrial contraction against the stiff LV.
- Features of CCF.

INVESTIGATIONS

- *ECG* – Evidence of LVH/strain, LBBB or RBBB.
- *Echo* – Evaluation of severity of valvular lesion and LV function (Table 2.1) – note in the presence of LVF the pressure gradient can underestimate severity.
- *Cardiac atheterization* – Evaluation of LV function, valve gradient and concurrent CAD.
- *Exercise testing* – Consider to identify and evaluate coexisting CAD.

Table 2.1 Grading of severity of aortic stenosis by echocardiography

	Valve area (cm^2)	Mean valve gradient (mmHg)
Mild	>1.5	<20
Moderate	1–1.5	20–40
Severe	0.6–1	40–60
Critical	<0.6	>60

MONITORING

- *ECG* – At least two leads (II and V5) for early detection of ischaemia, although sensitivity may be reduced due to underlying LVH.
- *Invasive arterial pressure (preinduction)* – Allows rapid recognition of haemodynamic changes
- *TOE* – Allows accurate evaluation of ventricular filling and provides the most useful information where rapid volume shifts are expected, can also identify regional wall motion abnormalities (RWMA), an early sign of ischaemia. However, this requires an experienced operator.
- *Pulmonary artery catheter* – Not recommended as measurement of filling will be inaccurate and can induce dysrhythmias.
- *Noninvasive cardiac output monitoring* – Not validated, and values for stroke volume and cardiac output will be inaccurate although trends pre-/post-fluid boluses have been shown to be accurate with oeseophageal Doppler and pulse contour wave analysis.

PHYSIOLOGICAL TARGETS

- *Heart rate* – Aim for a low-normal rate. Avoid tachycardia – Reduces diastolic LV filling, coronary artery perfusion and LV ejection time in systole, resulting in reduced cardiac output and myocardial ischaemia.
- *Blood pressure* – Aim for normal blood pressure and high-normal SVR using direct alpha-agonists (metaraminol and phenylephrine). Pay meticulous attention to volume status to ensure adequate filling. Consider preloading with intravenous fluid boluses when fluid shifts are expected.

- *Dysrhythmias* – Treat promptly. A defibrillator should always be available in theatre if not connected to the patient for rapid DC cardioversion if required.

ANAESTHETIC TECHNIQUE

Rapid changes in SVR and ventricular filling are poorly tolerated due to a relatively fixed cardiac output. Falls in SVR result in reduced cardiac output, profound hypotension and reduced coronary blood flow with resulting myocardial ischaemia. Pain will cause increased catecholamine levels and resultant tachycardia; therefore, good analgesia is essential.

General anaesthesia +/- regional limb blocks is considered the best approach, although drugs should be titrated carefully. Avoid inappropriate use of premedication. A central venous catheter (CVC) may be considered for safe administration of vasoactive substances in symptomatic patients or patients undergoing high-risk surgery.

Central neuraxial blockade potentially causes significant reduction in SVR. This should generally be avoided in patients with severe AS, and caution exercised if undertaking this approach.

POSTOPERATIVE MANAGEMENT

Maintain a low threshold for transferring the patient to ICU even following a minor procedure, due to inexperience of ward staff in dealing with patients with severe AS. Continue direct arterial pressure monitoring postoperatively, with the same considerations regarding haemodynamic parameters as intraoperatively. Provide adequate postoperative analgesia and a plan for escalation if this is not achieved.

AORTIC REGURGITATION

The incidence of aortic regurgitation (AR) increases with age, with a peak at >80 years old. Estimates of prevalence range from 2%–30% for all severities, with less than 1% of the population having severe disease. The Framingham study demonstrated a significant gender difference with prevalences of 13% in men and 8.5% in women, although this may be accounted for by the higher incidence of bicuspid aortic valves and Marfan syndrome in males.

AETIOLOGY

Congenital or degenerative diseases of the valve leaflets or aortic root are the most common causes (Table 2.2). However, worldwide, rheumatic fever remains a significant cause of AR.

PATHOPHYSIOLOGY

Aortic regurgitation causes both volume and pressure loading of the left ventricle due to regurgitant flow from the aorta back into the LV during diastole. The resulting increased LV end-diastolic volume is compensated for by an increased stroke volume, hence increased systolic LV pressure and pulse pressure (Figure 2.1). The radius of the regurgitant orifice is the primary determinant of regurgitant flow, with up to 60% of the stroke volume returning to the LV in severe disease.

The LV remodels to accommodate this, with an increase in cavity dimension and mixed eccentric and concentric hypertrophy resulting in a more spherical shape. These changes and increased heart rate allow preservation of the ejection fraction (EF) until late stages of disease. Whilst the remodelling means patients can be asymptomatic for long periods, eventually the eccentric hypertrophy will fail to compensate for the increased volume and the concentric hypertrophy will fail to maintain normal wall stress. This results in raised filling pressures, reduced cardiac output and development of congestive cardiac failure.

Table 2.2 Causes of aortic regurgitation

Valvular	Aorta
Bicuspid aortic valve	Marfan's syndrome/ Ehlers–Danlos Syndrome
Infective endocarditis	Syphllis
SLE	Hypertension
Rheumatic fever	Ankylosing spondylitis/ RA/Behcet disease
Appetite suppressant drugs	Trauma

CLINICAL MANAGEMENT

PREOPERATIVELY

In asymptomatic patients with AR and preserved LV function, there is no increased risk of cardiovascular complications in noncardiac surgery. If patients have symptomatic CCF, NYHA III or IV, or EF <30%, they should be considered for valve replacement surgery. If not suitable for surgical replacement, medical optimisation, with ACE-I's, ARB's or mineralocorticoid antagonists and titrated cardio-selective beta-blocker therapy should be undertaken prior to elective surgery. For new diagnoses of heart failure, where possible, surgery should be delayed for 3 months to allow optimization.

HISTORY

Many patients will by asymptomatic; however, the most common symptom is dyspnoea. Patients may also develop other features of CCF and can develop angina. Pay particular attention to symptoms of AR or cardiac failure. Medications should be reviewed and a history of comorbid disease sought. Recent cardiovascular investigations, in particular echo and exercise testing, should be reviewed and noted as part of the preoperative assessment.

EXAMINATION

- Collapsing pulse and wide pulse pressure.
- Displaced apex inferolaterally.
- Early diastolic high-pitched murmur classically maximal at lower left sternal edge although if aortic root involvement is present, this may be heard in the aortic area. The volume of murmur is not indicative of severity of regurgitation. The murmur is best heard with the patient sitting-up, leaning forward in expiration.
- An Austin-Flint murmur may be heard in mid-diastole at the apex due to regurgitant flow directed at the anterior MV leaflet.
- **De Musset's sign** – Head nodding with each pulse.
- **Corrigan's sign** – Visible carotid pulsation.

INVESTIGATIONS

ECG – LVH/strain, LAD
CXR – Cardiomegaly +/– pulmonary congestion +/– dilated ascending aorta
Echo – Measurement of vena contracta, regurgitant fraction and evaluation of LV function.

INTRAOPERATIVELY

MONITORING

- *ECG* – At least two leads (II and V5) for early detection of ischaemia, although sensitivity may be reduced due to underlying LVH.
- *Invasive arterial pressure (from preinduction)* – To allow rapid recognition of haemodynamic changes, where CCF is present.
- *TOE* – May be considered to monitor filling, although an experienced operator is required. This has a strong indication for mixed valvular disease where early detection of RWMAs is of greater importance.
- *Noninvasive cardiac output monitoring* – Not validated and values for stroke volume and cardiac output will be inaccurate.

PHYSIOLOGICAL TARGETS

- *Heart rate* – Aim for a high normal heart rate. Avoid bradycardia as this increases diastolic time and hence regurgitant volume.
- *Blood pressure* – Aim for a low normal blood pressure with low-normal SVR ensuring adequate filling with IV fluid. Patients will be very sensitive to reduced preload.
- *Dysrythmias* – Treat promptly particularly if associated with hypotension. A defibrillator should always be available in theatre for rapid DC cardioversion of tachyarrhythmias if required. Persistent bradycardia may require treatment with anti-cholinergics or beta-agonists.

ANAESTHETIC TECHNIQUE

Unlike in AS, neuraxial techniques are well tolerated and the aim should be to maintain a low-normal afterload. Ensure good analgesia to prevent

catecholamine release and associated hypertension. Consider a CVC for safe administration of vasoactive agents in symptomatic patients or patients undergoing high-risk surgery.

POSTOPERATIVELY

Following high-risk procedures, or where large volume shifts have occurred, monitoring in ICU is indicated with invasive blood pressure monitoring, to detect early signs of CCF.

PROSTHETIC VALVES

Patients with prosthetic valve replacement will normally undertake regular review with the cardiology team, and should have recent assessment of valvular function. If this is not available locally, echocardiography should be performed preoperatively where possible.

ANTICOAGULATION

Tissue valves do not require lifelong anticoagulation except in special circumstances. Mechanical valves will require life-long anticoagulation normally with warfarin. The risk of thromboembolic events largely depends on the site and type of valve. Older aortic valve and mitral valve replacements have a much higher embolization rate of approximately 5% per annum. For these, bridging should occur for the full period INR remains <2, with either unfractionated heparin infusions or treatment dose low molecular weight heparins. For modern bileaflet mechanical aortic valves, technological advances have reduced the thromboembolic rate to less than 5% per annum, some studies report as low as 1%. For these valves, bridge with prophylactic dose low molecular weight heparin may be considered and an alternative to full therapy in low risk procedures.

