Anaesthesia for non-cardiac surgery in patients with congenital heart disease

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Congenital cardiac abnormalities occur in 1% of the live-born population, with a wide spectrum of disability ranging from trivial to lethal. With rapid developments in the scope of surgery for congenital heart defects, there are a growing number of medium-term survivors. These may require anaesthesia for elective and emergency surgery, trauma and obstetric intervention. The aim of this review is to aid the anaesthetist, working in a non-specialist environment, to differentiate between straightforward and complex cases and to have a management strategy for each category.

We also wish to cover briefly the management of five important conditions that might be encountered by non-specialist anaesthetists: (i) the cyanosed neonate; (ii) the child with Fallot’s tetralogy; (iii) the obstetric patient with congenital heart disease; (iv) the patient with a single ventricle; and (v) the patient with a transplanted heart.

The spectrum of disease

Congenital heart defects may be divided into those where, with appropriate surgery, physiological correction is achievable and those where it is not. An example of the former is an atrial septal defect where surgery closes the shunt and is corrective. An example of non-correctable anatomy is the hypoplastic left heart syndrome where inadequate left ventricular tissue, combined with an hypoplastic aorta, is not amenable to correction. Surgery, other than transplantation, can only be palliative.

After consideration of the cardiac anatomy, the pathophysiology of each lesion should be considered. Symptoms of cyanosis, chronic lung disease, heart failure and dysrhythmia should be elicited. Each of these symptoms and their investigation and management will be dealt with below. However, a crucial point in the history of each symptom is its time course. New dyspnoea with rapid progression being of more significance than long-standing, well medically managed exercise dyspnoea.

Once full details of the anatomy, surgical history and current pathophysiology are obtained, patients can be divided into the following categories:

1. Congenital heart disease, yet to be surgically treated.
2. Surgically corrected, symptom free with no new developments.
3. Surgically corrected with symptomatic heart disease.
4. Surgically palliated, symptoms stable with no new developments.
5. Surgically palliated with severe symptoms or new developments.

The questions facing anaesthetists are:

1. Should the patient be referred to a specialist cardiologist before surgery?
2. Should surgery be performed in a centre specializing in congenital cardiology?
3. How should anaesthesia be conducted safely in the presence of congenital heart disease?

The answer to the first two questions will depend on the degree of urgency as well as details of the individual patient. If the situation permits, patients with palliated heart disease will benefit from input from specialists in congenital heart disease as they often have grossly deranged circulations. Those with new, or rapidly progressive, symptoms also warrant further investigation before all but the most urgent surgery.

The anatomy of the common lesions along with the surgical treatment most frequently
Normal heart. Surgical treatment of congenital heart disease aims to correct anatomy to allow blood flow to pulmonary and systemic circulation in sequence. This may be impossible due to congenital absence of structures. In this case, palliation is attempted and blood flow is abnormal through surgically created shunts and baffles. Cyanosis, arrhythmia and heart failure are common. Even with corrective surgery, leaks across repaired valves and potential for heart failure, arrhythmia and endocarditis exist. Each congenital anomaly is a disease spectrum and may vary from trivial to severe.

Ventriculoseptal defect. L-R shunt leads to heart failure and pulmonary hypertension. Those not closing spontaneously are closed with a surgical patch. After correction, potential problems with endocarditis and arrhythmia.

Atrial septal defect. L-R shunt leads to pulmonary hypertension in later life. Corrected with catheter insertion of umbrella or surgical patch. Few postoperative problems, but risk of arrhythmia or endocarditis.

Patent ductus arteriosus. Commoner in premature. If medical closure fails, transcatheter coil obstruction or surgical ligation performed. Late problems rare.

Atrioventricular septal defect. Most frequent lesion found in Down's syndrome patients. Results in severe heart failure and early onset of pulmonary hypertension. Surgical correction performed but mitral valve regurgitation may lead to long-term problems. Endocarditis and arrhythmia risk.

Hypoplastic left heart syndrome. Before surgery, pulmonary circulation depends on patent duct. Three stage palliative surgery leads to single ventricle and pulmonary flow through cavo-pulmonary connections. Palliation may result in problems with heart failure and arrhythmias.