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CROSS-REFERENCES

Heart failure, Chapter 2

Preoperative assessment of cardiovascular risk, Chapter 25

ATRIAL SEPTAL DEFECTS

Atrial septal defects (ASD) account for 10% of all congenital heart disease (CHD), and 20%–40% of CHD presenting in adult life. It is twice as common in women as men. Ostium secundum is the most common (75%), with primum (15%–20%) and sinus venosus (5%–10%).

AETIOLOGY

The formation of the atrial septum occurs in several stages. The septum primum is a soft tissue structure that grows towards the endocardial cushions to form the initial division into left and right atria. The space between the septum primum is the ostium primum, which narrows as the septum primum grows. Before this is fully occluded, the ostium secundum forms due to resorption of a portion of the septum primum, allowing continued movement of blood between right and left atria.

The septum secundum is a muscular structure that develops anterior to the septum primum. As it grows along a similar path to the septum

primum, it leaves an opening, the foramen ovale, which is continuous with the ostium secundum. The septum primum gradually shrinks leaving only a small flap of tissue, the valve of the foramen ovale, which closes after birth when the lungs become functional and pulmonary vascular pressure falls. Atrial septal defects are due to errors in this process.

Ostium primum ASD – Incomplete fusion of the septum primum with the endocardial cushion leads to a defect adjacent to the atrio-ventricular valves, which may or may not be affected (mitral > tricuspid).

Ostium secundum ASD – Caused by an unusually large ostium secundum, or failure of the septum secundum to correctly align with the septum primum. These defects can be further classified into:

Patent foramen ovale – Arises due to inadequate development of the septum secundum, or excessive or abnormal resorption of the septum primum, resulting in failure of normal closure of the foramen ovale soon after birth. Occurs in up to 30% of the population.

Sinus venosus ASD – Secondary to abnormal fusion of the sinus venosus and the atrial septum, usually near to the entry of the superior vena cava into the right atrium. Partial anomalous pulmonary venous drainage can be a feature.

Coronary sinus ASD – Defect resulting in an unroofed coronary sinus and persistent left superior vena cava that drains into the left atrium. Right-to-left shunt can result in desaturation. This is diagnosed by contrast injection into the left upper extremity – the coronary sinus will opacify before the right atrium.

PATHOPHYSIOLOGY

The degree of shunting across an ASD is related to the relative compliance of the two ventricles and the cross-sectional area of the defect. In the neonate, right- and left-sided cardiac pressures are approximately equal and little or no shunting occurs. As pulmonary vascular resistance falls, a left-to-right shunt develops. This is normally well tolerated; however, with large shunts the right heart will become volume-loaded and pulmonary flow will increase resulting in pulmonary hypertension and its sequelae. If left untreated, eventually the compliance of the right side of the heart will decrease and pressures will equalize between the right and left side of the heart. This allows bi-directional shunting and further increases in pulmonary artery pressure (PAP) will result in a right-to-left shunt (Eisenmenger's syndrome). Paradoxical emboli are now able to enter the systemic circulation from the right side of the heart, increasing the risk of systemic emboli and resultant CVA or MI. Surgery can be undertaken to close the defect and halt the progression to cyanotic disease; however, if this is delayed and the pulmonary hypertension (PH) is permanent, right ventricular (RV) failure will follow.

ASD CLOSURE

In childhood an ASD secundum may close spontaneously; however, once adulthood has been reached this is extremely unlikely. The decision to repair an ASD is based on clinical and echocardiographic information, including the size and location of the ASD, the magnitude and haemodynamic impact of the left-to-right shunt, the presence and degree of pulmonary arterial hypertension, previous paradoxical emboli, and the presence of orthodeoxia-platypnea. In general, closure is recommended in the following circumstances:

- Presence of right ventricular enlargement with or without symptoms
- Following the occurrence of a paradoxical embolic event
- Documented ortodeoxia-platypnoea
- In patients with PH if the pulmonary vascular resistance (PVR) <5 Wood units, or >5 Wood units with PAP <2/3 SVR

CLOSURE METHOD

There are two main options for closure of an ASD – percutaneous and surgical. For an ASD primum, surgical closure is performed due to the larger size of the defect and possible involvement of the mitral valve cleft. Coronary sinus ASDs and sinus venosus ASDs are usually closed surgically although percutaneous devices are used as well. For ASD secundums

<40 mm with a rim of tissue at least 5 mm around the defect, percutaneous device closure and surgery have comparable mortality data although reintervention is slightly higher for the device closure group.

PERCUTANEOUS DEVICE CLOSURE

Percutaneous closure is classically done under general anaesthesia via a femoral venous approach under intracardiac echo (ICE) or TOE guidance. If ICE is utilized, a supraglottic airway device may be used but if TOE is needed intubation is required. Following closure, dual antiplatelet therapy is continued for at least 6 months.

SURGICAL CLOSURE

For surgical closure, a midline sternotomy is routine but a minimally invasive approach with or without the aid of robotic surgery is in development. Complete thorascopic procedures have also been undertaken. Patients <25 years have better long-term outcomes. Some centres propose anticoagulation for up to 3 months following closure, to minimise the risk of thrombus attaching to the atrial patch and subsequent embolic complications. This is more common in patients who have had intraoperative arrhythmias and thus may benefit from postoperative anticoagulation.

CLINICAL MANAGEMENT

PREOPERATIVE MANAGEMENT

History

Pay particular attention to symptoms of cardiac failure, pulmonary hypertension, recurrent chest infections in children and cyanotic episodes with or without a relationship to posture. Review medications and any comorbidities. Review cardiovascular investigations, in particular echo and ECGs. Uncomplicated defects are likely to be relatively asymptomatic and the only indication of pathology may be the incidental finding of a murmur.

Examination

- Soft ejection systolic murmur loudest over the pulmonary area
- Pansystolic murmur loudest at the left sternal edge if tricuspid regurgitation (TR) has developed
- Fixed splitting of S2 due to increased pulmonary flow
- Pulse may be regular or irregularly irregular
- A right ventricular heave may be palpable at the left sternal edge
- JVP may be normal or raised, giant CV waves may be seen in the presence of TR, or cannon A-waves in the presence of right ventricular hypertrophy
- Features of syndromes associated with ASDs may be present, e.g. Down's syndrome

Investigations

- *ECG* – Look for AF; first-degree heart block; evidence of RVH/strain; incomplete RBBB; left axis deviation (LAD – seen with primum defects); right axis deviation (RAD – with secundum defects).
- *CXR* – Cardiomegaly, with atrial enlargement and pulmonary congestion.
- *ECHO* – Transthoracic echocardiography (TTE) can often reveal the defects – if not visible initially, bubble studies may be performed. ICE should be used to assess for the suitability of device closure.
- *Cardiac catherisation* – Can provide detailed information on location and function of the defect, pulmonary systemic shunting, PAP and ventricular function.

INTRAOPERATIVE MANAGEMENT

Monitoring for noncardiac procedures is the same as for a patient without an ASD. For closure procedures, use invasive arterial monitoring, ICE or TOE.

The recirculation of blood from intracardiac shunts may lead to a slower onset of intravenous induction and an increased dose may be required. Moderately soluble inhalational volatile agents (e.g. sevoflurane and desflurane) will have a more rapid increase in alveolar concentration due to a left-to-right shunt. Standard hypnotic agents are considered safe for induction and maintenance and inhalational agents provide a smaller reduction in

SVR than TIVA. Physiological targets should be similar to those for patients with established PH in order to prevent increases in PVR and potential shunt reversal.

POSTOPERATIVE MANAGEMENT

Postoperative care should be the same as for a patient without an ASD, depending on the procedure undertaken and comorbid disease. Pay particular attention to volume status and electrolyte management due to the increased risk of AF.

ASD AND EISENMENGER'S SYNDROME

Patients with right-to-left shunts will appear cyanosed and have finger clubbing. Pulmonary regurgitation, if present, causes a decrescendo diastolic murmur on auscultation. Chest radiography shows right ventricular hypertrophy, prominent pulmonary arteries and increased lung markings. Cardiac catheterization will confirm increased right ventricular and pulmonary artery pressures.

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Congenital heart disease in adult life, Chapter 2
Herat failure, Chapter 2
Patients with pacemakers and implantable defibrillators, Chapter 2
Pulmonary hypertension, Chapter 2

CORONARY ARTERY DISEASE

Although coronary artery disease (CAD) is no longer the greatest cause of mortality in the UK it remains a major cause of perioperative mortality and morbidity. Cardiac complications account for up to 42% of deaths within 30 days of surgery. It remains a major burden on healthcare systems in the developed world and has increasing prevalence in developing countries; it is estimated that the presence of CAD increases the perioperative risk of major complications by approximately 2.5-fold compared to the general population. This is particularly evident in open vascular surgery where the perioperative myocardial infarction rate is 5% compared to 1% nonvascular/noncardiac surgery. Timing of surgery after a previous myocardial infarction is important, with 30-day mortality 14.2% in the first 30 days reducing to 10.5% after >60 days.

AETIOLOGY

Atheromatous disease remains the most common cause. Plaques consisting of lipids with localized smooth muscle proliferation restrict blood flow within the coronary arteries. Ischaemia results when myocardial oxygen demand increases beyond supply or when there is rupture of plaque which can precipitate thrombosis and result in complete occlusion of an artery. Risk factors are illustrated in Table 2.3.

PATHOPHYSIOLOGY

Perioperative cardiac complications are caused by an imbalance between cardiac muscle oxygen supply and demand resulting in ischaemia. Increased

Table 2.3 Risk factors for CAD

Fixed	Modifiable
• Age: >45 men, >55 women • Gender: male 5x risk vs. female at age >50 • Race • Family history of early IHD	• Smoking • Hypertension • Diabetes mellitus • Obesity • Increased LDL:HDL ratio

myocardial oxygen demand may result from tachycardia (increases myocardial VO_2 and reduces diastolic filling time), increased contractility (e.g. pain causing a sympathohumoral response), and increased wall tension (inotropic therapy, hypertension). Alternatively supply may be reduced by tachycardia, vasospasm, hypotension, increased LVEDP (reduces coronary blood flow), hypoxia or anaemia. This is particularly important in the context of perioperative blood loss. Patients undergoing surgery are also at increased risk of an occlusive event due to the hypercoagulable and proinflammatory state.

PREOPERATIVE ASSESSMENT

AHA and ESC/ESA 2014 guidelines have suggested a stepwise approach to the assessment of preoperative risk evaluation and perioperative management of cardiac patients undergoing noncardiac surgery.

If the urgency of surgery prevents necessary cardiac testing or treatment, surgery should proceed with adequate perioperative surveillance for cardiac and medical treatment as appropriate. A plan should be instigated for immediate postoperative monitoring, further investigations and management; this would often include observation in an ICU.

If there is no requirement for immediate surgery, the patient should be screened for the presence of any unstable cardiac conditions:

1. Unstable angina pectoris
2. Acute heart failure
3. Significant cardiac arrhythmias (e.g. high-grade heart block, symptomatic monomorphic ventricular rhythms, polymorphic ventricular rhythms, new VT, SVT with ventricular response >100 at rest, new prolonged QT_c)
4. Symptomatic or severe valvular heart disease
   a. Severe or critical aortic stenosis (valve area <1 cm^2)
   b. Symptomatic mitral stenosis, or symptomatic in the presence of severe stenosis – valve area <1.5 cm^2 with pulmonary artery pressure >50 mmHg
   c. Symptomatic aortic or mitral regurgitation, or asymptomatic with LVEF <30%
5. Recent myocardial infarction (<30 days) or residual ischaemia

If any of these are present, the benefit of further evaluation and potential optimization should be weighed against any deleterious effects of delaying surgery by discussion involving anaesthetic, surgical and cardiology teams.

Where no unstable cardiac conditions are present, the procedural risk of the surgery should be considered in combination with the functional capacity of the patient. If the surgery is considered to have a 30-day MI and cardiac death risk <1%, or >1% with the functional capacity of the patient >4 metabolic equivalents (METs), risk factors should be identified and recommendations advised on lifestyle and medical therapy in line with current ESC/NICE guidance. A preoperative ECG should be considered. In patients with known IHD or previous myocardial ischaemia, low-dose titrated cardio selective beta-blockade should be considered or titrated preoperatively. Where ventricular systolic dysfunction has been identified, consideration should be given to an ACE-I preoperatively and in all patients undergoing vascular surgery statin therapy should be considered.

In situations where the functional capacity of the patient is ≤4 METS, the patient should be considered for noninvasive stress testing if one or more risk factors from Table 2.4 are present.

NON-INVASIVE TESTING

RESTING 12-LEAD ECG

A resting standard configuration ECG may reveal underlying rhythm disturbances or evidence of ischaemia. It is recommended that a resting ECG is

Table 2.4 Clinical risk factors according to revised cardiac risk index

Heart failure
Stroke or TIA
Diabetes mellitus requiring insulin therapy
Ischemic heart disease angina pectoris and/or previous myocardial infarction
Renal dysfunction (serum creatinine >170 umol/L or CrCl <60 mL/min/1.73 m^2
Peripheral arterial disease (ankle brachial ratio <0.9, or previous revascularization)

Source: Modified from Lee TH et al. (1999). *Circulation 100*:1043–1049.

performed on all patients with known CAD or risk factors undergoing intermediate or high-risk surgery. It may also be considered for patients undergoing low-risk surgery and patients over 65 with no risk factors undergoing intermediate or high-risk surgery. It is not recommended for patients undergoing low-risk surgery unless clinical suspicion of arrhythmias exists.

ASSESSMENT OF VENTRICULAR FUNCTION

Resting LV function may be assessed by SPECT, cardiac gated CT or MRI, radionucleotide ventriculography or echocardiography. TTE provides the most versatile and readily available imaging modality, which can also provide useful information about valvular disease. It is not recommended as a routine for all patients undergoing surgery, however, in the absence of signs of cardiac disease it may be considered for patients undergoing high-risk surgery. Patients with signs or symptoms of new valvular disease or cardiac failure should have TTE evaluation as should patients with known disease with symptomatic change in the presence of existing CAD.

STRESS TESTING

Bicycle or treadmill testing can detect inducible ischaemia and provides an estimate of functional capacity; however, the accuracy of detection of ST-segment changes during exercise varies significantly between studies and operators. Also preexisting ST-segment abnormalities or bundle branch blocks hinder reliable analysis. Results provide a graded response with onset of changes at low workloads associated with a significantly increased risk of perioperative mortality, cardiac events and long-term cardiac events. Inducible changes at high work-loads indicate a minimal increase in risk from a normal test.

In patients with limited exercise tolerance or reduced mobility, stress echocardiography, stress myocardial perfusion scanning, or stress cardiac MR can be carried out. This is achieved by undertaking imaging pre-/postpharmacological stressor (dipyridamole, dobutamine or adenosine). Stress echocardiography has high negative predictive value but low positive predictive value and failure to reach target heart rate is common. These allow inducible ischaemia to be demonstrated where areas of the myocardium are at risk, and fixed scarring where revascularization would be impossible.

Stress testing is recommend for patients undergoing high-risk surgery with poor functional capacity (<4 METS) and two or more risk factors (Table 2.4). It may also be considered in high or intermediate risk surgery in patients with one or more risk factors and poor functional capacity (<4 METS).

CORONARY ANGIOGRAPHY AND REVASCULARIZATION

Invasive coronary angiography is rarely indicated for patients undergoing noncardiac surgery and when undertaken inappropriately can result in unpredictable delay to planned surgery. Although CAD may be present in a significant proportion of patients undergoing surgery, the indications are similar to those in a nonsurgical setting. Urgent angiography is recommend where a patient has acute ST-elevation, new acute bundle branch block not requiring urgent surgery or where clinical benefit from angiography outweighs that of surgery. Urgent or early angiography should be undertaken in patients who have ACS or NSTEMI not requiring urgent surgery. In patients requiring urgent surgery, late revascularization should be considered postoperatively and the benefit of timing of angiography and surgery should

be discussed with the specialist teams on a case-by-case basis.

Elective preoperative angiography is recommended in patients with proven MI and unstable angina with maximal medical therapy. It can also be considered in patients with stable ischaemic cardiac disease undergoing nonurgent carotid endarterectomy. In the perioperative period two-thirds of myocardial events were not due to plaque fissure or intraluminal thrombus, but were type II MI where a low flow high demand situation occurred intraoperatively.

Surgical timing postrevascularization should be considered with current recommendations advising, where possible, a delay of 2 weeks following balloon angioplasty, a minimum delay of 4 weeks following bare metal stent insertion (but ideally 3 months) and a minimum of 6 months for new generation drug eluting stents (DES) or 12 months for older DES. If surgery is required to be performed within these windows it should be discussed with the specialist cardiology team on a case-by-case basis and discussion concerning antiplatelet therapy is essential with risks outlined to the patient.

PHARMACOLOGICAL INTERVENTIONS

Currently there is an extensive and ever-expanding array of pharmacological agents used in the treatment of CAD. As a result, in-depth discussion of each agent is not possible within the scope of this book. For complex cases, there should be discussion between the anaesthetic, surgical and cardiology teams in a timely manner preoperatively to allow optimization of therapy. Below some of the larger groups are discussed but this is not an exhaustive list of therapies.

BETA-BLOCKERS

Recent trials such as POISE and POBBLE have questioned the role of beta-blockade in the perioperative period. Both trials demonstrated increased mortality but a reduction in cardiac events. However, these results have been questioned by the maVS and DIPOM studies. As a result, the current ESC and AHA guidelines recommend continuation of beta-blocker therapy in patients on established therapy, and preoperative initiation may be considered in patients with known IHD or patients with >2 risk factors (Table 2.4) or ASA >3 undergoing high-risk surgery.

STATINS

Statins should be continued in the perioperative period if patients are established on this therapy. Current ESC guidelines also recommend initiation of statin therapy in patients with peripheral occlusive arterial disease prior to surgery and in patients undergoing vascular surgery who are statin naïve. This should be done 2 weeks prior to surgery to allow early detection of any complications such as statin-induced myopathy and rhabdomyolysis. For maximal benefit, statins should be continued for a minimum of 1 month post-surgery.

ACE INHIBITORS/ARBS

Continuation of ACE-I therapy perioperatively provides much discussion due to the risk of hypotension under anaesthesia. Observational studies have demonstrated a less frequent reduction in hypotension when ACE-I/ARB therapy is discontinued 24 hours prior to surgery when used for treatment of hypotension although this benefit remains debatable. Current guidance recommends that, in the presence of heart failure and LV dysfunction, AECE-I/ARBs continuation in the perioperative period under close monitoring should be undertaken. When used in the treatment of isolated hypertension without heat failure or LV dysfunction, consideration should be given to discontinuation of the therapy 24 hours preoperatively and until the patient's blood pressure and volume status are stable postoperatively.