Transposition of great arteries. Balloon septostomy performed in first few hours of life. Arterial switch performed but older patients have atrial switch. Few long-term problems with arterial switch but atrial baffles risk endocarditis, arrhythmia and failure of systemic right ventricle.

Pulmonary atresia. Pulmonary flow maintained with prostaoglandin until surgery. Treatment and outcome depend on presence of VSD and right ventricular tissue. Many patients suffer repeat palliative operations and cyanosis.

Fallot's tetralogy. Variable cyanosis with potential for hypercyanotic spells. Corrected by VSD closure and relief of RV outflow tract obstruction. Damage to pulmonary valve may lead to RV failure. Arrhythmia and endocarditis risk.

Fig. 1 Anatomy of congenital heart disease, surgical treatment and its implications. From The Multimedia Encyclopedia of Congenital Heart Disease, courtesy of Scientific Software Solutions, Inc. (Charlottesville, VA, USA <www.pedheart.com>).
employed are shown in Figure 1. A thorough understanding of the physiological changes due to the abnormal anatomy of each individual lesion is more important in devising a management plan than the actual drugs or techniques used.

Cardiovascular impairment
Cardiovascular impairment is generally manifest in four ways: cyanosis, pulmonary disease, cardiac failure or arrhythmias – either alone or in combination.

Cyanosis
By its presence alone, cyanosis indicates some sort of persistent cardiac abnormality. The degree of cyanosis at rest should be determined, along with any history of hyper-cyanotic episodes (and their precipitating factors). An attempt should be made to distinguish between cardiac and pulmonary causes of hypoxaemia as active pulmonary infection would be an indication for postponing elective surgery. Chronic hypoxaemia results in polycythaemia and abnormal haemostasis. Whatever the underlying defect, the primary objective is to maintain adequate tissue oxygenation. In general:
• ensure hydration
• maintain systemic vascular resistance to reduce right to left shunt
• minimise pulmonary vascular resistance, utilising a high inspired oxygen concentration
• avoid sudden increases in oxygen demand, e.g. crying, inadequate anaesthesia
• meticulous removal of air from all intravenous lines is vital in the presence of any shunt, regardless of its direction

When hypoxaemia is due to mixing of systemic and pulmonary blood, arterial saturation depends on the ratio of pulmonary to systemic blood flow (Qp:Qs). Fully saturated blood should not be expected. Management should maintain ventricular function and control the Qp:Qs ratio by manipulating systemic and pulmonary vascular resistance.

Pulmonary disease
Congenital heart defects are frequently associated with other anatomical abnormalities of the airways and pulmonary vasculature. The trachea or bronchi may be compressed by vascular rings, an enlarged heart, pulmonary artery, aorta or artificial shunts. Phrenic or recurrent laryngeal nerve compression may also occur, or they may have been damaged by previous surgery.

Excess pulmonary blood flow, secondary to a left-to-right shunt, leads to increased pulmonary vascular resistance and pulmonary hypertension, which is irreversible. Lung compliance is reduced and airway resistance increases, leading to increased work of breathing. Early surgery is crucial to avoid this irreversible phenomenon. Chronic exposure to excess pulmonary blood flow ultimately results in a pulmonary vascular resistance that equals or exceeds systemic vascular resistance. If there is a communication between the two circulations at this point, Eisenmenger’s syndrome develops with a reverse of the shunt direction and a new onset of cyanosis.

If there is reduced pulmonary blood flow, physiological dead space increases. Also, mechanical ventilation causes a further increase in dead space. Therefore, end-tidal carbon dioxide monitoring consistently underestimates PaCO₂.