INTRAOPERATIVE STRATEGIES

ST-SEGMENT MONITORING

The occurrence of perioperative ST-segment changes has been associated with cardiac morbidity and mortality also in patients undergoing noncardiac surgery. Intra- and postoperative ST-segment monitoring with computerized ST-segment analysis is considered

useful for patients with known coronary artery disease or those undergoing vascular surgery.

PULMONARY ARTERY CATHETER

Perioperative use of a pulmonary artery catheter remains a controversial issue. While significant information can be obtained from its use, no differences have been observed in survival or cardiovascular morbidity compared to standard care in patients who underwent major noncardiac surgery.

TRANSOESOPHAGEAL ECHOCARDIOGRAPHY

The use of transoesophageal echocardiography has gained wide acceptance in the setting of cardiac surgery. However, to date there is not sufficient evidence to support its routine use as a diagnostic monitor or to guide therapy during noncardiac surgery.

ANAESTHETIC MANAGEMENT

Neuraxial techniques can result in sympathetic blockade and cause a decrease in preload and afterload. Although initially some randomized controlled trials suggested that the use of neuraxial techniques might have beneficial effects on outcome, these data have not been unequivocally confirmed in more recent studies on larger patient populations.

A comparison of the effects on outcome of general anaesthesia with opioid analgesia to combined general-epidural anaesthesia and analgesia in intra-abdominal aortic, gastric, biliary and colonic surgery revealed no overall differences in death or major complications. It seems that to date, there is insufficient evidence to confirm (or deny) that postoperative analgesic techniques affect major postoperative morbidity and mortality.

In recent years, increasing evidence has indicated that volatile anaesthetic agents may have cardioprotective properties. In the setting of coronary artery surgery, the use of these drugs was shown to be associated with a better preservation of postoperative myocardial function and less evidence of postoperative myocardial damage. In noncardiac surgery, however, there is at the moment no such evidence.

Other measures to be taken in the perioperative period that may help to improve outcome include maintenance of normothermia and adequate perioperative pain management.

POSTOPERATIVE STRATEGIES

PAIN MANAGEMENT

Postoperative pain may increase sympathetic drive and therefore constitute a risk factor for the development of postoperative cardiac complications. However, the potential benefits of invasive analgesic techniques should be weighed against the potential dangers of their application. This is especially a concern in patients on antithrombotic or anticoagulant drugs.

Patient-controlled analgesia may be an alternative for postoperative pain relief. Nonsteroidal anti-inflammatory drugs and cyclo-oxygenase-2 inhibitors may promote heart and renal failure as well as thromboembolic events and should therefore be avoided in patients with myocardial ischaemia.

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Cardiopulmonary exercise testing, Chapter 25
Coronary artery bypass grafting, Chapter 16
Perioperative assessment of cardiovascular risk in noncardiac surgery, Chapter 25

CARDIAC CONDUCTION DEFECTS

Cardiac arrhythmias are common during anaesthesia. Underlying conduction abnormalities that are benign in the resting heart may be exposed due to the additional physiological stress and pharmacological interventions experienced perioperatively. The presence of structural or ischaemic heart disease increases further the risk of perioperative arrhythmias. Conduction abnormalities can be classified into:

- Atrioventricular blocks
- Bundle branch blocks
- Accessory pathways
- Long QTc syndromes

ATRIOVENTRICULAR BLOCK

Atrioventricular (AV) block can be defined as a delay or interruption in the transmission of electrical impulses from the atria to the ventricles caused by a problem at the level of the AV node (AVN) or His-Purkinje System. This may be a transient or permanent alteration to normal conduction and has many causes (Table 2.5).

Table 2.5 Classification of causes of AV block

Causes of AV block	Mechanisms
Cardiac	MI, fibrosis in the conduction systems (Lev and Lenegre syndromes), cardiomyopathies, CHD, e.g. ASD, PDA, Ebstein's, congenital heart block, cardiac surgery, valvular heart disease, myocarditis
Drugs	Beta blockers, calcium-channel blockers, digoxin
Systemic disease	Collagen vascular disorders, e.g. SLE, RA.
	Neuromuscular disorders, e.g. myotonic dystrophy
Other	Hypothermia, hyperkalaemia, hypoxia, hypo/hyperthyroidism

FIRST-DEGREE AV BLOCK

In a first-degree block, there is delayed transmission of impulses through the AVN resulting in prolongation of the PR interval to >200 ms. This heart block is normally asymptomatic and may be treated with atropine or glycopyrollate acutely if required. Chronically some patients benefit symptomatically from a dual chamber pacemaker.

SECOND-DEGREE AV BLOCK

Mobitz type I (Wenckebach) – There is delayed conduction through the AVN with progressive lengthening of the PR interval, until a beat is dropped (nonconducted P wave). This rhythm is usually asymptomatic and does not require treatment.

Mobitz type II – The block is usually at the level of the His bundle rather than the AVN. The ratio of P waves to conducted QRS complexes is often used to describe this block (2:1, 3:1, 4:1, variable). It can be associated with symptoms, usually pre/syncopal in nature. Mobitz type II may progress to complete heart block.

Management of reversible causes should be undertaken, e.g. electrolyte correction, treatment of myocardial ischaemia and cessation of drugs that increase nodal delay. If no reversible cause is found, and the rhythm persists, a permanent pacemaker is indicated and caution should be taken to avoid drugs which slow nodal conduction.

THIRD-DEGREE AV BLOCK (COMPLETE HEART BLOCK)

This occurs when no impulses are conducted from the atria to the ventricles and an escape pacemaker takes over. The ECG will show no relationship between P waves and QRS complexes. If QRS complexes are narrow and their rate is 45–55 bpm, this indicates an AVN block and a junctional or His-bundle escape rhythm. If QRS complexes are

broad and the rate is slower, at 30–40 bpm, it suggests an infra-nodal block with a distal ventricular escape rhythm. This block should always be treated. Removal of any aggravating medications, correction of electrolytes, and initial therapy with an isoprenaline infusion should commence while arrangements for a temporary or permanent pacemaker are made. Atropine can be used with caution in haemodynamically unstable patients; however, this will only increase rate if the block is at the level of the AVN. When the block is below the level of the AVN the unopposed vagolysis will increase the atrial rate, potentially slow the ventricular rate, and can lead to potentially dangerous ventricular arrhythmias.

BUNDLE BRANCH BLOCKS

Bundle branch blocks result from damage to the His-Purkinje system. This causes broadening of the QRS complex (>120 ms). The most common causes are age-related fibrotic changes, ischaemic heart disease, hypertension, cardiomyopathies, infiltration from systemic disease, cardiac surgery or trauma, and, for RBBB, pulmonary embolism or cor pulmonale. Diagnosis relies on careful examination of the ECG to ensure a supraventricular origin of the impulse.

Left bundle branch block (LBBB) – The left bundle is made up of two branches, the smaller anterior fascicle supplied by septal braches of the left anterior descending artery (LADA) and the posterior fascicle supplied from both the LADA and right coronary artery (RCA). A delayed depolarisation of the left ventricle gives rise to prominent notched R waves in all leads and an 'M' shape is often seen in V6.

If involvement is limited to the anterior fascicle (anterior fascicle hemiblock), then the ECG will show left axis deviation (LAD), and minimal prolongation of the QRS. If limited to the posterior fascicle (posterior fascicle hemiblock), right axis deviation (RAD) greater than 120 degrees will be evident with minimal prolongation of the QRS. Most commonly LBBB is seen with ischaemic heart disease, and development of a new LBBB should be considered as an acute ischaemic event equivalent to ST-elevation, and treated as such.

Right bundle branch block (RBBB) – Delayed depolarisation of the right ventricle produces an RSR pattern in V1 and a prominent S wave in leads I and V6. RBBB is relatively common in the adult population and has been reported in up to 2% of inpatients. However, if a new RBBB develops, it should be considered to be the result of acute ischaemia until proven otherwise and treated as equivalent to ST-elevation.

Bifascicular block – RBBB and L anterior or posterior hemiblock. May progress to trifascicular block or complete heart block. No specific management is required unless the block progresses at which point a pacemaker should be considered.

Trifascicular block – Bifascicular block and first degree AV block. This rhythm may progress to complete heart block. If the first degree block is due to AVN disease, then it is less likely to progress to complete heart block. Assessment by a cardiologist should be sought for patients with this rhythm, and if symptomatic a permanent pacemaker should be inserted.

ACCESSORY PATHWAYS

Conduction from the atria to the ventricles normally occurs via the AVN and His-Purkinje system. Patients with additional conduction tracts are said to have pre-excitation syndromes, because the tracts allow rapid bi-directional conduction of electrical impulses.

The most common of these is the Bundle of Kent. It results in the typical Wolff-Parkinson-White ECG findings of PR shortening (<120 ms), broad QRS (>120 ms) and delta-waves. Patients with accessory pathways are predisposed to AV re-entrant and unstable tachyarrhythmias, which may occur particularly in the context of AF. This can deteriorate to VT or VF.

Such patients should be managed by a cardiac electrophysiology specialist prior to elective procedures. Intra-operative AF can be treated with procainamide or amiodarone. If the patient is compromised by the rhythm, prompt synchronized DC-cardioversion is recommended. AV nodal reentrant rhythms can be treated with IV verapamil or lignocaine, in the absence of haemodynamic instability, or synchronized DC cardioversion if

this is present. For patients with pharmacologically managed arrhythmias, all anti-arrhythmic drugs should be continued in the perioperative period. Intraoperative vagolytic drugs should be avoided and adequate analgesia and depth of anaesthesia should be ensured, particularly during laryngoscopy and surgical stimulus to avoid catecholamine surges precipitating tachyarrhythmias.

LONG QT SYNDROME

The long QT syndrome (LQTS) is a disorder of myocardial repolarization characterized by a prolongation of the QT interval on the ECG. Patients with LQTS usually report palpitations, syncope and seizures and are at high risk of developing torsades-de-pointes and sudden cardiac death. The LQTS may be congenital or acquired. The congenital forms are caused either by autosomal dominant or less commonly by autosomal recessive genetic mutations, almost all of which encode for abnormal cardiac ion channels. The acquired form is usually caused by drug therapy, hypokalaemia or hypomagnesaemia.

ANAESTHETIC MANAGEMENT

Preoperative management by a specialist cardiac electrophysiology team is recommended due to the high risk of sudden cardiac death. Commonly implantable cardiac defibrillators are used to mitigate the risk of fatal arrhythmias. Beta-blockers have also been shown to reduce the QT interval and some patients may be managed with this as single therapy. Responders to beta-blockers as a single therapy appear to have lower risk of malignant arrhythmias intraoperatively. A baseline 12 lead ECG should be performed preoperatively, and correction of electrolytes undertaken, with particular attention paid to potassium and magnesium.

INTRAOPERATIVE MANAGEMENT

General and regional anaesthesia are considered safe although catecholamine surges should be avoided. Where local anaesthetic agents are used, adrenaline should be avoided. Premedication with a narcotic and benzodiazepine on the morning of surgery is recommended.

Antiarrhythmic drugs, a defibrillator with pacing facility and a temporary transvenous pacemaker should be available in the operating room with defibrillator pads attached to the patient. In addition to routine monitoring, a minimum of two ECG leads should be continuously monitored (one limb and one chest lead), invasive arterial monitoring is advisable and continual temperature measurement is recommended. The allows rapid identification of arrythmogenic factors such as temperature changes, heart rate changes, worsening in ST-segment elevation in Brugada syndrome or lengthening QT interval.

Intraoperative ventricular dysrhythmias in patients who respond to beta-blockers are usually responsive to further beta-blockade. Primidone, bretylium or verapamil may be used in those who do not respond. In both groups, premature ventricular contractions usually respond to lidocaine. Standard advanced cardiac life support protocols (with the possible exception of using epinephrine last) should be followed for ventricular tachycardia or fibrillation.

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CROSS-REFERENCE

Patients with pacemakers and implantable defibrillators, Chapter 2

CARDIOMYOPATHIES

A cardiomyopathy is defined as a change to the heart muscle that results in structural and functional abnormalities in the absence of coronary artery disease, valvular disease, hypertension or congenital heart disease sufficient to explain the observed myocardial abnormality.

The WHO classifies cardiomyopathies in terms of anatomy and physiology as follows; each of these can be further classified into idiopathic or acquired:

- Dilated cardiomyopathy (DCM)
- Hypertrophic cardiomyopathy (HCM)
- Restrictive cardiomyopathy (RCM)
- Arrhythmogenic right ventricular cardiomyopathy/dysplasia (ARVC/D)
- Unclassified cardiomyopathies

DILATED CARDIOMYOPATHY

Idiopathic DCM has a reported prevalence of 0.4 per 1000, and DCM overall has a likely prevalence of 1 per 2500. It is most commonly diagnosed in the third and fourth decades of life (Table 2.6).

Table 2.6 Aetiology

Causes of dilated cardiomyopathy	Examples
Hypertensive disease	Secondary to chronic hypertension and cardiac remodeling.
Valvular heart disease	Haemodynamically significant lesions (MR, AS, AR, MS) can lead to cardiac remodeling, hypertrophy, chamber dilatation and DCM.
Viral myocarditis and other infections	~50% of those receiving a diagnosis of acute viral myocarditis develop DCM. Common causes: coxsackievirus B, adenovarius, parvovirus. Other causes: HIV, Lyme disease, *trypanosoma cruzi*, toxoplasmosis and malaria.
Toxic cardiomyopathies	Alcohol (4% of cases), cocaine, amphetamines, anthracycline chemotherapy, e.g. doxorubicin, Herceptin, postpartum cardiomyopathy
Metabolic conditions	Malnutrition, vitamin and nutrient deficiencies, e.g. B vitamins, adrenocortical insufficiency, hyper/hypothyroidism, acromegaly and phaeochromocytoma.
Stress cardiomyopathy	Takosubo cardiomyopathy
Familial cardiomyopathy	Up to 25%, usually autosomal dominant, X-linked autosomal recessive and mitochondrial inheritance also reported.

HYPERTROPHIC CARDIOMYOPATHY

Prevalence estimated at 1 in 500 adults.

AETIOLOGY

Usually autosomal dominant inheritance of a number of mutations in genes encoding sarcomeric proteins such as beta-myosin heavy chain and troponin T, including *MYH7, MYBPC3, TNNT2, TNN13*.

Other causes of hypertrophic cardiomyopathy include chronic hypertension and aging.

RESTRICTIVE CARDIOMYOPATHY

Uncommon in the West (<5% of all cardiomyopathies), estimated prevalence of between 1 in 1000 to 1 in 5000. However, endomyocardial fibrosis is a significant cause of heart failure in parts of Africa (Table 2.7).

Table 2.7 Aetiology

Causes of restrictive cardiomyopathy	Examples
Idiopathic/ primary	Endomyocardial fibrosis or Loeffler eosinophilic endomyocardial disease
Infiltrative	Haemochromatosis, amyloid, sarcoid, cardiac carcinoid, glycogen storage disease affecting the heart, malignant infiltration
Treatment-related	Radiotherapy, anthracyclines

ARRHYTHMOGENIC RIGHT VENTRICULAR CARDIOMYOPATHY/ DYSPLASIA (ARVC/D)

The prevalence of ARVC/D is estimated to be between 0.2 and 1 cases per 1000 in the general population. Under-recognition remains a significant problem, so this may be an underestimate of the actual burden of disease. ARVC/D is an important cause of sudden cardiac death worldwide, representing the cause of 2%–5% of deaths in adults below the age of 40.

AETIOLOGY

The aetiology of ARVC/D remains unknown. In autopsies, the presence of inflammatory infiltrates in the myocardium has been demonstrated and ARVC/D has been documented post-myocarditis. Presently it is thought that it arises as the result of myocardial inflammation in the context of genetic susceptibility.

ANAESTHETIC CONSIDERATIONS FOR CARDIOMYOPATHIES

PREOPERATIVE MANAGEMENT

Pay particular attention to symptoms of cardiac failure, palpitations, syncope and associated conditions. Review medications and comorbidities. Elicit an accurate family history particularly with regards to sudden cardiac death or early onset CCF. Recent cardiovascular investigations in particular echocardiogram should be reviewed. In the early stages of the disease patients with cardiomyopathy may have no signs and symptoms. However, with the progression of the disease the classical clinical symptoms of cardiac failure will present.

INTRA- AND POSTOPERATIVE MANAGEMENT

Patients with cardiomyopathies generally behave like patients with heart failure of any cause, and should be managed as such in the perioperative period. Notable exceptions are patients with HOCM, restrictive cardiomyopathies and ARVC/D. Elective admission to a critical care environment should be considered postoperatively for patients with cardiomyopathies.

In patients with HOCM, the LVOT is prone to obstruction, resulting in loss of CO and fatal arrhythmias. This can be exacerbated by reduced LV filling. As such, in HOCM patients CO is said to be filling-dependent. In the perioperative period it is essential to avoid hypovolaemia and tachycardia, and hypotension should be primarily treated with fluid administration. In these patients, the use of invasive arterial BP monitoring and noninvasive cardiac output monitoring or TOE is strongly recommended to guide volume status.

In patients with restrictive cardiomyopathy, the stroke volume is relatively fixed and small, so rapid changes in SVR are not well tolerated. Nodal infiltration may also occur in diseases such as cardiac amyloid resulting in bradyarrhythmias. These should be fully investigated preoperatively. A high-normal heart rate and SVR should be targeted, and bradycardias treated promptly with vagolytic drugs.

In patients with ARVC/D, fatal arrhythmias are the most common cause of death. Such arrhythmias may be triggered during anaesthesia, and usual precautions with optimization of electrolytes preoperatively should occur. If an ICD is *in situ,* the normal precautions of this should apply. An external defibrillator should be connected to patient intraoperatively and only disconnected once the ICD has been reactivated.

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CHILDREN WITH CONGENITAL HEART DISEASE FOR NONCARDIAC SURGERY

The global incidence of congenital heart disease (CHD) is approximately 1 in 125 live births. These patients may present for emergency or elective surgery to nonspecialist centres with corrected, partially corrected or uncorrected disease. The key is to understand the underlying defect, the anatomy you are dealing with, and the functional physiology, then conduct a thorough preoperative evaluation. Despite advances in understanding and management, children with severe or major CHD still have greater than twice the 30-day mortality of matched controls.