Cardiac failure
Cardiac failure occurs when the heart cannot pump enough blood to meet the metabolic demands of the body. Compensatory mechanisms for a failing heart include ventricular hypertrophy, increase in circulating catecholamines, and sodium and water retention. Most patients presenting for non-cardiac surgery will not be in overt failure. However, if this is the case, elective surgery should be postponed until medical or interventional treatment optimises function. Cardiac reserve should be estimated – exercise tolerance is probably the most informative. In cases where functional limitation exists but surgery cannot be deferred, the details of the anaesthetic technique chosen are less important than the care with which the patient is managed. Adequate premedication avoids anxiety and tachycardia. Care in induction of anaesthesia is necessary to avoid myocardial depression, vasodilation and hypotension. Mechanical ventilation with tracheal intubation may be beneficial for many patients. However, where there is passive blood flow to the lungs (e.g. palliative cavo-pulmonary shunts), positive intrathoracic pressure may reduce pulmonary blood-flow and an anaesthetic technique avoiding positive pressure ventilation may be more appropriate. In any patient with cardiac failure, there should be a low threshold for invasive monitoring and adequate consideration given to close supervision in the postoperative period.

Arrhythmias
Arrhythmias are a significant long-term problem for patients with congenital heart defects due to disorders of the conduction system – both congenital and acquired. Common arrhythmias
include complete heart block, supraventricular tachycardia and ventricular tachycardia.

**Complete heart block**

If permanent pacing is not indicated, have a temporary pacing device readily available. Transcutaneous pacing has been effective in children as small as 6 kg. Avoid drugs which decrease sympathetic tone or increase vagal tone. Avoid hypovolaemia as cardiac output may depend on stroke volume alone.

**Supraventricular tachycardia**

Premedication is useful to decrease sympathetic activity. Vagolytic drugs should be avoided. If circulation is compromised, synchronised DC shock is recommended. Other options include vagal manoeuvres, overdrive pacing, adenosine, digoxin, amiodarone, β-blockers or verapamil.

**Ventricular tachycardia**

The presence of ventricular ectopics on the ECG is ominous; 30% of these patients will eventually die suddenly. Therefore, if possible, refer for medical treatment or further surgical intervention. Otherwise, avoid excess sympathetic stimulation, hypercarbia, acidosis and hypoxaemia.

**Antibiotics and anticoagulation**

Endocarditis is a risk following most cardiac operations especially if prosthetic material is present. Procedures resulting in a normal circulation, using no prosthetic material, are now regarded as low-risk for endocarditis. Up-to-date antibiotic prophylaxis regimens are regularly published in the British National Formulary. Patients who are anticoagulated can present a dilemma. However, in general, the risk of allowing coagulation to normalise during the peri-operative period is preferable to the difficulties with haemostasis if anticoagulation is continued unchanged. Warfarin should be stopped 1–3 days pre-operatively (use fresh frozen plasma in an emergency situation) and heparin commenced. Heparin should be discontinued 4–6 h pre-operatively and restarted within 12 h postoperatively. Warfarin may then be restarted 1–7 days postoperatively.

**The cyanosed neonate**

At birth, the sudden increase in PaO₂, leads to functional closure of the ductus arteriosus within 6 h – anatomical closure occurring around 6 weeks later. Thus, a neonate with a duct-dependent circulation (*e.g.* pulmonary atresia) will develop problems shortly after birth when functional closure of the duct occurs, becoming hypoxaemic, tachycardic and shocked. The treatment is to open the arterial duct pharmacologically using a prostaglandin infusion. Ventilation is frequently required due to the collapsed state and the respiratory depressant effects of prostaglandin E₂. Referral to a specialist centre for evaluation and surgical treatment follows.

**Tetralogy of Fallot**

Tetralogy of Fallot consists of a ventricular septal defect with right ventricular outflow tract obstruction, right ventricular hypertrophy and an over-riding aorta. Although the lesion can be compatible with survival to adulthood, most cases are detected and repaired in childhood. The principal symptom is cyanosis. Patients may develop hypercyanotic episodes known as ‘spells’. A hypercyanotic spell frequently starts in response to crying or emotional upset. Increased catecholamines cause muscular contraction of the right ventricular outflow tract causing an increase in the right-to-left shunt. The resulting cyanosis results in increased catecholamine secretion and acidosis, further magnifying the right-to-left shunt. Ultimately, this leads to a profound and dangerous cyanotic collapse.