There are more than 100 different types of CHD although the underlying pathophysiology allows a useful classification of CHD from a clinical perspective (Table 2.8).

PATHOPHYSIOLOGY

SHUNT LESIONS

Left-to-right shunts

Acyanotic congenital heart disease results from shunting of blood from the left side of the heart to the right side and into the pulmonary circulation. This is the most common pathophysiology in CHD patients. These conditions often manifest in the first two weeks of life due to the high PVR *in utero*. As this falls following delivery, the shunt may become evident with features of cardiac failure. As a consequence of shunting a proportion of the cardiac output to the right ventricle becomes volume overloaded with pulmonary hyperperfusion. The magnitude of the shunt for large defects is mainly dependent on the ratio of pulmonary vascular resistance (PVR) and systemic vascular resistance (SVR). As the defect size reduces, the shunt becomes largely independent of the

Table 2.8 Classification of CHD based on pathophysiology

Shunts	Left-to-right	Right-to-left
	ASD	Tetralogy of Fallot
	VSD	Truncus arteriosus
	AVSD	Complete Transposition
	PDA	Total anomalous pulmonary venous connection
	PFO	L-to-R shunt plus Eisenmenger's syndrome
Obstructive	LVOT	RVOT
Lesions	Aortic stenosis/atresia	Pulmonary stenosis/atresia
	Mitral stenosis	Tricuspid atresia
	Coarctation of aorta	Ebstein anomaly with intact septum
	Interrupted aorta	

resistances and predominantly determined by the size of the defect.

The increased pulmonary flow over time results in pulmonary hypertension as the pulmonary circulation adapts to the increased volumes and right-sided pressures. Once this has developed (a PAP >25 mmHg at rest or >30 mmHg during exercise), these children are eight times more likely to experience a major perioperative complication.

Pulmonary hypertensive crisis may occur preoperatively following the development of pulmonary hypertension where the PVR exceeds the SVR resulting in pressure overload of the RV and reduced pulmonary blood flow and compression of the LV +/- flow reversal across the shunt. This compromises cardiac output resulting in hypotension, hypoxia and a mixed respiratory and metabolic acidosis and ultimately biventricular failure. If this occurs, treatment to reduce the PVR should be administered: 100% O_2; inhaled nitric oxide or inhaled/intravenous prostacyclin; inotropic support of the RV; reduction in arterial $PaCO_2$.

Eisenmenger's Syndrome

Any uncorrected left-to-right shunting lesion may undergo flow reversal and become a right-to-left shunt. Once a patient has developed pulmonary hypertension in response to the increased flow and resultant damage to the pulmonary capillary beds, the RV undergoes compensatory hypertrophy to maintain forward flow. Right-sided heart pressures now exceed the left, and flow reversal occurs, leading to mixing of desaturated blood in the left heart and resultant systemic cyanosis.

Right-to-left shunts

Cyanotic heart disease results from shunting of desaturated blood from the right heart to the left and to the systemic circulation, bypassing the pulmonary circulation. These lesions carry an increased risk of paradoxical embolism, so particular care to avoid even small volume air embolism or accidental particle administration when administering IV medications and fluids should be exercised. The shunt volume may be reduced by manoeuvres that reduce the PVR and increase SVR, e.g. squatting, administration of 100% O_2, inhaled NO or prostacyclin. Conversely, this can be exacerbated by falls in systemic vascular resistance and increase in PVR seen in anaesthesia.

Polycythaemia may occur in patients with a right-to-left shunt. Arterial hypoxia is detected by the renal erythropoietin-producing oxygen-sensing cells in the juxtamedullary cortex of the kidneys, and erythropoietin (EPO) is produced in response. The increased erythropoiesis in response to high levels of EPO causes polycythaemia, high reticulocyte count, serum hyperviscosity and a resultant increased risk of thrombosis.

OBSTRUCTIVE LESIONS

Lesions leading to outflow tract obstruction of either the right or left heart, if uncorrected, leave patients with a relatively fixed cardiac output. The increased afterload for either ventricle results in pressure loading and compensatory hypertrophy. Obstructive lesions tend to have higher rates of arrhythmias which may be refractory to classic agents. Patients with severe lesions will rapidly develop symptoms of venous congestion on exertion or in illness. There is also an increased risk of myocardial ischaemia due to the increased ventricular mass and the relatively fixed cardiac output being unable to adapt to increases in demand.

PREOPERATIVE MANAGEMENT

In this patient group is it vital that the anaesthetist has a clear understanding of the functional physiological status of the patient, prior to proceeding with the case. This can broadly be split into two sections underlying circulation and presence of any of the four key risk factors (Table 2.9).

Table 2.9 Physiological risk factors

Physiological risk factors	
Cardiac failure	Arrhythmias
Pulmonary hypertension	Cyanosis

Source: Adapted from White MC and Peyton JM. (2012). *Contin Educ Anaesth Crit Care Pain* **12**(1): 17–22.

FUNCTIONAL CIRCULATION

SERIES "NORMAL" CIRCULATION

A series circulation is where there is a separate pulmonary and systemic circulation, working together. This is seen following complete repair procedures and with some forms of unrepaired lesions such as ASD or VSD. In the latter, blood mixing may occur down the pressure gradient resulting in shunts, as discussed.

PARALLEL CIRCULATION

A parallel circulation is where there is communication between the systemic and pulmonary circulations. Here the blood flow is determined by the resistances to flow, so blood will flow down the path of least resistance. It can be thought of as a balance between SVR and PVR determining the flow of blood to lungs and body. CCF occurs if pulmonary flow is too high with low cardiac output, and cyanosis occurs in the reverse. This situation is seen in infants with large VSDs, modified Blalock-Taussig (BT) shunts and hypoplastic left heart syndrome. Caution should be taken to avoid high concentrations of O_2, and avoid large drops in SVR.

SINGLE VENTRICLE CIRCULATION

In some cases the abnormality may not be amenable to full repair so palliative surgery can be performed. This may be a BT-shunt, Glenn shunt or a Fontan circulation. In these, the blood flow to the lungs is passive down the pressure gradient from the PA to LA. Varying degrees of cyanosis are observed with each of the shunts. These procedures may often be carried out in a stepwise manner in the child's perinatal and infant life. Here careful consideration should be given to ventilation strategies because the pulmonary blood flow is augmented by the negative intrathoracic pressure generated on inspiration in spontaneous breathing. Positive pressure ventilation will restrict pulmonary blood flow so inspiratory times should be kept short, PEEP optimized and high peak pressures avoided.

PREOPERATIVE ASSESSMENT

A thorough history should be taken including the original abnormality, any surgical correction undertaken, current medications reviewed, any issues or hospital admissions since birth for cardiac complications or coexisting disease. Particular attention should be paid to a history of recent upper or lower respiratory tract infections, pulmonary hypertension, CCF, arrhythmias, cyanosis, seizures, failure to thrive, developmental delay and reactive pulmonary disease. All cardiac investigations should be reviewed carefully, and baseline oxygen saturations, heart-rate and blood pressure should be fully documented.

EXAMINATION

A full examination of the child should be undertaken, particularly looking for features of cardiac failure, pulmonary hypertension, cyanosis and evidence of recent respiratory tract infection. These children will normally have cardiac murmurs and should be further investigated if atypical for the type of CHD or repair present. A full airway assessment should be carried out as CHD may be associated with syndromes involving the upper respiratory tract and cervical spine instability.

INVESTIGATIONS

These should be guided by history and examination with the child's underlying condition taken into consideration and any previous investigations performed. Where any doubts occur, these should be discussed with the child's cardiologist and/or specialist team.

ECG – All CHD patients should have a preoperative ECG. Whilst RBBB is common and unlikely to progress to third-degree heart block, the presence of ventricular ectopics (VE) warrants further investigation and consideration of transfer to a specialist centre. This is due to the high incidence of sudden cardiac death in CHD patients with VEs.

Echo – If there is no recent imaging, or there is a recent change in the child's condition, echocardiography should be performed.

Full blood count – Consider in the presence of cyanosis to evaluate for polycythaemia. If present, coagulation and viscosity studies should be considered.

U&Es – Many of the cardiac medications will affect electrolyte balance and renal function.

CXR – Should be performed if there is clinical evidence of cardiac failure or recent respiratory infection.

Cardiac MRI/catherisation – Should be undertaken on advice of specialist team or child's cardiologist.

RISK CLASSIFICATION

A significant part of the preassessment is to allow risk stratification and to enable the surgery to proceed in the safest environment. White and Peyton have proposed a very useful risk stratification system (see reference). For elective procedures, high-risk patients should be transferred to a specialist centre, intermediate-risk patients should be discussed with a specialist centre and considered for transfer, and low-risk patients should be operated on locally if the skill-set allows. In emergency situations, all high- and intermediate-risk patients should be discussed with the paediatric transfer service for feasibility of transfer to a specialist centre. If this is not possible, the case should be discussed with the specialist team and transferred at the earliest safe opportunity. Low-risk patients may be managed locally if the skill-set allows and any concerns discussed with the specialist team.

INTRAOPERATIVE MANAGEMENT

Full monitoring should be undertaken in line with paediatric AAGBI guidance. It should be noted that in CHD the $ETCO_2$ will correlate poorly with $PaCO_2$ and the gap will vary. Consideration of invasive monitoring should be undertaken for all intermediate- and high-risk patients.

PREMEDICATION

In patients with arrhythmogenic conditions, the prevention of a catecholamine surge may be beneficial. However, it may be harmful in patients with pulmonary hypertension if there is fixed cardiac output. Volume status should be considered and prolonged fasts avoided to prevent dehydration, as this has a twofold effect, both reducing preload and also increasing serum viscosity.

ANTIBIOTIC PROPHYLAXIS

In the current ESC guidelines on the use of prophylactic antibiotics for prevention of infective endocarditis, patients with cyanotic CHD, all CHD where a repair has been made in the previous 6 months with synthetic material or those with a residual shunt or valvular regurgitation are considered high risk. These patients should receive prophylactic antibiotics for high-risk procedures in keeping with local guidelines.

ANAESTHETIC TECHNIQUE

There is a wide variety of anaesthetic techniques described with no significant evidence that any one technique betters another. Propofol and ketamine have been well studied in children with propofol causing a reduction in SVR and CO. In some situations this may not be desirable such as in parallel circulations. Ketamine has relative small effects on SVR, PVR, PAP and MAP making it an ideal induction agent in children with pulmonary hypertension or parallel circulations. Whilst inhalational induction is considered safe, slower inductions with low concentrations of volatile agents should be considered. Intravenous access where possible should be obtained preinduction, or a second consultant anaesthetist or senior paediatrician should be present in the anaesthetic room such that IV access can be quickly secured postinduction.

Maintenance of anaesthesia with sevoflurane or isoflurane has been well studied and both have minimal deleterious effects. Ketamine and opiate infusions are also considered safe in this population although propofol infusions should be avoided due to the reduction in SVR and cardiac output. Regional anaesthesia is considered safe in CHD if it is routine practice for the procedure being undertaken.

POSTOPERATIVE MANAGEMENT

Provision of monitoring and timely intervention is key to successful postoperative recovery, with meticulous fluid balance and maintenance of baseline parameters with supplemental oxygen and drugs

as required. Elective admission to a paediatric critical care unit should be considered for all high- and intermediate-risk patients, or those with slow progression to baseline function postoperatively.

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CROSS-REFERENCES

Cardiac conduction defects, Chapter 2
Pulmonary hypertension, Chapter 2
Patients with heart failure, Chapter 2

CONGENITAL HEART DISEASE IN ADULT LIFE

With advances in surgical techniques and supportive management, there are increasing numbers of adults with grown-up congenital heart disease (GUCHD). Up to 90% now survive to adult life. In 2010 it was estimated that there were approximately 200,000 GUCHD patients in the UK and 1.2 million in Europe. There are significant physiological and anatomical consequences that impact anaesthetic assessment and practice in this specialized patient group.

Broadly speaking, GUCHD can be classified into simple, moderate or severe complexity rather than using an extensive anatomical classification (Table 2.10). These patients can also be classified into uncorrected, corrected and palliated.

Table 2.10 Congenital heart lesions by complexity

Simple	Moderate	Severe
Isolated aortic valve disease	Aorta to LV fistula	Conduits
Isolated mitral valve disease	Partial or total anomalous pulmonary drainage	Cyanotic congenital heart disease
PFO, small ASD or VSD	AV canal defects	Fontan procedure or TCPC
Previously repaired PDA	Coarctation of the aorta, or unrepaired PDA	Single ventricle circulation
Sinus venosus or secundum ASD (repaired)	Moderate to severe pulmonary stenosis or regurgitation	Transposition of the great vessels
VSD (repaired)	Tetralogy of Fallot	Mitral, tricuspid or pulmonary atresia
Mild Pulmonary stenosis	VSD with associated abnormality	Other rare congenial heart disease

Abbreviations: ASD = atrial septal defect; PDA = patent ductus arteriosus; PFO = patent foramen ovale; VSD = ventricular septal defect; TCPC = total cavo-pulmonary connection.

COMPLICATIONS OF GUCHD

Patients who have undergone surgical repair, palliation or unrepaired lesions are at risk of developing cardiac and noncardiac complications. The most common are electrophysiological disturbances, but this also includes cardiac failure, sudden cardiac death, infective endocarditis, bleeding and thromboembolic disease, renal impairment, airway stenosis, nerve palsies, altered blood flow and chronic hypoxaemia.

ELECTROPHYSIOLOGICAL CHANGES

These are both the most common and most significant consequences of GUCHD. Most commonly abnormalities of conduction are seen rather than SA nodal dysfunction. These can occur due to the altered anatomy and physiology, chronic hypoxia or as a direct consequence of surgical injury. Patients most prone to arrhythmias include those with history of ventriculotomy or lesions resulting in structural changes to the right side of the heart.

Anti-arrhythmic drugs are often tolerated poorly due to the negative ionotropic effects and should be used with caution. Bradycardia and deterioration to nonsinus rhythm is generally poorly tolerated, and should be avoided where possible. Cardioversion should only be carried out following anticoagulation and echocardiographic evaluation except in life-threatening circumstances due to the increased risk of cardiac thrombus.

CARDIAC FAILURE

Cardiac failure commonly develops as a consequence GUCHD due to the physiological and anatomical changes. Cardiac failure is generally treated in line with guidance on acquired disease and anaesthetic management should be similar to that for all patients with cardiac failure, with a low threshold for ionotropic support. For many GUCHD patients with cardiac failure, heart transplant is the only option although this surgery has increased complexity than in normal patients.

THROMBOEMBOLIC DISEASE AND BLEEDING TENDENCY

Abnormal anatomy, increases in turbulent flow and chronic hypoxia with compensatory erythrocytosis lead to an increased risk of intracardiac thrombus formation and also venous thrombosis. The use of pharmacological and mechanical thromboprophylaxis is essential perioperatively, with early mobilization post-surgery.

Conversely, large numbers of patients have established anticoagulation or antiplatelet therapy following shunt insertion, valvular surgery, device closure or following arrhythmias. Patients will often run a low-normal platelet count and may have abnormalities of function, particularly where the cardiac abnormality is part of a systemic syndrome.

INFECTIVE ENDOCARDITIS

Patients with GUCHD have an increased risk for the development of infective endocarditis. In recent years, guidelines have restricted the scope of use of prophylactic antibiotics. In general, prophylactic antibiotics are recommended for all untreated cyanotic heart disease and in patients with postoperative palliative shunts or other prosthesis. After surgical repair where no residual defects remain once endothelialisation has occurred normally at six months the risk is significantly reduced. Patients with a prosthetic valve and GUCHD should be treated as other patients with prosthetic valves. Current European Society of Cardiology guidelines recommend that inantibiotic prophylaxis is restricted to the above groups unless high-risk procedures are being undertaken. In patients with GUCHD, strict asepsis should be observed when inserting invasive lines or undertaking any invasive procedures.

HYPOXAEMIA

In patients with congenital heart disease, chronic as opposed to acute hypoxaemia is a common symptom and is usually associated with reduced pulmonary blood flow and/or right-to-left shunting. Hypoxaemia is a concomitant feature of arrhythmias, concurrent cardiac failure and pulmonary disease.

AIRWAY ABNORMALITIES

Tracheal or bronchial stenosis is commonly seen in patients with GUCHD. This may result from

prolonged periods of intubation and ventilation or external compression from enlarged or malpositioned vessels. Prosthetic conduits may also result in scarring and stricture formation.

PREOPERATIVE MANAGEMENT

Congenital heart disease confers an increased perioperative risk from cardiac and noncardiac causes. It is vital that all but the simplest of conditions and situations be discussed with a specialist centre and transfer to a cardiac centre be considered.

A thorough history should be elicited, with particular attention to symptoms of heart failure, palpitations, arrhythmias, syncope, previous infective endocarditis and functional capacity. Details of the original abnormality and any corrective/palliative surgery undertaken and any recent reviews from their specialist team. Medications should be reviewed, and a history of comorbid disease sought. All recent cardiovascular investigations, in particular echo and exercise testing, should be reviewed. If present, arrhythmias may be symptomatic with palpations, dizziness or syncopal episodes. Features of heart failure may also be described.

On examination look for:

- Scars suggestive of previous thoracotomy or permanent pace maker.
- Pulse – Normal; low volume in low cardiac output states; irregularly irregular in AF.
- Central or peripheral cyanosis.
- JVP – May be raised, and/or distended neck veins may be seen.
- Apex bet – Displaced in the context of volume-overload.
- Cardiac murmurs.
- S_3 with gallop rhythm may be audible on auscultation.
- Basal crepitations may indicate pulmonary oedema.
- Peripheral oedema.

INVESTIGATIONS

- *ECG* – Arrhythmias, conduction defects, evidence of current or previous ischaemia.
- *CXR* – Cardiomegaly, pulmonary congestion (bat-wing shadowing), Kerley B lines (interstitial oedema), pleural effusions. Shunts, permanent pacemakers and artificial valves may be visible.
- *Echo* – Recommended for all patients undergoing intermediate or high-risk noncardiac surgery – consider presence, quantification, timing and degree of ventricular dysfunction, structural abnormalities and valvular lesions. Regional wall motion abnormalities may be seen with ischaemic heart disease, or myocardial thickening in myocarditis.
- *FBC* – Low platelet count may be present, or reactive polycythaemia.
- *U&Es* – most cardiac medications affect electrolyte balance and renal function; look for cardio-renal syndrome, AKI or CRF. Evaluate and optimise preoperatively.