The aims of treatment of a hypercyanotic spell are to decrease the shunting of blood and break the right ventricular outflow tract spasm. Patients spontaneously find that kinking of the femoral arteries (squatting) is beneficial and this may be reproduced by pressure on the femoral arteries by an assistant. Oxygen (100%) should be administered, although this will do little alone to remedy the situation. Reduction in the right-to-left shunt is achieved by increasing the systemic vascular resistance with vasoconstrictors and fluid boluses. The right ventricular outflow tract obstruction is relieved by reducing catecholamine production using morphine as sedation. β-Adrenoceptor blockade may be used to prevent further attacks, pending surgery.

**Obstetric interventions**

The presence of pulmonary hypertension is particularly important as maternal mortality approaches 30% if there is significant pulmonary vascular disease. Of women with cyanotic heart disease, 50% will suffer functional deterioration during pregnancy compared with 15% of those with acyanotic defects. Likewise, 30% of those with existing cardiac failure will deteriorate compared with 5% of those without.
Women who can be expected to labour in an essentially normal manner include those with surgically corrected lesions who are asymptomatic. Those conditions which are palliated may tolerate the stress of pregnancy badly. Valvular stenosis results in a limited functional reserve whereas regurgitant lesions with good myocardial function are better tolerated.

Anaesthetic management involves preservation of ventricular function, maintenance of adequate preload, and prevention of undesirable changes in magnitude or direction of intracardiac shunts. Vaginal delivery is the aim, but Caesarean section may be required if adverse fluctuations in cardiac output and blood pressure occur during active labour, in addition to possible obstetric indications.

Extradural block, produced slowly and incrementally with appropriate fluid loading and vasopressor use, can minimise rapid fluctuations in preload, cardiac output and heart rate. However, this may be undesirable in the fixed cardiac output state associated with severe aortic valve disease when general anaesthesia may be safer with modification of the technique in order to reduce the response to tracheal intubation and ventilation. Before regional techniques are employed, defective haemostasis should be considered. The use of endocarditis prophylaxis should also be remembered.

The patient with a single ventricle
Severe forms of congenital heart disease may result in a single ventricle (e.g. hypoplastic left heart syndrome, or severe forms of pulmonary atresia with intact ventricular septum). Surgery, performed shortly after birth, aims to provide pulmonary blood supply from a shunt from the subclavian artery to the pulmonary artery. As blood mixes from the systemic and pulmonary veins, a degree of cyanosis is inevitable. Anaesthetists faced with a child with this ‘Blalock-Taussig’ shunt must accept a degree of cyanosis and manipulate pulmonary blood flow using systemic vasoconstrictors.

As the child grows, pulmonary blood flow decreases relative to the size of the child. Cyanosis worsens and further surgery is performed connecting first the superior vena cava and then, later, the inferior vena cava to the pulmonary artery. The subclavian artery shunt is taken down and blood flow is then passive to the lungs. The final circulation, where blood is pumped from a single ventricle to the systemic circulation and then passive venous return carries de-oxygenated blood to the lungs and back to the heart, has become known as the ‘Fontan’ circulation. Successful circulation is dependent on good ventricular function and low pulmonary vascular resistance. Blood flow to the lungs is augmented by the negative intrathoracic pressure developed during active inspiration.

Anaesthesia in these patients should aim to maintain good ventricular function, low pulmonary artery pressure and, if at all possible, maintain the patient’s own respiratory efforts. Ultimate outcome for these patients is partly dependent on the morphology of the ventricle. Left ventricles being much better able to cope with systemic pressures than morphological right ventricles. Patients with the Fontan circulation are at high risk of arrhythmias and ventricular failure.

Recipients of a heart transplant
The anaesthetic assessment and management of patients who have received a heart transplant was described recently in this journal (see key references).

Conclusions
With increasing numbers of patients with congenital heart disease living to middle age and beyond, and the increased number of heart transplant recipients, the chance of being called upon to anaesthetise for non-cardiac surgery is increased. Anaesthesia may safely be performed in patients with corrected congenital heart disease in a non-specialist centre. More problems may be expected in patients after palliative surgery, where haemodynamics will be grossly abnormal. These patients and those with new or rapidly progressive symptoms after corrective surgery, should be referred to a specialist cardiology centre before all but the most urgent surgery. In all cases, the management strategy involves an understanding of the anatomy of the underlying defect and its pathophysiological effects.

Key references
See multiple choice questions 108–111.