INTRAOPERATIVE MANAGEMENT

MONITORING

- *ECG* – At least two leads (II and V5) for early detection of ischaemia, although sensitivity may be reduced with abnormal baseline ECG.
- Invasive arterial blood pressure (start before induction).
- *TOE* – To monitor filling, although an experienced operator is required. Strongly indicated for mixed valvular disease, where early detection of regional wall motion abnormalities is of greater importance.
- *Noninvasive cardiac output monitoring* – Useful for monitoring filling status. Although absolute values are unlikely to be accurate, response to fluid boluses do show incremental changes with pulse contour wave analysis, and Doppler flow.

PHYSIOLOGICAL TARGETS

- *Heart rate* – Aim for a normal resting heart rate. Patients are particularly sensitive to tachy- and brady-arrhythmias and rhythm changes.
- *Blood pressure* – Aim for a normal blood pressure. A low normal SVR should be

maintained by meticulous attention to volume status to ensure adequate filling but avoid overload. Consider preloading with intravenous fluid boluses when fluid shifts are expected.
- *Dysrhythmias* – Treat in line with current ALS recommendations. A defibrillator should always be available in theatre for rapid DC cardioversion if required.
- *Other* – Avoid hypoxia, hypothermia, hypercarbia and acidosis, which may increase reduced myocardial function and increase PVR.

ANAESTHETIC TECHNIQUE

Anaesthetic technique should take account of the specific anatomical and physiological impact of the malformation. The uptake of inhalation agents can be affected by intra-cardiac shunting. An R-L shunt will reduce the pulmonary blood flow and hence the partial pressure in blood going to the brain resulting in a slower uptake; however, intravenous agents will have a more rapid onset. Peripheral nerve blocks are recommended where possible to reduce the haemodynamic impact of general anaesthesia. Central neuraxial blockade should be used with extreme caution due to the sensitivity of these patients to preload and poor tolerance of bradycardia.

POSTOPERATIVELY

All patients should have supplemental oxygen and good analgesia is essential to minimise catecholamine release. Following medium- or high-risk procedures, or where large volume shifts have occurred, monitoring in ICU is indicated with invasive blood pressure monitoring, to detect early signs of decompensation. All patients with cyanotic heart disease should be managed in ICU or a specialist cardiac centre.

PREGNANCY AND GUCHD

A significant proportion of maternal deaths during pregnancy are accounted for by GUCHD patients. However, the majority of patients, particularly those with simple lesions, can undergo pregnancy without difficulty to mother or fetus. Counselling should be advised prior to pregnancy and this should be lesion-specific by a specialist team. For example, patients with R-L shunts are more susceptible to changes in SVR and flow across the shunt will increase potential resulting in cyanosis.

During labour there is an increased risk of developing infective endocarditis. Caesarean section should be reserved for obstetric reasons as far as possible, with planning by the obstetric, cardiac and anaesthetic teams crucial throughout the pregnancy. Induction of labour should only be considered in a specialist centre if there is evidence of cardiac decompensation.

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CROSS-REFERENCES

Atrial septal defect, Chapter 2
Cardiac conduction defects, Chapter 2
Medical problems and obstetric anaesthesia, Chapter 12
Heart failure, Chapter 2
Patients with pacemakers and implantable defibrillators, Chapter 2
Pulmonary hypertension, Chapter 2
Tetralogy of Fallot, Chapter 2

HEART FAILURE

Heart failure (HF) can be defined as any structural or functional cardiovascular abnormality leading to systemic perfusion unable to meet the body's

metabolic demands without excessive increase in LV filling pressures.

Two major classification systems are in clinical use. The New York Heart Association (NYHA) scale classifies severity in terms of functional limitation and degree of activity required to elicit symptoms (Box 2.1). The American College of Cardiology/American Heart Association (ACC/AHA) guidelines (Box 2.2) categorizes the stages of heart failure into distinct groups. This emphasizes the progressive nature of heart failure and allows for intervention based on stage.

BOX 2.1: NYHA functional classification of heart failure severity

- Class I – Patients with heart disease that does not limit physical activity and an absence of symptoms (fatigue, palpitation, dyspnoea) on ordinary physical activity.
- Class II – Patients with heart disease resulting in mild limitation of physical activities or symptoms of HF developed with ordinary physical activity but absent at rest.
- Class III – Patients with heart disease resulting in significant limitation of physical activity or symptoms of HF on minimal activity but absent at rest.
- Class IV – Patients with heart disease resulting in the inability to carry on any physical activity without discomfort. If any physical activity is undertaken discomfort increases.

BOX 2.2: AHA/ACC stages of heart failure development

- Stage A – At risk of HF but without structural heart disease or symptoms of HF (fatigue, palpitation, dyspnea).
- Stage B – Structural heart disease but without signs or symptoms of HF.
- Stage C – Structural heart disease with prior or current signs or symptoms of HF.
- Stage D – Refractory HF requiring specialized interventions.

EPIDEMIOLOGY

Prevalence is estimated at 2–20 per 1000 population and increases steeply with advancing age. Median age at diagnosis in the UK is 76 years. More than 37.7 million people are thought to suffer from heart failure worldwide. The 900,000 patients in the UK account for 5% of all emergency admissions and 2% of inpatient bed-days.

AETIOLOGY

Heart failure most commonly occurs in developed countries as a sequel of chronic hypertension or valvular heart disease (Table 2.11).

PATHOPHYSIOLOGY

Changes seen in HF result from physiological changes in response to cardiac injury. Raised EDV is compensated for by LVH and increased wall stress, with initial increase in SV and contractility. This is unsustainable with continuing volume overload and leads to fibrotic changes in the myocardium and LV

Table 2.11 Classification of cardiac failure

	Causes
Left-sided heart failure	Ischaemic heart disease or hypertensive heart disease (commonest cause) Valvular heart disease Cardiomyopathy Myocarditis
Right-sided heart failure	Lung disease with pulmonary hypertension, e.g. end-stage COPD, ILD, and resultant cor-pulmonale Right-sided cardiomyopathy, e.g. arrhythmogenic RV dysplasia (rare)
Congestive or biventricular heart failure	RV failure due to pulmonary hypertension and fluid overload as a result of LV dysfunction
High output failure	Anaemia Pregnancy Hyperthyroidism Paget's disease of bone Arteriovenous malformations

dilatation with impaired active relaxation and hence ventricular filling.

Various pathways are involved in the development of HF and are targeted therapeutically. The Renin-Angiotensin-Aldosterone System (RAAS) plays a key role in pathogenesis. It is up-regulated due to renal hypoperfusion and sympathetic stimulation, resulting in increased circulating angiotensin II, which causes vasoconstriction, salt and water retention and ventricular remodeling.

CLINICAL MANAGEMENT

Patients with clinical evidence of HF, active symptoms or features present on examination are at significantly increased risk of perioperative complications. This is reflected by its inclusion in the majority of risk indices. In the developed world, the prevalence of heart failure continues to increase steadily as the population ages resulting in more patients presenting for surgery with active disease. Patients with severe heart failure LVEF <30% and/or NYHA class IV have high rates of perioperative cardiovascular complications and increased mortality. Where urgency of surgery allows, these patients should be optimized medically preoperatively and counseled about the risks. There is no clear evidence demonstrating difference in mortality for patients with isolated diastolic HF (preserved LVEF) versus systolic HF highlighting heart failure and functional limitation as key determinants perioperatively.

Acute decompensation provides the greatest risk to the patient and should be evaluated and treated by a cardiologist. Elective surgery should be deferred until this has occurred and ideally a stable baseline achieved for at least two weeks for patients with known chronic HF or at least three months following a new diagnosis of severe HF although urgency of surgery may preclude this. For emergency surgery, haemodynamic optimization should occur with echo evaluation as soon as feasible and pre-/postoperative natriuretic peptide levels. Postoperative care will likely be in an ICU with involvement of the cardiology team.

PREOPERATIVE MANAGEMENT

Pay particular attention to symptoms of HF, history of associated conditions and functional capacity. Review medications and comorbidities. Recent cardiovascular investigations, in particular echo and exercise tests should be reviewed and noted. If a new diagnosis of HF is made, or presence of new symptoms or poor symptom control is elicited, a cardiology opinion should be sought preoperatively where possible. Symptoms include:

- Fatigue
- Exertional dyspnoea
- Paroxysmal nocturnal dyspnoea
- Orthopnoea

If present, arrythmias may be symptomatic with palpations, dizziness or syncopal episodes. If HF is secondary or in the presence of coexisting CAD, the patient may report angina episodes. Signs of gout may be present and signs of chronic renal failure in the presence of cardiorenal syndrome.

EXAMINATION

- Pulse may be regular, of low volume if a low cardiac output state is present or irregularly irregular in the presence of AF.
- JVP may be raised and/or distended neck veins may be seen.
- The apex may be normal or displaced in the context of volume-overload.
- S_3 with gallop rhythm may be audible on ausculataion of the precordium.
- Bibasal crepitations are heard on auscultation of the posterior aspect of the chest wall with pulmonary oedema
- Peripheral oedema will result in pitting ankle and sacral swelling and weight gain.
- Hepatomegaly (may be pulsatile) can result from right-sided failure with or without TR portal flow reversal on Doppler studies.

INVESTIGATIONS

- *ECG* – Look for arrhythmias, conduction defects, or evidence of current or previous ischaemia.
- *CXR* – Cardiomegaly and pulmonary congestion (bat-wing shadowing) can be seen; Kerley B lines may be present at the lung margins from interstitial oedema; bilateral pleural effusions may also be seen.